

Paparella: Volume III: Head and Neck

Section 2: Disorders of the Head and Neck

Part 7: The Neck

Chapter 48: Malignant Melanoma: Cutaneous and Mucosal

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Malignant melanoma is of increasing importance as the incidence of the disease rises throughout the world. Cutaneous melanoma is, in the majority of cases, visible on the skin and so should be easily diagnosed if patient and physician are aware of its characteristics. Mucosal melanoma may be less easily identified, depending on its location and its clinical presentation, and may be diagnosed only when histologic reports are received. It is clear that early diagnosis and treatment will produce the best results, and these goals will be obtained by increasing the awareness of both doctors and the public concerning the signs that may suggest a lesion is a malignant melanoma. In this chapter, we propose to deal with cutaneous and mucosal melanomas of the head and neck separately, as they do have pronounced differences in their behavior and responses. Ocular melanoma will not be discussed, as it falls more into the field of the ophthalmologist, apart from the treatment of advanced disease.

Cutaneous Melanoma of the Head and Neck

Incidence

The incidence of malignant melanoma varies according to the country of origin and its latitude, the racial origin of the population, and the degree of their skin pigmentation. The persons most prone to the development of malignant melanoma are those of Celtic origin, a group whose original distribution was in northern and central Europe, Scandinavia, and the British Isles. They tend to have fair or reddish complexion, may freckle easily, and are subject to sunburn when exposed to ultraviolet radiation for only short periods. By comparison, those races with deeper skin pigmentation and with less sensitivity to sunlight exposure have a much lower incidence of cutaneous melanoma, except on plantar skin and in subungual areas.

The highest incidence of malignant melanoma is in Queensland, Australia, where the Celtic makeup of the population is high and the climate and life style lead to high levels of exposure to sunlight. The incidence of cutaneous melanoma in the other Australian states falls as the distance from the equator increases and the intensity of ultraviolet radiation consequently decreases. The incidence of invasive malignant melanoma in Queensland has risen from 15.1 cases/100,000 population in 1966 to 28.4 cases/100,000 population in 1979 to 1980. Figures from the Queensland Cancer Registry suggest that the incidence is continuing to rise.

Cutaneous melanoma of the head and neck composed 16 per cent of the 624 invasive melanomas recorded in Queensland in 1979 to 1980. Thirty-two per cent of these lesions were lentigo maligna melanoma, 37 per cent were superficial spreading melanoma, 21 per cent were nodular melanoma, and the remainder were not classified. Of the 247 Clark's level I (in

situ) melanomas removed in the same period, 48 per cent were of the lentigo maligna type.

The incidence in other countries is lower; however, as they do not all have compulsory cancer registries, complete figures are not available. In Denmark, the incidence was shown to have risen by 25 per cent in two periods from 1968 to 1972 and from 1973 to 1977. In Sweden, the rate rose by more than 400 per cent between 1958 and 1980. The incidence has also risen markedly in England and Scotland. The reason for this world-wide rise in the incidence of cutaneous melanoma is conjectural, but it is a rise that is seen in cohorts of the same age. This suggests that life style may play a part, and the increased affluence of many countries and the tendency to vacation in sunny countries may indicate that increased exposure to ultraviolet radiation from sunlight may play a part.

History

A history of change in a pre-existing mole, or the development and continued growth of a new pigmented lesion, should lead to suspicion of melanoma. The changes are usually observed over a period of weeks or months and may include an irregular increase in area or elevation, or both; a change in color, most often to a darker shade of brown or to black; the development of a fine scaling of the epidermal surface over the lesion; an itch or irritation in or around the lesion; and in more advanced ulcerated lesions, serous oozing or bleeding.

Clinical Diagnosis

The aim of the clinician should be to diagnose malignant melanoma at a biologically early stage, that is, when it is less than 0.76 mm thick and therefore is likely to be cured by adequate surgery. Melanoma is a potentially fatal disease if the diagnosis and definitive treatment are delayed until the later stages of its biologic development. The experience of the members of the Queensland Melanoma Project is that any competent and aware practitioner can be taught to recognize a melanoma from the history of its development and the macroscopic appearance of the lesion. Although not every melanoma will be diagnosed in this way, the majority of these lesions will show characteristic features. Of course, confirmation of the diagnosis remains with the pathologist.

