Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 5: Skull Base

Chapter 53: Otologic Manifestations of Retrocochlear Disease

Timothy T. K. Jung, Rick L. Nissen

Lesions in cranial nerve VIII, in the cerebellopontine angle, and along the central auditory and vestibular pathway can produce otologic symptoms such as hearing loss, tinnitus, disequilibrium, and vertigo. These retrocochlear abnormalities may be congenital, infections, neoplastic, vascular, traumatic, metabolic, toxic, demyelinating and degenerative, or idiopathic in nature (Table 1). Some of these diseases may involve both central and peripheral auditory and vestibular pathways. The term *retrocochlear* is applied broadly in this chapter to include some of the systemic diseases affecting auditory and vestibular end-organ as well as central connections. Recent advances in audiometric diagnostic methods and neuroradiologic imaging technology made it possible to localize the lesion and to correlate with otologic symptoms much more accurately than previously. The purpose of this chapter is to describe otologic manifestations, diagnosis, and management of these retrocochlear diseases.

Evaluation of Patients for Retrocochlear Disorders

When a patient presents with hearing loss, tinnitus, or dizziness, evaluation should proceed in a logical and practical manner to rule out a retrocochlear lesion. The evaluation begins with the history, followed by physical examination, audiologic and vestibular testing, other laboratory testing, and radiographic evaluations, as necessary.

History

Retrocochlear lesions produce auditory and vestibular symptoms and careful history can provide clues that indicate the possible presence and location of central lesions. Unfortunately, there is no uniform history that is typical for retrocochlear lesions. In general, otologic manifestations of retrocochlear lesions are similar to those of peripheral lesions.

In the pediatric population, gestational and perinatal history is of special importance. Prenatal infections, such as rubella, toxoplasmosis, influenza, cytomegalovirus infection, and syphilis, can cause hearing impairment (Bergstrom and Stewart, 1971). Certain medications, such as streptomycin, quinine, and chloroquine phosphate, can have a deleterious effect on the developing ear. Asphyxia and anoxia during birth may lead to hearing loss through toxic damage to the cochlear nuclei (Hall, 1964). Kernicterus may cause damage to the cochlear nuclei or other central auditory pathways (Crabtree and Gerrard, 1950; Swaiman and Wright, 1983).

Table 1. Retrocochlear Disorders With Otologic Manifestations

Congenital Disorders

Usher syndrome
Maternal rubella
Prematurity
Kernicterus
Birth trauma
Arnold-Chiari syndrome

Infectious Disorders

Meningitis
Encephalitis
Brain abscess

Neoplastic Disorders

Glioma
Medulloblastoma
Astrocytoma
Ependymoma
Acoustic neuroma
Meningioma
Cholesteatoma
Facial nerve schwannoma
Metastatic tumors

Vascular Disorders

Vertebrobasilar artery insufficiency
Lateral medullary syndrome
Lateral pontomedullary syndrome
Vascular loop and other disorders

Traumatic Disorders

Skull fracture
Concussion
Cervical vertigo (whiplash injury)

Metabolic and Toxic Disorders

Diabetes mellitus
Thyroid dysfunction
Malnutrition
Drugs

Demyelinating and Degenerative Disorders

Multiple sclerosis
Spinocerebellar degenerations

Other Retrocochlear Disorders

Epilepsy
Autoimmune disease.

A slowly progressive unilateral hearing loss is one of the most frequent presenting signs of retrocochlear lesion. The hearing loss can be fluctuant or sudden in onset. The patient may complain of difficulty in understanding words rather than in hearing actual sounds. Tinnitus is commonly present with unilateral hearing loss. Acceptance of the tinnitus by the patient varies widely.

Most patients with retrocochlear lesions have a mild balance disturbance with a sensation of unsteadiness, and not the violent vertigo typically present in peripheral disorders. Vertigo

present in central disorders is characterized by the lack of latency and fatigability. Patients may have headache, gait disturbance, hypesthesia of the face, otalgia, nausea, or facial palsy.

Physical Examination

A complete head and neck and otoneurologic examination is performed. The physical examination frequently is normal. The corneal reflexes may be reduced with decreased facial tactile sensation. Facial function should be noted and the cerebellar function test should be done.

Audiologic and Vestibular Tests

Basic Audiometric Battery. Basic hearing tests include air, bone, speech reception threshold, and speech discrimination. Typical hearing loss in a patient with a retrocochlear lesion is a unilateral sensorineural hearing impairment with a poor speech discrimination score. The retrocochlear audiologic test battery formerly included short-increment sensitivity index (SISI), tone decay tests, and Békésy audiometry. These tests are not always reliable, and auditory brain-stem response (ABR) audiometry and acoustic reflex tests have replaced them.

Acoustic Reflex Tests. In acoustic reflex tests, reflex threshold, decay, and latency are measured. Absent or elevated acoustic reflex threshold in the absence of a significant hearing loss, or the presence of reflex decay (50 per cent decay from the initial reflex amplitude over a 10-second period) may indicate a retrocochlear lesion (Jerger and Jerger, 1975). Of patients with confirmed acoustic tumors 88 per cent have either pathologic acoustic reflex decay or absence of the acoustic reflex. One drawback of this test is that it has a high false-positive rate (approximately 30 per cent) (Sheehy and Inzer, 1976).

Auditory Brain-Stem Response Audiometry. Auditory brain-stem response (ABR) audiometry is sensitive to intra- and extra-axial tumors, as well as other neural lesions such as multiple sclerosis and vascular lesions such as arteriovenous malformation or stroke. Retrocochlear pathology is suspected when any of the following quantitative measurements are present: (1) abnormally prolonged peak latencies, (2) abnormal peak interval measures (I-V interval of 4.4 msec), (3) interaural latency differences of wave V greater than 0.4 msec, and (4) V/I amplitude ratios less than 0.5 (Hosford-Dunn, 1985). ABR is the most sensitive and specific test in the audiology battery for detecting disorders of the brain stem. For example, 98 per cent of patients with confirmed acoustic neuromas have positive findings based on the interaural latencies or the central conduction time (Barrs et al, 1986).

Vestibular Tests. On alternate bithermal caloric testing, a difference of 30 per cent is considered a significant reduction of response. Only 80 per cent of patients with acoustic neuroma had a significantly reduced caloric response (Linthicum et al, 1979). Because of relatively low sensitivity, ABR audiometry and acoustic reflex tests have reduced the need for vestibular tests.

Central Auditory Perceptual Tests. Central auditory perceptual tests (CAPTs) help to evaluate central auditory pathways. The integrity of the central nervous system can be assessed by the use of a dichotic speech test such as the staggered spondiac word (SSW) test (Katz, 1968), the synthetic speech sentence identification (SSI) test (Jerger and Jerger, 1974), and the binaural fusion and rapidly alternating speech tests (Willeford, 1976). Speech tests presented monaurally, such as the filtered speech test (Willeford, 1976), and other distorted monosyllable work materials such as speech in noise are helpful in the diagnosis of central auditory disorders.

Radiologic Examination

Conventional Radiography. Plain (Stenvers' or transorbital view) or polytomographic studies of the internal auditory canals are obtained for screening purposes. Bony erosions and widening of the canal suggest existence of acoustic neuroma.

Computed Tomographic Scans. Computed tomographic (CT) scans provide a definitive diagnosis of retrocochlear lesions. CT scans have replaced both polytomography and isophendylate (Pantopaque) myelography of the cerebellopontine angle. CT pneumocisternography is used to detect intracanalicular tumors.

Magnetic Resonance Imaging. Magnetic resonance imaging (MRI) is the study of choice for retrocochlear pathology occurring in the internal auditory canals, cerebellopontine cisterns, and brain stem (Curati et al, 1986; Valvassori, 1986). MRI may well replace CT scans in the diagnosis of retrocochlear lesions. For the assessment of bony lesions and peripheral auditory and vestibular disorders, CT scans remain the best study.