Differential Diagnosis

The clinician must use the history and the clinical findings to attempt to make a differential diagnosis of the lesion. Benign melanocytic tumors, including lentigo, junctional, compound, and intradermal nevi; blue nevus; halo nevus; and Spitz nevus must all be considered as they will frequently have features that may suggest melanoma. Other lesions such as seborrheic keratosis, pyogenic granuloma, hemangioma, and sclerosing angioma (dermatofibroma) must be included. Malignant skin lesions that at times may be difficult to differentiate from melanoma include pigmented basal cell carcinoma, squamous cell carcinoma, and Kaposi's sarcoma.

Dysplastic Nevus Syndrome

The dysplastic nevus syndrome was first described by Clark and associates in 1978, and the understanding of this important predisposition to the development of melanoma is

now becoming widespread. Patients who are identified as having dysplastic nevus syndrome should be classified as being at high risk for melanoma, and any doubtful lesions should be excised for histologic examination. Dysplastic nevi are more commonly seen on sun-exposed skin and are more frequent on the trunk and limbs, but they are also on the neck, face, and scalp. Careful and regular examination of persons in this high-risk group will lead to early identification of changing lesions and thus to improved prognosis.

Clinical Features

The clinician can recognize features that separate melanomas into groups, and these groups are identified by the pathologist from their histologic patterns. There are three major groups in the clinicopathologic classification of melanoma, with two smaller groups that cover only a very few lesions. There are also some special variants that are of significance.

Superficial Spreading Melanoma

Superficial spreading melanoma lesions are usually greater than 5 mm in diameter and may be considerably larger. They may have a variety of coloration throughout their substance and may be irregular in shape. The use of a hand lens with x 10 magnification enables a much clearer examination of the features of a melanoma, and in a superficial spreading melanoma, the fine pattern of skin lines may appear coarsened and partly lost. Scaling of the epidermis may be visible and nodules, either pigmented or amelanotic, may develop within the lesion.

Lentigo Maligna Melanoma

Lentigo maligna melanoma is common on the sun-exposed, and consequently ultraviolet radiation-damaged, skin of the head and neck. It arises in a pre-existing lentigo maligna (Hutchinson's melanotic freckle), which is a premalignant lesion that may exist for many years before undergoing malignant change. The freckle may be irregular in shape, advancing at some edges and regressing at others, varying in color in different parts, with brown, black, and reddish areas. When malignant change occurs, the surface skin pattern is disturbed, nodules or plaques of elevation arise, and progression is then as for other melanomas.

Nodular Melanoma

Clinically, nodular melanoma is a raised lesion with no peripheral area of flat in situ change. It may be small in diameter but is always at least Clark's level II in invasion and usually is at deeper levels when diagnosed. The lesion is palpable, has a defined edge, is usually black, and may be scaly or crusted on the surface.

Desmoplastic Melanoma

Desmoplastic melanoma (and its neurotropic variant) is a special type of melanoma that is frequently situated on the head and neck. It is not a common variant but is of importance because of difficulty in diagnosis, both for the clinician and for the pathologist. These lesions are often lightly pigmented or nonpigmented and may have none of the normal

features of melanoma on inspection. The neurotropic type is frequently found occurring as a nodule on the lip. These lesions may be diagnosed only when the pathologist examines a lesion excised for diagnostic purposes.