Other Laboratory Studies

Cerebrospinal fluid analysis for protein in the diagnosis of acoustic neuroma is used infrequently. When a child is suspected of having congenital hearing loss, the following laboratory studies may be helpful to find the cause: (1) antibody assay for toxoplasmosis, rubella, cytomegalovirus, and herpes virus (TORCH studies); (2) VDRL (Venereal Disease Research Laboratory) or FTA (fluorescent treponemal antibody) tests when congenital syphilis is suspected; (3) thyroid function tests when Pendred's syndrome is suspected; and (4) electrocardiography (ECG) when Jervell and Lange-Nielsen syndrome is suspected.

Congenital Disorders

The majority of congenital disorders that accompany otologic symptoms are associated with peripheral or cochlear abnormalities. Congenital sensorineural deafness can be due to either an abnormal bony labyrinth or abnormal neuroepithelium (organ of Corti). An abnormal bony labyrinth is frequently associated with abnormalities of central nervous system (Ruben, 1983). It has been shown that destruction of the inner ear or sound deprivation can cause changes in the central auditory pathways (Webster and Webster, 1977). During the developmental stage there seems to be an interaction between the cochlear and retrocochlear auditory pathway. Congenital

disorders are divided into either genetic or acquired. It is estimated that of all congenital deafness, about 50 per cent is due to acquired disease, 34 per cent to autosomal recessive inheritance, 15 per cent to autosomal dominant inheritance, and 1 per cent to X-linked inheritance.

Genetic Disorders

More than 70 different genetic syndromes are associated with congenital deafness (Konigsmark and Gorlin, 1976). Most of them are rare and are associated with abnormalities of either the bony labyrinth or the organ of Corti. Only the more common types with retrocochlear abnormalities are discussed here. The most common types of genetic congenital deafness found in humans are Waardenburg's syndrome, Usher's syndrome, Pendred's syndrome, and the Jervell and Lange-Nielsen syndrome. In all of these, except for Usher's syndrome, the pathology is primarily in the cochlea. Usher's syndrome is transmitted as an autosomal recessive and consists of congenital sensorineural deafness, progressive retinitis pigmentosa, night blindness, tunnel vision, cataracts, vestibular impairment, mental retardation, loss of olfaction, aphasia, psychosis, spinocerebellar ataxia, and nystagmus (Nuutila, 1970). The histopathology shows degeneration of the organ of Corti and spiral ganglion cells, mainly in the basal turn (Belal, 1975). Retrocochlear defect with atrophy of auditory portion of the central nervous system was noted in Usher's syndrome (Vernon, 1969).

Acquired Disorders

Acquired congenital deafness is usually caused by injury to the developing fetus. Two major causes of perinatal injury are the ingestion of various ototoxic and teratogenous substances and intrauterine infection. Well-known ototoxic substances include streptomycin, quinine, and chloroquine phosphate. Common intrauterine infections that cause deafness include rubella, toxoplasmosis, cytomegalovirus, and syphilis. Other common causes of acquired congenital deafness are meningitis, prematurity, kernicterus, and birth trauma (Vernon, 1967). These intrauterine and perinatal injuries affect multiple areas of the body, including retrocochlear and cochlear sites and the central nervous system. The chance of deafness in premature infants is 20 times greater than that in the child with normal birth weight (Ruben, 1983). The underlying mechanism of sensorineural hearing loss in the premature infant is suspected to be from central nervous system pathology, as is hearing loss with kernicterus (Dublin, 1976). There are changes throughout the central nervous system, with the greatest amount of nerve cell injury in the ventrocochlear nucleus of the central auditory pathway.

Arnold-Chiari Syndrome

Malformation of the cranial vertebral junction of the base of the skull may cause herniation of cerebellar tonsils and can cause vestibular symptoms and hearing loss. Some patients may develop ataxia and long tract signs. Surgery may alleviate some of the symptoms by relieving the herniation (Longbridge and Malinson, 1985).

Management of Congenital Deafness

The prompt diagnosis of congenital deafness after a thorough evaluation of the patient is the most important step in management. Once the diagnosis is made, management is directed toward education, habilitation, and genetic counseling.

Infectious Disorders

Infectious disorders of central nervous system, such as meningitis, encephalitis, and brain abscess, can cause hearing loss and vertigo. Infection may involve only the central nervous system or may extend to the inner ear through a hematogenous or preformed route. Since the inflammatory process involves wide areas of the central nervous system, specific otologic symptoms may not be apparent until late in the course of the infection.

Meningitis

Meningitis is one of the most common causes of unilateral or bilateral sensorineural hearing loss in childhood. The incidence of hearing loss due to meningitis in a retrospective study of 185 children was 31 per cent with *Streptococcus pneumoniae,* 10.5 per cent with *Neisseria meningitidis,* and 6 per cent with *Haemophilus influenzae* infections (Dodge et al, 1984). Meningitis was reported by Vernon (1967) to be the cause of deafness in 20 per cent of all deaf children.

The most common organism that causes purulent meningitis is *Haemophilus influenzae* (Gower and McGuirt, 1983). Others pathogens frequently found are *S. pneumoniae, N. meningitidis, Staphylococcus aureus,* beta-hemolytic streptococci, and *Escherichia coli.* The early clinical manifestations of meningitis are headache, fever, neck rigidity, and abnormal reflexes. Definite diagnosis is made by a lumbar puncture and analysis of the cerebrospinal fluid. Because the auditory and vestibular damage from meningitis is irreversible, every effort should be exerted to prevent meningitis or to treat it vigorously.

Patients with meningitic deafness almost invariably have an absence of vestibular function. Other severe handicaps such as cerebral palsy, mental retardation, and visual pathology may result from meningitis.

The histopathology of hearing loss following meningitis has been described (Schuknecht, 1974). The bacteria from meningitis can invade the inner ear through the nerves, vessels, cochlear aqueduct, endolymphatic duct and sac, and possibly the modiolus. Once bacteria invade the inner ear, labyrinthitis follows. The membranous labyrinth is destroyed and replaced by granulation tissue and eventually by new bone (labyrinthitis ossificans).

Temporal bone histopathology in deafness caused by cryptococcal meningitis showed the primary site of invasion at the cochlear nerve in the internal auditory canal and the modiolus with the loss of ganglion cells and nerve fibers (Harada et al, 1979).

Other organisms may cause meningitis besides the usual bacteria discussed above. Although uncommon, tuberculous meningitis is associated with a high incidence of deafness (Lorber, 1961). Viral meningitis, although usually benign, can cause hearing loss and other complications in the central nervous system. Viruses that may cause meningitis include mumps, herpes zoster, coxsackievirus, herpes simplex, and enterovirus (Nadol, 1978).

One example of viral infection with otologic manifestations is herpes zoster oticus (Ramsay Hunt syndrome). This syndrome presents with facial paralysis, vesicular rash of the auricle, and occasional hearing loss, tinnitus, and vertigo. It is believed that herpetic meningitis and neuralgia of the sensory ganglia and nerve roots cause the syndrome. Histopathologic findings in a case of Ramsay Hunt syndrome included perivascular, perineural, and intraneural round cell aggregations in the facial nerve, cochlea, and mastoid process is symptomatic. The facial nerve paralysis may progress to the point that facial nerve decompression may be necessary.

Encephalitis

Encephalitis is an inflammation of the brain and may be caused by a multiplicity of agents including viruses, spirochetes, bacteria, mycobacteria, fungi, and protozoa.

Viral Encephalitis

Mumps is the single most common causative agent of viral encephalitis (Johnson, 1982). In the prenatal period, rubella virus and cytomegalovirus are common viral agents. Viral encephalitis is characterized by alteration in consciousness in addition to headache, fever, and nuchal rigidity. It may progress to mild lethargy, confusion, stupor, and coma. Paralysis of cranial nerves III and VI may develop, owing to increased intracranial pressure. Auditory and vestibular defects may result from neuritis of cranial nerve VIII, especially with mumps (Krugman and Ward, 1968). Labyrinthitis may accompany viral meningoencephalitis (Lindsay et al, 1960). Although infections with mumps, measles, and rubella usually produce a permanent deafness, other forms of viral encephalitis may be associated with only mild symptoms of central nervous system involvement.

Infrequently, systemic viral infections or administration of certain vaccines give rise to central nervous system abnormalities that do not appear to depend on direct invasion of the brain, but rather to be the result of autoimmune or toxic mechanisms. Examples of this type of disease are acute disseminated encephalomyelitis, acute hemorrhagic encephalomyelitis, and acute inflammatory polyneuropathy (Guillain-Barré syndrome). Auditory or vestibular nerves may be affected by similar autoimmune or toxic mechanisms following viral infections. Several cases of delayed onset of Ménière's disease with hearing loss and intermittent vertigo after several years following viral infection have been reported (Schuknecht and Gulya, 1983). Mild to moderate sensorineural hearing loss was seen in a patient who was given influenza virus vaccine several weeks before. Hearing returned after a short course of treatment with corticosteroids (author's unreported case).

Neurosyphilis

Infection of the leptomeninges or brain parenchyma with *Treponema pallidum* causes neurosyphilis. Neurosyphilis may cause a variety of auditory and vestibular abnormalities.

Approximately 70.000 or more persons develop primary syphilis and 13.000 persons develop secondary syphilis per year in the USA, as of 1980 (Johnson, 1985). Neurosyphilis may develop within 1 to 3 years following acute infection, especially after inadequate treatment. The clinical presentation and pathology of neurosyphilis vary widely. It may present with acute meningitis, a meningovascular form with arteritis and infarction of neural tissue, general paresis, tabes dorsalis, and rare granulomatous accumulations (gumma). Among the cranial nerves, nerve VIII is most frequently affected by neurosyphilis, resulting in deafness and dizziness.

The diagnosis of neurosyphilis should be suspected in any patient with sensorineural deafness associated with other stigmata of syphilis infection, such as Hutchinson's teeth and interstitial keratitis. The hearing loss may be bilateral or asymmetric. Vertigo may accompany hearing loss, and caloric tests may be hypoactive (Karmody and Schuknecht, 1966). A positive fistula test may be elicited in the absence of a fistula (Hennebert's sign). In fact, syphilis infection of inner ear causes endolymphatic hydrops and often presents symptoms indistinguishable from Ménière's disease.

The laboratory diagnosis of neurosyphilis is made by the analysis of cerebrospinal fluid and serum. Changes in cerebrospinal fluid include increased lymphocytes, proteins, and immunoglobulin G and occasional presence of spirochetes detected by the immunofluorescent method. The serologic tests for syphilis are divided into two types: nontreponemal or specific treponemal tests. A commonly used nontreponemal test is the VDRL test. The specific treponemal serologic tests include the readily available fluorescent treponemal antibody-absorbed (FTA-ABS) test and the treponemal immobilization test (TPI), which is not routinely available. The nontreponemal test is used for screening purposes, and the specific test is used for confirmation. The FTA-ABS test is frequently used in a deaf or dizzy patient suspected of having neurosyphilis, because this test remains positive even when the VDRL test is negative. Since there are false-positive results using the FTA-ABS test, it should be repeated, and when it is weakly positive, TPI testing should be done.

Penicillin is the antibiotic of choice in the treatment of neurosyphilis. Because the organism divides slowly and antibiotic is effective only during the dividing stage, prolonged antibiotic therapy is necessary for a cure. In addition to antibiotics, corticosteroid is recommended to improve hearing. Medications are discontinued if there is no improvement in hearing after one month of treatments (Hendershot, 1978).

Brain Abscess

Brain abscess is a localized collection of encapsulated or free pus in the parenchyma of the brain. Brain abscess can occur by extension from infections within the cranium, by

introduction of bacteria from head trauma, or by hematogenous infection from other parts of the body. The primary infection can be from otitis media, sinusitis, pulmonary disease, cardiac disease, or generalized sepsis. Otogenic brain abscesses are usually located in the temporal lobe cerebellum. Approximately 0.5 per cent of patients with acute otitis media and 0.3 per cent of patients with chronic otitis media will develop brain abscess (Harter, 1985). When there is bony erosion at the attic or mastoid antrum and the middle fossa dural plate is eroded, otitis media can extend into the temporal lobe and cause an abscess. In cerebellar abscess, there is often evidence of retrograde thrombosis from the lateral sinus. Paranasal sinus infection accounts for about 5 to 10 per cent of all brain abscesses. Frontal sinusitis can extend into the anterior part of the frontal lobe, and infection from the ethmoid sinus can cause deep temporal lobe infection.

Microorganisms responsible for brain abscesses include (1) gram-positive cocci (group A *Streptococcus, S. pneumoniae, S. viridans,* and *Staphylococcus aureus*), (2) gram-negative bacteria (*Haemophilus influenzae*), (3) gram-negative enteric bacilli (*Escherichia coli, Proteus species,* and *Pseudomonas aeruginosa*), and (4) anaerobic bacteria (*Bacteroides* species) (Heineman and Braude, 1963).

The symptoms of brain abscess are generally the same as far as space-occupying intracranial lesions, including fever, headache, nausea or vomiting, increasing obtundation, seizures, and localizing neurologic findings. Depending on the site of involvement, focal signs may develop. Temporal lobe abscess may cause aphasia, paresis of the opposite side of the face and mouth, and visual field defects. Cerebellar abscess may produce ataxia on the same side, intention, tremor, and vertical or oblique nystagmus (Shambaugh, 1967).

The treatment of brain abscess consists of early diagnosis, antimicrobial therapy, corticosteroids to reduce brain edema, and evacuation or resection of the abscess. The common use of CT scans has greatly improved the diagnosis and treatment of brain abscess. CT scans have made it possible to treat more brain abscesses medically, because they can be monitored closely. When the source of the brain abscess is found, it should be removed whenever possible - for example, mastoidectomy for the otogenic brain abscess and frontal sinus trephination for the abscess originating in the frontal sinuses. The mortality rate from all brain abscesses is approximately 30 per cent (Morgan et al, 1973).

Neoplastic Disorders

Tumors of the Central Nervous System

Neoplasms of the central nervous system are capable of producing symptoms similar to those for neoplasms of a retrocochlear origin. Depending on their location, above or below the tentorium, they may produce early vertigo and deafness or manifest vague auditory-vestibular symptoms. Spontaneous or vertical nystagmus with calorically induced optokinetic directional preponderance may be present with central nervous system neoplasms; however, these tests are of limited value in localizing the problem (Fredrickson et al, 1969). Usually, central nervous system tumors will have other neurologic signs such as weakness in the extremities, paresthesias,

deranged sensorium, or symptoms suggestive of a mass effect. Hence, as opposed to tumors located in the cerebellopontine angle that produce the "syndrome of the cerebellopontine angle", patients with central nervous system tumors will not usually present initially to the otolaryngologist.

In the cerebellum, the more common tumors are the medulloblastoma, astrocytoma, and ependymoma. In this location, a slowly progressive vertigo, ataxia, or incoordination may be present. With obstruction of the cerebrospinal fluid circulation, there will be signs of increasing intracranial pressure. When they occur in children, most of these tumors are treated with surgery, occasionally with combined radiotherapy (Dekaban, 1970).