Pathologic Features

Clark and co-workers described the concepts of radial and vertical growth phases, which can be identified in all melanomas other than those described as nodular melanoma, which is diagnosed by the lack of the radial growth phase. The radial growth phase is the proliferation of atypical melanocytes and upward epidermal invasion, which if present without dermal invasion would constitute a Clark's level I (in situ) melanoma. When there is an associated vertical growth phase, the lesion is an invasive melanoma. Clark's levels of invasion give an indication of the penetration of the melanoma into the skin and are useful in determining prognosis, though they are not as favored for this purpose as is the measured thickness by the Breslow method. A schematic presentation of the Clark levels is given. A full discussion of the pathologic features of malignant melanoma should be sought in a specialized text.

Superficial Spreading Melanoma

The most common form of melanoma in most series is the superficial spreading type. The radial growth phase has the characteristics of in situ melanoma, and the vertical growth phase shows melanoma cells infiltrating into the dermis. There are differences between the cellular appearances of the radial growth phase of superficial spreading melanoma and those of lentigo maligna melanoma.

Nodular Melanoma

Nodular melanoma is seen less commonly than is superficial spreading melanoma, but it is considered to be more malignant. There is no radial growth phase to be identified, possibly because it is small in area and enveloped in the early developing invasive vertical growth. These lesions are often diagnosed when invasion has progressed deeply, and thus the prognosis is not as good as in the thinner, more superficially invasive lesions.

Lentigo Maligna Melanoma

Melanoma arising in a Hutchinson's melanotic freckle is classified as lentigo maligna melanoma.

Acral Lentiginous Melanoma

A less frequently occurring type of lesion is acral lentiginous melanoma, which is found peripherally on palmar and plantar skin and in the subungual area. The radial growth phase shows features resembling the radial growth phases of both superficial spreading and lentigo maligna melanoma. Diagnosis of these lesions is often difficult.

Melanoma of Special Types

Malignant melanoma may arise in blue nevi and giant congenital nevi. In these lesions, the change occurs in the deeper part of the lesion, and diagnosis may be delayed until a late stage. Biopsy is necessary if a nodule arises in a giant congenital nevus. A history of change in a blue nevus suggests the need for excision for diagnostic purposes.

Special Variants

A special variant of melanoma is the desmoplastic melanoma, which is a spindle cell melanoma producing a stromal fibrous response. They are seen predominantly on the head and neck, and a group of them showing a neurotropic variation are frequently found on the lip. These variants are often amelanotic and may be misdiagnosed both clinically and histologically. They are associated with a poor prognosis when the neurotropic variant invades along nerve fibers, and they may become intracranial by direct infiltration.

Treatment

Surgery has been the treatment of choice for malignant melanoma, and present results show that about 80 per cent of patients treated by surgical methods alone survive without recurrence of disease. The width of the margin of normal skin from the edge of the lesion has been under consideration, and the very wide margins previously recommended are now believed to be neither necessary for thin lesions or beneficial for patients with thick lesions. Because of the anatomic constraints, wide excisions have not often been practicable on most areas of the head and neck. Our general policy is to seek a margin 1 cm wide from the edge of the melanoma, planning the incision to gain a good cosmetic result when possible by primary suture, but if necessary, by rotation flap or skin graft. The lesion should be excised down to, but not including, the fascia over muscle when possible, but the various sites on the head and neck may require modification of this general approach.

Ionizing radiation has been used in the treatment of primary malignant melanoma, especially for lentigo maligna melanoma, and reports of good results have appeared. It is not our policy to use radiotherapy for these lesions, and we do not use radiotherapy for the premalignant lentigo maligna (Hutchinson's melanotic freckle). Surgery can be adequately performed with good cosmetic results in nearly all patients, and those too frail to undergo surgery with modern local, neuroleptic, or general anesthetic techniques may well be better managed by expectant observation rather than by operative intervention.

The techniques of cryotherapy, topical chemotherapy, curettage, and cautery have all been used with some success, but also with some local and distant recurrences, and our policy is to seek surgical excision and full histologic examination of all suspicious lesions for diagnostic and prognostic purposes.

Lentigo Maligna Melanoma

A common form of melanoma on the head and neck is lentigo maligna melanoma. It arises in a pre-existing lentigo maligna (Hutchinson's freckle), and many such lesions can be prevented by the prophylactic excision of the premalignant Hutchinson's melanotic freckle

before it undergoes malignant change. When invasive melanoma occurs in a Hutchinson's melanotic freckle, the major prognostic features in the histologic examination are the measured thickness, the presence or lack of ulceration, and the mitotic rate. The fact that the lesion arose in a Hutchinson's melanotic freckle does not confer a survival benefit over patients with lesions of the other types if these prognostic factors are similar.

Superficial Spreading Melanoma

Surgical excision is the preferred treatment for superficial spreading melanoma, and only in exceptional circumstances, such as severe medical illness, would other techniques be contemplated. As with melanoma on other parts of the body, there is doubt whether radical excision margins (2 cm plus) are necessary, and the limitations of area and anatomy on the head and neck have meant that 1-cm excisions have been practiced. There is no evidence that this leads to poorer results, and a 1-cm margin, when feasible, is adequate.

Nodular Melanoma

Often a nodular melanoma of the head and neck may be diagnosed when it has become deeply invasive, because of obscured visibility (in the scalp covered by hair), rapid growth, or small diameter. Again, surgical excision with an adequate margin of 1 cm when possible is the preferred treatment.

Regional Limitations

Face and Neck. Good cosmetic results can be achieved on the face and neck with adequate excision margins in most cases. Problems may arise when the lesion is near an eye, on the eyelid, or close to an ear or lip. Lentigo maligna melanoma is more common in older patients who frequently have lax facial skin with loss of elasticity, and closure is relatively easily achieved even with quite large lesions. Superficial spreading melanoma and nodular melanoma are usually smaller in area than is lentigo maligna melanoma, and these lesions present fewer problems with adequate excision and primary closure.

Scalp. Melanoma on the scalp may be diagnosed at a biologically advanced stage and is thought to have an overall poor prognosis. If the primary lesion is not accompanied by satellites, and the regional nodes are clear, excision with a 1- or perhaps 2-cm margin and a partial-thickness skin graft will usually give a good local result. If there are satellites, a wider excision will be needed to eliminate the local disease, and consideration of elective lymph node dissection will be indicated.

Ear. Melanoma on the ear needs to have the excision planned to remove all primary disease, and it usually requires partial excision of the ear rather than removal of skin alone. On the helix, a wedge excision will allow a reasonable cosmetic reconstruction, whereas the ear lobe can be removed if the melanoma is situated in that area. Fortunately, it should rarely be necessary to remove the entire ear, as the resulting deformity and inconvenience, especially to wearers of eyeglasses, is severe.

External Auditory Canal. Melanoma arising in the external auditory canal may develop for some time before it produces symptoms, so it may be diagnosed only when deep

invasion has occurred. Surgical excision seeks to eradicate the primary lesion and any local extension. Recurrent disease at this site is very difficult to manage, and a multidisciplinary approach may be required.

Lip. Surgical texts have often referred to the need to excise pigmented lesions of the lip because of a suggested high risk of change to malignant melanoma. When these lesions are removed, the report is most often of freckle or lentigo. Occasionally, a compound nevus is reported, but junctional nevus is uncommon. This may be because the lentiginous lesions are removed before they progress to become lentiginous junctional nevi. Pigmented malignant melanoma on the lip is uncommon, though it sometimes is seen as an extension of melanoma arising in a nearby Hutchinson's melanotic freckle on the chin. Nonpigmented nodules occurring in the skin or in the substance of the lip can prove to be spindle cell lesions with neurotropic features, and the pathologist may have difficulty in differentiating between neurofibrosarcoma and melanoma. Special stains may be of assistance in the differential diagnosis. When neurotropism is reported in lip lesions, or in lesions at any other site, the excision margins should be wide. We have seen several patients in whom the tumor has spread along the branches of the mandibular nerve and reached the base of the intracranial fossa, becoming untreatable. The tendency of the neurotropic variety of desmoplastic melanoma to extend proximally along nerves makes these lesions particularly difficult to eradicate. The advent of magnetic resonance imaging has aided the assessment of these patients, as infiltration of nerves and the extent of disease can be clearly shown.