Supratentorial tumors with vague auditory-vestibular symptoms usually have associated symptoms, depending on their location. Thus, tumors in the frontal lobe usually have associated personality changes and mental symptoms, whereas tumors in the temporal lobe frequently stimulate seizures and visual/olfactory hallucinations. Tumors in the occipital lobe often have associated visual field defects and visual hallucinations. Most of these central nervous system tumors are diagnosed and treated by specialists in neurosurgery/neurology.

Tumors of the cerebellopontine angle, on the other hand, are usually accompanied by more initial and definitive auditory-vestibular signs and symptoms. Hence, these patients are often initially seen, diagnosed, and treated by the otolaryngologist or otologist. These tumors include the acoustic neuroma, meningioma, glioma, cholesteatoma, metastatic tumor, cholesterol granuloma, and arachnoid cyst.

Tumors of the Cerebellopontine Angle

Acoustic Neuroma

The acoustic neuroma is by far the most common lesion of the cerebellopontine angle and thus deserves special emphasis. In a series of 1354 cerebellopontine angle tumors, 90 per cent were acoustic neuromas (Brackmann and Bartels, 1980). Acoustic neuromas reportedly account for about 9 per cent of all intracranial tumors and have an incidence of approximately 2.4 per cent in the general population (Glasscock, 1980).

Acoustic neuromas are benign encapsulated tumors that usually arise from the Schwann cells of the superior division of the vestibular nerve. Less frequently, they may arise from the inferior vestibular nerve; rarely, they arise from the cochlear nerve. The usual site of origin is within the internal auditory canal in the region of Scarpa's ganglion where the neurilemmal sheath terminates (Nager, 1969). As the tumor slowly enlarges, growth is toward the cerebellopontine angle with widening of the canal and involvement of cranial nerve VIII by compression. Occasionally, the tumor may originate within the angle and may spare cranial nerve involvement until the tumor reaches a large size.

Histologically, the tumor is composed of streams of elongated spindle cells, with elongated nuclei, arranged in a parallel fashion. Each group spreads in a different direction, so that the appearance is that of latticework. Tumors in which there is a thick concentration of cells are called Antoni type A, whereas those with a loose arrangement are called Antoni type B (Nager, 1969). Usually there is a combination of the two types within a given tumor. Degenerative changes with cyst formation often occur as the tumor enlarges.

Symptoms

An acoustic neuroma produces symptoms and findings by pressure on nerve tissue and occasionally on blood vessels (Sheehy, 1979). Initial symptoms are usually referred to the auditory-vestibular apparatus and consist of tinnitus, hearing loss, and dizziness. The natural history of acoustic tumors is one of slowly progressive growth (growth rate of 0.2 cm per year) (Silverstein et al, 1985), and symptoms have a slow, insidious onset.

Unilateral sensorineural hearing loss and tinnitus are common presenting complaints, usually associated with poor speech discrimination on the involved side. Distortion of hearing using the telephone is a common complaint. In general, auditory symptoms are unilateral and progressive. Occasionally, auditory symptoms present as a sudden sensorineural hearing loss. It is thought that this may result from interference of the blood supply and, as such, may fluctuate or seem to respond to medical treatment (Sheehy, 1979).

Dizziness is a common complaint in patients with acoustic tumors. It usually appears in the form of unsteadiness (Pulec et al, 1964). This has been reported in 70 to 80 per cent of these patients. True vertigo is much less frequent, because vestibular destruction is due to slow growth of the tumor, allowing time for compensation by the patient (Hitselberger and House, 1971). Occasionally, the vertigo presents with paroxysms, as in Ménière's disease.

If the tumor has its origin in the cerebellopontine angle, tinnitus and hearing loss may not become evident until the tumor reaches a large size. As it continues to grow and expand into the cerebellopontine angle, other cranial nerves and structures become involved. The sensory part of cranial nerve V is usually the first cranial nerve involved, outside of nerve VIII. This manifests as diminished or absent corneal reflex and anesthesia of the ipsilateral face. Continued growth will show involvement of cranial nerves IX, X, XI, and XII. Cranial nerve VII shows surprising resilience under even extreme pressure and stretching. The sensory division of nerve VII may show earlier involvement than the motor division, and this will manifest as anesthesia over the posterior aspect of the external auditory canal (Hitselberger's sign). Very large tumors may obstruct the flow of cerebrospinal fluid, causing headaches, nausea, vomiting, and dullness of the mental faculties (Merritt, 1959).

The important point to remember is that there is no typical clinical history. To make a diagnosis of an acoustic neuroma at an early stage, complete evaluation must be made of every patient who have suspicious inner ear symptoms.

Diagnosis

The diagnosis of acoustic neuroma is made through a thorough history, as well as audiologic, vestibular, and radiologic studies. CT scans, and more recently MRI, have become the "gold standards" for diagnosing these tumors. In today's cost-containment environment, not every patient with unilateral auditory-vestibular symptoms will undergo these tests. Hence, it is important to use preliminary screening tests with a high degree of sensitivity for retrocochlear pathology, as mentioned in the first part of this chapter. Although screening tests are extremely valuable adjuncts, of equal or greater importance is a complete, thorough history and a thorough neurotologic examination.

Audiometric Studies. The audiogram is crucial for early diagnosis, as this usually is what catches the attention of the physician initially. A hearing loss in high tones is most common (66 per cent) (Johnson, 1977) with a loss in low tones being present in only 9 per cent of cases. A flat or trough type of curve may also be seen. Poor speech discrimination is also seen commonly and leads one to be highly suspicious of an acoustic tumor. In a series of 500 patients, discrimination scores of less than 60 per cent were seen in 72 per cent of patients (Johnson, 1977).

Special audiometric tests were used in the past as initial screening tests; however, because of their low sensitivity, these have been replaced by more sensitive tests. The Short Increment Sensitivity Index (SISI) and Alternate Binaural Loudness Balance (ABLB) were used as measures of recruitment in the diagnosis of retrocochlear pathology. In a series of 500 acoustic tumor patients, both tests were positive in approximately 50 per cent of cases tested (Johnson, 1977). In the same series, Békésy recordings of type III or IV, indicative of acoustic neuromas, were found in only 60 per cent of patients. The acoustic reflex (absence or decay) as demonstrated by impedance audiometry, has proved to be the most useful of these tests, with 80 per cent of confirmed tumors having an absence or decay of the acoustic reflex (Sheehy et al, 1976). These tests are helpful but cannot be relied on as absolute proof of the presence or absence of a tumor.

One of the most sensitive screening tests for acoustic tumors is auditory brain-stem response (ABR) audiometry. This computerized, objective hearing test has been shown to have an accuracy of greater than 90 per cent for diagnosing acoustic tumors (House et al, 1979). In this test, a large series of clicks is presented to the ear being tested. A vertex electrode picks up the average electrical response as it passes through cranial nerve VIII and the central pathways, toward the auditory cortex. Pressure on the auditory nerve can cause an increase in latency of response waves reported on an oscilloscope. The waves are labeled I through V. Comparisons are made by evaluating the latency from both ears. Intra-auricular latency of greater than 0.2 millisecond is considered significant if found in the suspect ear. The main drawback of this test is that the patient must have a hearing threshold of at least 70 to 75 dB in order to "hear" the click stimuli. Advantages are that the test does not require the patient's cooperation and is a completely objective test of the auditory nerve and central pathway.