Management of the Regional Nodes

Elective lymph node dissection is no longer practiced as a routine after excision of the primary lesion but is limited to carefully selected patients in whom the risk of regional lymph node metastases is high. These patients are those in whom the primary lesion was thick (1.5 mm or more), the nodal drainage is predictable, the probability of distant metastases is not high, and when, after full explanation of the reasons for and against the procedure, the patient is prepared to accept the recommendation. The aim of elective dissection is to remove micrometastases in the nodes, and the procedure can be justified only if more than a small proportion of the patients so treated prove to have nodal metastases on histologic examination, or if an appreciably increased survival rate can be shown for those patients undergoing operation.

Therapeutic lymph node dissection is recommended when clinical suspicion of lymph node involvement is formed. Even in the presence of distant metastases, operation may still be indicated as a palliative procedure to prevent the local problems of pressure, ulceration, and infiltration by large malignant masses.

The techniques of dissection of the cervical lymph node fields are well described in available texts and will not be further detailed here.

Adjuvant Therapy

Attempts to improve the prognosis for patients with high-risk melanoma have included use of adjuvant agents to suppress or eliminate occult metastatic disease. Immune stimulation by agents such as smallpox and bacille Calmette-Guérin (BCG) vaccines and by

Corynebacterium parvum have not been shown to improve the prognosis. Using chemotherapeutic substances as adjuvants produces problems of toxicity, and no obvious benefits has been demonstrated. Adjuvant studies using biologic-response modifiers are proposed, but the toxic effects of these agents are not negligible, and it will be some years before the results of trials can be assessed. At present, there are no known adjuvants that can be used to enhance the prognosis in high-risk melanoma patients.

Management of Distant Metastases

Malignant melanoma that has metastasized beyond the regional nodes may appear in any tissue or organ of the body. It is commonly seen in lungs, liver, heart, spleen, kidney, adrenal gland, gut, and brain, as well as in the subcutaneous tissues and bone.

No one treatment has been shown to be useful in all patients with advanced disease, and consideration of each individual case is necessary before formulating a plan of management. Modalities to be considered are surgery, radiotherapy, chemotherapy, and biologic-response modifiers. In addition, local ablative procedures may be of use for cutaneous, subcutaneous, and mucosal deposits. Immunotherapy has been extensively investigated, but there is no evidence that it offers benefit, and it will not be discussed further in this chapter.

Surgery may be used to excise recurrent disease, both when there is hope of thereby eliminating the disease and when excision of a lesion will offer palliation by improving the quality of life and when the quantity of life can be increased without undue impairment of the quality of life. Each patient needs careful individual assessment to determine the nature and extent of surgery indicated.

Radiotherapy is useful for metastatic disease occurring in bone, for inoperable lymph node metastases, and in some patients in whom surgical excision of a metastasis is contraindicated because of a medical condition. Special sites on the head and neck include retro-orbital metastases and occasionally nasopharyngeal melanoma that is unsuitable for surgery.

Chemotherapy with single agents has been used to attempt control of metastatic melanoma, but success rates are usually no higher than 15 per cent. It has been claimed that multiple-agent chemotherapy offers better results, but the toxic effects are such that quality of life may be greatly reduced, and the gain in quantity of life often does not justify these measures.

Biologic-response modifiers are still in the early stages of use in melanoma, and investigative trials are proceeding. Recombinant interferons have been shown to produce objective responses in about 16 per cent of patients, and when used in combination with dacarbazine (DTIC), the response rate has been 30 per cent. Similar results have been reported with recombinant interferon and other chemotherapeutic agents. Interleukin 2 has also produced promising results, and further investigation of both agents in various combinations is being pursued.

Diathermy coagulation of cutaneous and mucosal metastases is often effective in palliation, and cryotherapy of cutaneous and mucosal metastases has also been used. Laser treatment to reduce or eliminate masses may offer benefit.

Cutaneous malignant melanoma of the head and neck is best managed by prevention when possible, early diagnosis, and correct definitive treatment. Recurrent disease requires appropriate treatment regimens, but once the disease has metastasized, the prognosis is poor.

Mucosal Melanoma of the Head and Neck

Incidence

Malignant melanomas arising from mucous membranes represent 0.5 to 2 per cent of all malignant melanomas. The incidence as a percentage of head and neck tumors in general, however, is probably of more clinical relevance. Of 1264 new cases of various head and neck tumors (benign and malignant) presenting at the Head and Neck Clinic at the Princess Alexandra Hospital, Brisbane over an 8-year period, 6 cases were mucosal malignant melanomas (Table 1). This is an incidence of 0.5 per cent of tumors in general involving the head and neck region. Further breakdown to specific sites is similar to the Toronto experience, with melanoma representing less than 0.5 per cent of oral cavity and oropharyngeal malignancies but a very significant 20 per cent of malignant nasal cavity lesions.

Table 1. Incidence of Mucosal Malignant Melanoma Related to Total Number of Head and Neck Tumors (Benign and Malignant)

Primary Site	Number	Malignant Melanoma
Oral cavity	244	nil
Oropharynx	123	1
Nasopharynx	41	1
Nasal cavity	20	4
Nasal sinuses	46	nil
Other	790	nil
Hypopharynx		
Esophagus		
Skin		
Salivary glands		
Benign lesions		
Total	1264	6 (0.5 per cent).

There is an apparent male predominance, with occurrence being most frequent between 40 and 70 years of age. It is generally inferred from the age occurrence that mucosal malignant melanoma appears at a more advanced age than does malignant melanoma of the skin. The majority arise in the nasal and oral cavities. Rarely are they found more caudally in the pharyngolaryngeal region or upper esophagus. The etiology remains unknown.

As mentioned earlier with cutaneous melanomas, there seem to be some racial differences. In Japanese persons, for example, melanoma of the skin is exceedingly rare when compared with white persons, but mucosal melanoma is relatively more common and is frequently associated with pre-existing pigmentation or melanosis. Benign racial pigmentation also occurs with bluish black lesions of the oral mucosa in black persons, being so common as to be considered physiologic. They are symmetric and are generally limited to the buccal and labial surfaces of the gingiva. Focal pigmentation from hemoglobin breakdown products are a dental amalgam tattoo may present a different diagnostic problem.

Clinical Features

The clinical features and natural history of mucosal melanomas clearly depend on the site of origin.

Melanoma involving the nasal cavity presents with the same symptoms as other nasal tumors, that is, airway obstruction, discharge, crusting, and bleeding. Sites of predilection are said to be the septum and the inferior and middle turbinates; however, the lesion is often bulky and polypoid, and it is impossible to determine the exact site of origin. Pain is uncommon unless the tumor mass has caused secondary obstructive inflammatory changes in the sinuses. The duration of signs and symptoms before medical attention is sought is variable, in part because of the similarity of symptoms to those of chronic rhinitis and allergic nasal polyposis. Anterior rhinoscopy reveals a polypoid mass with possible areas of necrosis that bleed easily. Of particular note is that pigmentation is often lacking or not readily visible.

Melanomas involving the oral cavity, however, are generally pigmented with a predilection for the palate and alveolar ridge. This is in sharp contrast to oral squamous carcinoma, which more often involves the lower jaw and floor of the mouth. Bleeding and ulceration are the common symptoms, but because the latter lacks induration, pain is not a common feature. Hence, there is a similar delay in the patient's seeking medical attention.

Regional lymph node involvement occurs with melanomas of the tonsil, being most aggressive with early metastatic, or even direct, spread to the jugulodigastric region. In a review of the literature, Snow and Van der Wall state that primary melanoma involving the nasal mucosa, however, rarely presents on admission with regional lymph node involvement. Spread to the retropharyngeal and upper deep cervical nodes may eventually occur. Distant metastatic spread to lung, liver, and brain is the major cause of death.