Vestibular Studies. In previous years, a unilateral reduced vestibular response was found in 90 to 95 per cent of cases of acoustic neuromas. More recently, the frequency of abnormal response has been found to be around 75 per cent (Linthicum, 1983). This drop in accuracy is directly attributable to deficiencies in the earlier diagnosis. There are now more sophisticated tests and a greater awareness of these tumors; hence, these tumors are now found at a smaller size, possibly before vestibular destruction has occurred. Further evaluation shows a reduced electronystagmographic response in 98 per cent of patients with a tumor arising from the superior vestibular nerve, whereas only 60 per cent of those with tumors of the inferior vestibular nerve have a reduced response. This is due to the fact that standard electronystagmography tests the lateral semicircular canal and hence the superior vestibular nerve. It is thought that if a test for the inferior vestibular nerve is found, the accuracy will be increased to levels found in the past. It is possible that caloric tests may be better evaluated in conjunction with rotatory tests. Methods utilizing sinusoidal angular acceleration have shown asymmetry of response in cases in which caloric tests were normal.

Radiologic Evaluation. Radiologic evaluations are the mainstay of definitive diagnosis of acoustic neuromas. Preliminary studies include plane films and polytomograms. Two commonly used views for plane film evaluation are the transorbital and Stenvers' views. As little a difference between the two internal auditory canals as a 1-mm enlargement on the symptomatic side may be significant (Glasscock, 1980). The accuracy of well-made plane films of the internal auditory canal (IAC) is approximately 90 per cent. Polytomograms (with 1-mm cuts) will show the area of the IAC with extreme clarity. In most patients, however, plane films will adequately visualize the IAC. In the recent past, radiopaque polytomograms were used to visualize filling defects at the porus acusticus and were the definitive diagnostic criterion. With the recent addition of CT scans and MRI, this test has fallen by the wayside.

CT scans have proved their importance in the diagnosis of acoustic neuromas. Since acoustic neuromas have tissue densities equal to those of surrounding brain tissues, it is important that these lesions be enhanced with a bolus of intravenous iodine-based dye (Davis et al, 1977). Many tumors will not be demonstrated on CT scans without enhancement. Tumors 2 centimeters or smaller may be missed (false-negative) even with contrast. Hence, in highly suspect patients with negative CT scans with contrast, an air CT scan is used for the definitive diagnosis.

In this procedure, a lumbar puncture is performed and 5 to 7 cc of CO_2 or O_2 is injected into the subarachnoid space. The patient is then tilted head up, and the air is allowed to enter the cerebellopontine angle and each internal auditory canal. If a tumor is present, it will bloc the entrance of air into the canal. With the recent advent of MRI, a new noninvasive method for definitely diagnosing acoustic tumors is available. In a direct comparison with CT scans of 44 patients with acoustic neuromas, MRI was able to identify all tumors with equal sensitivity (House, 1986). Again, since this is a noninvasive test, it may well become the definitive diagnostic tool of choice, especially with the use of injectable contrast agents such as gadolinium. Its main limitation at this point is with the claustrophobic patient (who has difficulty staying within the close confines of the testing environment) and with patients who have metallic implants of some type.

In summary, a reasonable workup for the patient may include an initial audiogram documenting a unilateral hearing loss. With mild or low suspicion of acoustic neuroma, ABR audiometry, plane films of the IAC, and possibly electronystagmography will be ordered. These three tests together have a sensitivity of over 90 per cent. With an abnormal response of any of the three, CT scan or MRI are ordered. In the highly suspect patient, preliminary studies may be bypassed and the evaluation proceed directly to CT scan/MRI.

Management

The management of acoustic neuromas will depend upon their size and the patient's health and age. There are many different surgical approaches to these tumors, which will be influenced by the variables. Surgical approaches are discussed in detail in the following chapter (Chap. 55).

Although acoustic neuromas account for the vast majority of lesions within the cerebellopontine angle, other lesions occasionally present in this area. In a series of 1354 cerebellopontine angle tumors, approximately 10 per cent were lesions other than acoustic neuromas (Brackmann and Bartels, 1980). In order of frequency of occurrence, these included meningiomas (3 per cent), primary cholesteatomas (2.5 per cent), facial nerve neuromas (1 per cent), neuromas of other cranial nerves in the posterior fossa. Other rare tumors (1.5 per cent) include arachnoid cysts, tumors of vascular origin, metastatic tumors, and cholesterol granuloma. In spite of their rare occurrence, familiarity with these tumors is important from a differential diagnostic and therapeutic standpoint.

Meningiomas

Fourteen per cent of intracranial tumors and about 3 per cent of cerebellopontine tumors are meningiomas. Meningiomas are benign but locally invasive tumors that arise from endothelial cells of arachnoid villi found in the walls of the cranial venous sinuses (Glasscock, 1980). Meningiomas arising on the surface of the temporal bone usually appear as a firm, well-circumscribed tumor without a capsule. Less frequently, they appear as flap plaquelike lesions within the dura. These tumors usually are very vascular and infiltrating.

Meningiomas may produce symptoms of the cerebellopontine syndrome, especially if the tumor originates in or near the internal auditory canal (Hitselberger and House, 1971). In these cases, they may be indistinguishable from an acoustic neuroma, producing unilateral hearing loss, tinnitus, and imbalance. This tumor has a greater tendency within the posterior fossa to arise outside the auditory meatus and may involve other adjacent cranial nerves earlier than cranial nerve VIII. These tumors have a greater tendency to invade the temporal bone itself, and frequently the patients will have chronic otorrhea with involvement of the temporal bone.

Diagnosis

The symptoms of meningiomas are similar to those of acoustic neuroma. Routine audiometry and acoustic reflex tests will show similar findings as the acoustic neuroma. The detection rate for meningioma using ABR audiometry or vestibular tests is lower than that for acoustic neuroma. Conventional radiography may show hyperostosis of the pyramid, but rarely widening of internal auditory canal. CT scans or MRI provides definitive diagnosis of the presence of a cerebellopontine tumor. Meningiomas are more dense in CT scans and are often demonstrated in non-iodine-enhanced scans. In CT scans, meningiomas are more sessile in their attachment to temporal bone, are not centered on the porus acusticus, and often do not enlarge IAC.

Treatment

The treatment of meningiomas is surgical removal, as they do not respond to radiation therapy. Since they have the tendency to infiltrate bone, it is often difficult to achieve total removal in some cases. In these cases, extensive surgical removal may relieve symptoms for a period of years. (See the following chapters for more extensive discussion on surgery of temporal bone lesions.)

Primary Cholesteatoma of the Petrous Pyramid

Primary cholesteatoma is a congenital lesion believed to arise from embryonic epidermal rest cells. When located deep within the petrous pyramid, it may mimic the cerebellopontine angle syndrome, with symptoms of hearing loss, tinnitus, and imbalance (Hitselberger and House, 1971). It is very common for these lesions to result in early weakness/paralysis of the facial nerve, which is different from the acoustic neuroma. The fact that these cholesteatomas usually occur in a younger age group and frequently present with facial nerve problems should help distinguish these patients from the patient with an acoustic neuroma. Again, radiologic studies in a suspect patient give presumptive evidence. In CT scans, primary cholesteatomas show less density than the brain, no enhancement with intravenous iodinated contrast material, and irregular margins and are not centered on the porus acusticus.

The usual treatment for primary cholesteatoma is surgical exteriorization through a radical mastoidectomy. If residual hearing is present and the surgeon is confident of complete removal of all epidermal cells, a more conservative procedure may be attempted (Glasscock, 1980). With erosion into the posterior or middle cranial fossa, the dura must be exposed to remove the cholesteatoma completely. Usually the dura remains intact, and it is possible to remove the matrix without entering the intradural space.

Occasionally these tumors may arise within the cerebellopontine angle outside the temporal bone (House, 1962). Usually cranial nerve V will show involvement before nerve VII, from a tumor in this location. To prevent operative complications, a thorough knowledge of the anatomy of the temporal bone is needed to remove these deep-seated lesions.

Cholesterol Granulomas

Although often confused with cholesteatomas, these lesions are a distinct, separate entity. They result as a nonspecific tissue reaction to a foreign body (cholesterol crystals). Breakdown of local tissue or blood and the products of the breakdown appear to be the main sources of cholesterol. Although most commonly seen in the mastoid, cholesterol granulomas may arise within the petrous apex, secondarily to blocked air-cell tracts in a well-pneumatized petrous apex (Gherini et al, 1985). This may be due to an obstruction of the eustachian tube, mucosal edema, fracture of the temporal bone, or cholesteatoma.

Cholesterol granulomas grow silently until they exert pressure on cranial nerves. Most commonly they will then produce symptoms of unilateral hearing loss, tinnitus, vertigo, facial twitching or weakness, and symptoms suggestive of retrocochlear pathology. Physical examination is usually unremarkable. The preoperative diagnosis is made with a CT scan. The finding of a cystic lesion of the petrous apex that does not enhance with intravenous contrast material and is isodense with brain tissue suggests cholesterol granuloma.

Treatment is surgical drainage with establishment of permanent aeration to prevent recurrence of the lesion. Aeration may be provided with a catheter from the mastoid or middle ear to the apex. With adequate drainage and aeration, it is not necessary to excise the cystic wall. The surgical approach will depend on the status of hearing and may involve translabyrinthine, infralabyrinthine, or a middle fossa approach. A follow-up CT scan at 1 year postoperatively is recommended. If air is seen in the apex, there is little chance of recurrence; with no air, recurrence is possible and a CT scan should be performed yearly.

Facial Nerve Schwannoma

Facial nerve schwannomas originate from Schwann cells and may develop anywhere along the course of the facial nerve. Symptoms of facial nerve schwannomas depend on the location of the tumor. There may be conductive hearing loss when tumors are located within the middle ear or mastoid, and symptoms similar to those of acoustic neuroma when tumors are located in the internal auditory canal or in the cerebellopontine angle. Usually, facial palsy does not appear until the tumors become large. Facial twitching may be present.

Diagnosis is made from the history, physical findings, audiometry, auditory reflex tests, ABR audiometry, and radiographic studies. Most of the diagnostic studies show findings similar to those for an acoustic neuroma. Polytomography may show abnormality of the fallopian canal. CT scans will demonstrate identical findings as an acoustic neuroma when it is located in the cerebellopontine angle or may show enlarged geniculate ganglion and fallopian canal.

Treatment is surgical excision through mastoidectomy or a translabyrinthine, suboccipital, or middle fossa approach, depending on the tumor's location. A small tumor can be dissected off from the nerve. In the majority of cases, the facial nerve has to be excised with the tumor and repaired with a cable graft.

Metastatic Tumors

The most common sites of origin of metastatic tumors to the temporal bone, in order of frequency, include the breast, kidney, lung, stomach, larynx, prostate, and thyroid gland (Hughes, 1985). Frequently, the duration of retrocochlear symptoms will be relatively short and can include all the symptoms of the cerebellopontine angle syndrome. The rapid development of hearing impairment in association with other cranial neuropathies and/or brain stem dysfunction suggests a malignant neoplasm of the posterior fossa (Brackmann, 1980). These symptoms, when seen with a past history of another malignant tumor, are highly suspicious for metastatic tumor. Usually CT scans will identify the lesion. Frequently, hearing is poor and, when this is the case, the translabyrinthine approach offers the lowest morbidity and greatest ease for obtaining a biopsy for diagnosis. If the preoperative hearing is good, an initial retrolabyrinthine approach may be used to establish a diagnosis and preserve hearing. The usual treatment will be to establish the diagnosis, to partially remove or debulk the tumor, and to give postoperative radiation. Prognosis is poor with or without treatment.

Miscellaneous Tumors

Other, rare types of tumors may originate in the cerebellopontine angle or petrous apex and produce the symptoms of the cerebellopontine angle syndrome. These include other cranial nerve neuromas, which may grow to considerable size before producing symptoms related to compression of cranial nerve VIII. An arachnoid cyst may produce symptoms similar to those for an acoustic neuroma. Often a CT scan will show such a mass of less density than that of brain tissue. It is not necessary to remove the entire arachnoid cyst wall, although permanent drainage is important. Deciding between a translabyrinthine and a retrolabyrinthine approach will depend on the residual level of hearing.

Vascular tumors such as hemangiomas have also been reported in the retrocochlear area. It has been reported that these patients develop retrocochlear symptoms more rapidly than patients with acoustic neuromas (Sundresan, 1976). The severity of symptoms and signs, such as facial weakness or fasciculations, is out of proportion to those expected with a small tumor. It is recommended that these vascular tumors be removed, if it can be accomplished without risk of major neurologic injury. CT scans offer a means of following those patients in whom total removal is not accomplished.

Other rare tumors reported within the cerebellopontine angle include dermoids, leiomyomas, medulloblastomas, chordomas, chondrosarcomas, and teratomas.

Vascular Disorders

Cerebrovascular disease represents the most common devastating disease affecting the central nervous system. Among the white population in the USA, the annual incidence rate is between 1 and 2 per 1000, and the death rate is between 0.5 and 1 per 1000. It is the third leading cause of death, ranking behind heart disease and cancer. The two major causes of

cerebrovascular accident are ischemia and hemorrhage. Cerebral ischemia may be caused by atherothrombosis or embolism. The injury caused by ischemia or hemorrhage in specific areas of the brain will produce specific signs and symptoms. The pathologic processes can result in a permanent or a temporary loss of function. Many of these injuries are associated with otologic manifestations. As they have with many other diseases of the central nervous system, CT scans and MRI have revolutionized the diagnosis and treatment of cerebrovascular disorders.

Vertebrobasilar Artery Insufficiency

Vertebrobasilar artery insufficiency is usually caused by atherosclerosis of the subclavian, vertebral, and basilar arteries. This is a common cause of vertigo in the elderly (Fischer, 1967). Vertigo in vertebrobasilar artery insufficiency is abrupt in onset and lasts several minutes to hours. It is frequently associated with nausea, vomiting, visual disturbances, drop-attacks, weakness, visual field defects, diplopia, dysphagia, and headache. The ischemia may affect the vestibular nuclei or the neural elements of labyrinth (Toglia, 1967). However, there is no specific pattern of abnormality on vestibular tests diagnostic of vertebrobasilar insufficiency. These symptoms may occur alone or with vertigo. Episodes of vertebrobasilar artery insufficiency may be precipitated by postural hypotension or by mechanical compression from cervical spondylosis (Naritomi et al, 1979). The subclavian steal syndrome caused by occlusion or stenosis of the subclavian or innominate artery just proximal to the origin of the vertebral artery can cause vertebrobasilar insufficiency by siphoning of blood down the vertebral artery from the basilar systems to the upper extremities. Exercise of the upper extremities can precipitate vertigo and other symptoms of vertebrobasilar artery insufficiency.

Treatment consists of symptomatic care and antiplatelet drugs such as aspirin and dipyridamole (Persantine) to prevent a subsequent cerebrovascular accident.

Lateral Medullary Syndrome (Wallenberg's Syndrome)

Lateral medullary syndrome, or Wallenberg's syndrome, is caused by infarction of a wedge at the dorsolateral medulla just posterior to the olive and caudal to the cochlear nuclei. The syndrome is due to the occlusion of the ipsilateral vertebral artery and rarely from occlusion of the posterior inferior cerebellar artery (Fisher et al, 1961).

The signs and symptoms of this syndrome include sudden onset of vertigo, hypesthesia of the same side of the face and contralateral side of the body, ipsilateral Horner's syndrome, diplopia, dysphagia, and dysphonia. Vestibular tests may show a horizontal or rotatory nystagmus to the side of the lesion, depressed ipsilateral caloric response, and directional preponderance. Hearing usually remains intact.

Lateral Pontomedullar Syndrome

Occlusion of the anterior inferior cerebellar artery causes infarction of the dorsolateral pontomedullary region and the inferolateral cerebellum (Adams, 1943). Since the labyrinthine artery arises from the anterior inferior cerebellar artery, this syndrome is strongly associated with otologic symptoms. Severe vertigo, nausea, vomiting and ipsilateral sudden hearing loss are the initial prominent symptoms. Other associated symptoms include tinnitus, ipsilateral facial palsy, ipsilateral cerebellar dysfunction, and loss of pain and temperature sensation on the same side of the face and the opposite side of the body. Vertigo may persist for several weeks to months. Vestibular testing usually shows depressed response of the involved side. Isolated labyrinthine (internal auditory) artery occlusion is an uncommon lesion. If this happens, sudden hearing loss and vertigo occur, resembling a peripheral labyrinthine disorder.

Vascular Loop and Other Disorders

A vascular loop in the internal auditory canal pressing upon the vestibular nerve was found to be the cause of episodic vertigo and severe motion intolerance (McCabe and Harker, 1983). A vestibular nerve section medial to the vascular loop offered relief of symptoms in all patients. It is possible that pressure on the vestibular nerve by an arterial loop my be the cause of intractable vertigo in many more patients (Janetta, 1975).

Aneurysms of the vertebral artery and arteriovenous malformations of the posterior fossa may mimic lesions of the cerebellopontine angle. Aneurysms of the anterior inferior cerebellar artery may present with acute subarachnoid hemorrhage due to rupture or with an insidious onset, as a cerebellopontine angle mass. Erosion of the internal auditory canal may be present, as well as hearing loss and vertigo. If an enhancing cerebellopontine angle mass appears atypical, and dynamic CT confirms rapid enhancement, vertebrobasilar angiography is essential to establish an aneurysm of the inferior cerebellar artery as the cause of the problem (Dalley et al, 1986).

Traumatic Disorders

Trauma is the leading cause of death in persons 1 to 44 years of age. Each year 50.000 Americans die as a direct consequence of traumatic brain injury, and another 50.000 to 60.000 survive severe head injury with varying degrees of disability (Becker et al, 1982). Head trauma may affect the central nervous system, cranial nerve VIII, and the inner ear to produce auditory and vestibular problems. One of the most common disabilities after head and neck trauma is dizziness. About 90 per cent of all patients note dizziness after head trauma (Linthicum and Rand, 1931). About one-half of the patients demonstrate sensorineural hearing loss, and 15 to 43 per cent of these patients have positional nystagmus (Barber, 1964).

Skull Fracture with Temporal Bone Involvement

Fractures of the base of the skull commonly involve the temporal bone. Fractures of temporal bone may be either a *longitudinal fracture,* in which the fracture runs parallel to the petrous ridge, coursing through the middle ear and tympanic ring, and sparing the labyrinth, or a *transverse fracture,* in which the fracture line crosses the petrous ridge through the internal auditory canal and/or the otic capsule. Longitudinal fractures are more common (70 to 90 per cent) (Cannon and Jahrsdoerfer, 1983), are usually caused by a blow to the side of the head, and frequently involve the tympanic membrane, ossicles, and roof of the middle ear, causing conductive hearing loss. Transverse fractures are less common (20 to 30 per cent), are usually caused by a blow in the occipital or frontal area of the head and involve internal auditory canal and labyrinth, more often causing sensorineural hearing loss and facial nerve paralysis (Schuknecht, 1974).

Concussion

Head trauma may be associated with brief unconsciousness without appreciable neurologic deficits or radiographic evidence of structural brain damage. The symptoms of post-concussion syndrome include headache, irritability, dizziness, difficulty in concentrating, intolerance to loud noise, and insomnia. Tinnitus with or without detectable hearing loss is another common complaint.

The cause of post-concussion syndrome is not known. Possible causes include derangement of vestibular apparatus, endolymphatic hydrops, and minimal brain stem injury. Fortunately, most patients recover without too many sequelae.

Cervical Vertigo (Whiplash Injury)

Neck injury may be associated with a benign, paroxysmal type of vertigo. About one-half of patients demonstrate objective evidence of either caloric abnormality or nystagmus (Toglia et al, 1970). Whiplash injury is also associated with hearing loss and tinnitus.

The cause of the vertigo following neck injury is not clear. Possible mechanisms of cervical vertigo include alteration of the tonic neck reflex, irritation of cervical nerve roots, involvement of the sympathetic nervous system, and compression of the cervical artery (Ryan and Cope, 1955; Sheehan et al, 1960). However, some doubt the existence of cervical nystagmus (Barnes and Forbat, 1979).

Metabolic and Toxic Disorders

Auditory and vestibular symptoms are common with metabolic or hormonal imbalance, alcohol or other drug intoxication, and malnutrition. The pathophysiology of auditory and vestibular symptoms caused by these disorders is poorly understood. The central nervous system and cranial nerve VIII as well as end-organs are probably affected.

Diabetes Mellitus

Sensorineural hearing losses have been reported in approximately 50 per cent of diabetic patients (Jorgensen and Bach, 1961). Diffuse vascular pathology involving the inner ear, cranial nerve VIII, and the central nervous system is the probable mechanism of auditory and vestibular problems in diabetes mellitus. For example, a high incidence of PAS-positive thickening of the capillary walls, especially in the stria vascularis, was noted in the temporal bones of diabetics (Jorgensen, 1961). Similar vascular pathology was found in Alloxan-induced diabetes in rabbits involving the stria vascularis, Scarpa's ganglia, and vestibular nerve fibers (Cojazzi and Boetner, 1950).

Hypoglycemia may cause dizziness and vertigo. Hearing loss was correlated with hypoglycemia in patients who complained of fluctuation in their hearing before meals (Parkin and Tice, 1970).

Thyroid Dysfunction

The relationship between hypothyroidism and hearing loss is well known in the pediatric genetic disorder of Pendred's syndrome. In this syndrome, thyroid enlargement, which is present at birth or occurs at puberty, is associated with severe sensorineural hearing loss.

The relationship between hearing loss and thyroid deficiencies in adults is not so clear. There have been several reports documenting hearing improvement in patients with hypothyroidism after treatment with thyroid extract (Cody, 1971; Vant Hoff and Stuart, 1979). In other reports, no improvement in hearing threshold was demonstrated after thyroid replacement therapy in elderly patients with myxedema (Parving et al, 1983). Well-controlled studies are needed before a firm conclusion can be made.

Malnutrition

Nutritional deficiencies, especially certain vitamin deficiencies, are associated with neuropathies including auditory and vestibular symptoms. Thiamin (vitamin B_1) deficiency, which commonly occurs as a result of alcoholism, can cause vomiting, horizontal nystagmus, ophthalmoplegia caused by weakness of the rectus muscles, ataxia, and gradual onset of sensorineural hearing loss (Denny-Brown, 1974). Deafness was reported in patients with nicotinic acid deficiency (Merritt, 1959). A temporal bone from a patient with malabsorption syndrome and bilateral progressive severe sensorineural hearing loss showed marked loss of cellular elements of the spiral ligament, degeneration of the organ of Corti, necrosis of the stria vascularis, degeneration of the saccular macula with otolithic membrane displacement, and bilateral segmental demyelination of vestibular and cochlear nerves (Gussen, 1974). Hypervitaminosis A and D can cause vertigo and headache (Swaiman and Wright, 1983).

Drugs

The number of drugs that cause vertigo, tinnitus, and deafness continues to grow. The auditory and vestibular symptoms are side effects of these drugs and may occur from a toxic effect on the end-organ or on the central pathway.

Aminoglycoside antibiotics have auditory and vestibular toxicity that is primarily due to damage of the end-organ. In a randomized, blind assessment of the ototoxicity of gentamicin, tobramycin, and netilmicin, cochlear damage was found to be highest with gentamicin (16.4 per cent), followed by tobramycin (9.6 per cent) and netilmicin (2 per cent). In the same study, vestibular damage was highest with gentamicin (11.8 per cent), followed by tobramycin (3.3 per cent), and the least with netilmicin (0 per cent) (Lerner et al, 1984). Other ototoxic drugs with primary effects on end-organs include diuretics such as furosemide and ethacrynic acid, cisplatin, and erythromycin (Rybak, 1982; Moroso and Blair, 1983; Schweitzer and Olson, 1984).

Alcohol produces vestibular symptoms in both acute and chronic intoxication. In acute intoxication, the effects probably originate at the periphery, producing positional nystagmus (Aschan et al, 1956). With chronic intoxication the pathology is found at central sites, and major lesions occur in the periventricular regions of the diencephalon, midbrain, and brain stem and in the superior vermis of the cerebellum. Clinical manifestations of Wernicke's encephalopathy include a clinical triad of ophthalmoplegia, ataxia, and global confusion as well as diplopia, disturbed balance, horizontal nystagmus on lateral gaze, and vertical nystagmus on upward gaze in about 50 per cent of cases (Charness and Diamond, 1984). Other drugs that affect central auditory and vestibular system include sedatives, tranquilizers, and hypnotics (Spector and Wilf, 1967).

Demyelinating and Degenerative Diseases

Auditory and vestibular anomalies often are early symptoms of demyelinating and degenerative diseases. Since pathologic lesions are usually widespread throughout the central nervous system, symptoms are also multiple and diverse.

Multiple Sclerosis

Multiple sclerosis is a demyelinating disease of unknown etiology characterized by lesions disseminated in time and space within the central nervous system. It is a disease of multiple neurologic signs and symptoms, characterized by remissions and exacerbations.

Clinical manifestations include blurring of vision, diplopia, limb weakness, ataxia of gait, intention tremor, incoordination of limbs, paresthesias, and sphincter impairment. Vertigo is the initial symptom in about 5 per cent of patients and occurs sometime during the course of disease in as many as 75 per cent (Baloh, 1983; Toglia, 1967). Nystagmus in multiple sclerosis is often bounding or pendular (Aschan et al, 1956). The abnormalities most often encountered in electronystagmograms of patients with multiple sclerosis were smooth pursuit (96 per cent),

saccadic eye movement (76 per cent), optokinetic nystagmus (53 per cent), abnormal caloric reactions (40 per cent), and defective visual suppression of the nystagmus (43 per cent) (Grenman, 1985). Hearing loss occurs in about 10 per cent of patients (Noffsinger et al, 1972). Hearing loss may be a part of the initial symptoms or may occur later in the course of disease. It can be sudden or gradual in onset. Pure-tone audiometry has no characteristic pattern for lesions of multiple sclerosis, but a battery of special audiometric studies such as acoustic reflex, tone decay, and speech audiometry is helpful in localizing the lesions. ABR audiometry is abnormal in 32 to 93 per cent of patients with multiple sclerosis and can detect subclinical lesions of multiple sclerosis (Stockard et al, 1977; Hosford-Dunn, 1985). The ABR in patients with multiple sclerosis is characterized by prolonged latency and diminished amplitude of wave V, increased I-V interval, and abnormal waveforms. ABR audiometry in the early diagnosis of multiple sclerosis is valuable because, first, it identifies silent lesions and, second, it can differentiate multiple sclerosis from other diseases (Chiappa et al, 1980). There is no specific laboratory test for multiple sclerosis. Cerebrospinal fluid examination shows a high percentage of elevated gamma globulin and myelin basic protein (Waxman, 1983). MRI shows characteristic lesions at the paraventricular area and brain stem and is promising as a definitive diagnostic test.

There is no definitive treatment for multiple sclerosis. Corticosteroids and adrenocorticotropic hormone (ACTH) may be used during an acute exacerbation.

Spinocerebellar Degenerations

Spinocerebellar degenerations are groups of inherited progressive degenerative disorders characterized by ataxia and dysmetria resulting from degeneration of the cerebellum. Auditory and vestibular symptoms are common in these disorders, which are associated with hearing loss and absent vestibular function. Ataxia, not vertigo, is the predominant symptom, because of the bilateral progressive loss of vestibular function. Many are characterized by pathologic nystagmus such as gaze-paretic nystagmus, spontaneous vestibular nystagmus, positional nystagmus, and rebound nystagmus (Baloh and Honrubia, 1979).

Friedreich's Ataxia. This is the commonest form of spinocerebellar degeneration. It begins in childhood and is characterized by dysarthria, nystagmus, moderate mental retardation, arched feet, scoliosis, and cardiomegaly with fibrosis. Inborn errors of metabolism with deficiencies of pyruvate oxidation may be the cause of this disorder (Blass et al, 1976).

Refsum's Disease. This disorder is characterized by pigmentary retinal degeneration, ichthyosis, bilateral sensorineural hearing loss, cerebellar ataxia, and peripheral neuropathy. The cause of Refsum's disease is the deficiency of an enzyme necessary for the oxidation of phytanic acid.

Roussy-Lévy Disease. Roussy-Lévy disease begins in childhood with ataxia, areflexia, clubfoot deformity, and kyphoscoliosis. It differs from Friedreich's ataxia in sparing position and vibratory sensation and in the absence of nystagmus, dysarthria, and extensor plantar responses (Brown, 1983).

Olivopontocerebellar Degeneration. This disorder is characterized by atrophy of the cerebellum, brain stem, and spinal cord. Essential clinical features include progressive ataxia, dysarthria, dysmetria, nystagmus, dysdiadochokinesia, hyperreflexia, extensor plantar responses, and optic nerve atrophy. It begins in mid-adulthood and may be inherited with either an autosomal recessive or an autosomal dominant pattern. CT scans show pontine and cerebellar atrophy and ABR audiometry shows a delay between waves II and III and reduce voltage levels in all components (Gilroy and Lynn, 1978).

Other Retrocochlear Disorders

Epilepsy

Vertigo may be a part of an aura of focal seizure, especially when it develops from the temporal lobe. Epilepsy is a possibility whenever there is a transient attack of vertigo and loss of consciousness. In a study of 120 patients with focal seizures who experienced vestibular symptoms as part of their aura, the most common vestibular symptom was a sense of spinning (55 per cent), followed by a sense of linear movement (30 per cent) (Smith, 1960). Other associated symptoms included visceral and autonomic symptoms (62 per cent), visual symptoms (45 per cent), auditory symptoms (28 per cent), and somatosensory symptoms (22 per cent).

Initial evaluation and treatment of these patients are directed at finding the underlying pathology, such as brain tumor or subdural hematoma. Medical therapy can be started after the underlying pathology is fully evaluated.

Autoimmune Disease

Cogan's syndrome with interstitial keratitis, episodic vertigo, tinnitus, and deafness is an example of an autoimmune disease with otologic manifestations (Cogan, 1945). In other collagen vascular diseases, vestibular symptoms have been reported in 50 per cent of patients with relapsing polychondritis (Damiani and Levine, 1979) and in 13 per cent of patients with systemic lupus erythematosus (Bowman et al, 1986). McCabe (1979) reported that patients with autoimmune-induced sensorineural hearing loss responded to therapy with corticosteroid and cyclophosphamide. An animal model of autoimmune sensorineural hearing loss and vestibular dysfunction using type II collagen has been developed (Yoo et al, 1983). In these animals, changes in auditory and vestibular nerves and ganglia were noted. Treatment for the patient with autoimmune medicated disease includes corticosteroids, cytotoxic drugs, and plasmapheresis.