Mucosal malignant melanoma is characteristically unpredictable in its clinical course. In some cases, the tumor progresses rapidly, with early widespread dissemination, or it can remain relatively dormant for periods of 10 years or more with low-grade recurrences that respond well to simple debulking.

Diagnosis

As a concept, biopsy is always indicated for any lesion that does not have an indisputably innocuous cause. Clinical symptoms of mucosal melanoma really are of little assistance in a prebiopsy situation. Prompt and early diagnosis, therefore, is based on (1) a high index of suspicion and (2) appropriate submission of excised tissue for histologic

assessment. (The emphasis here is with respect to nasal polyps.)

Generally, it is not feasible to apply the concept of excisional biopsy, and any biopsy material is incisional by virtue of the sites involved. Hence, accurate assessment of the thickness and penetration of the primary lesion is, at the very best, difficult and is virtually impossible with polypoid nasal melanomas. Assessment of the thickness remains important, however, as far as microstaging and prognosis is concerned, and such assessment should always be attempted. This requires the surgeon to submit tissue of sufficient quality and orientation. Clark's levels of invasion have no application with mucosal lesions because of the lack of layers analogous to the papillary and reticular dermis.

It is important to remember that skin melanomas can metastasize to all organs of the body, including the mucous membranes of the head and neck, although this is uncommon. Before the diagnosis of primary mucosal malignant melanoma can be made, three criteria need to be satisfied: (1) the demonstration of clinical and microscopic tumor, (2) the presence of intraepithelial activity, and (3) the inability to demonstrate any other primary lesion.

Difficulties associated with the microscopic evaluation in some circumstances, plus the role of S100 immunohistochemical methods, are detailed in standard pathology texts. Again, it is worth emphasizing that approximately one third of mucosal melanomas are amelanotic.

Treatment

Treatment has been determined over the years by the experience with cutaneous melanomas, with surgery being the preferred option.

Wide en bloc excision is recommended for the primary lesion, with the detailed nature of the surgery being determined by the individual primary site and extent of spread at presentation. Nasal melanomas will require, as a minimal procedure, removal via a lateral rhinotomy approach incorporating resection of the lateral bony wall of the nose (i.e. medial wall of the maxillary and ethmoidal sinuses). When the middle turbinate or upper septum is involved, it must be assumed that the entire ethmoidal complex is involved. Adequate resection in this situation can be undertaken only by a combined craniofacial approach. Even with lesions that are primarily in the oral cavity, some form of nasal or partial maxillary removal will be required because of the predilection for occurrence on the palate and alveolar ridge. Therapeutic dissection is naturally in order when there is clinical suspicion of lymph node metastases. Elective-prophylactic dissection, however, is not appropriate because (1) the previously mentioned primary sites are essentially midline structures with bilateral lymph drainage patterns and the nodal area removed with a standard radical neck dissection is discontinuous with the primary nasal sites. (The lymphatic drainage is usually to the retropharyngeal nodes.)

For primary lesions of the lower oral cavity and oropharynx, surgical clearance will most likely involve a jaw-neck resection of the Commando type. In this situation, elective-prophylactic dissection would seem appropriate because of (1) the high incidence of occult nodal disease, (2) the continuity between the primary sites and lymphatic drainage, and (3) the fact that the neck will be entered for both resection and reconstructive reasons.

The mandatory 5-cm margin advocated years ago is no longer recommended, applicable, or even necessary in view of advances in the microstaging, and this concept is particularly important in the head and neck region in order to better preserve function and appearance. By the same token, more aggressive surgical resection can now be undertaken because of improved anesthetic and intensive care facilities, as well as reconstructive options.

Because of the characteristic, unpredictable course of malignant melanoma, recurrent disease is always worth treating. Local and regional recurrences are best treated, if possible, by further definitive radical surgical resection. For recurrent nasal melanoma, long-term and quite reasonable palliation cannot infrequently be obtained by simple debulking or cryotherapy. For many years, mucosal melanomas have been regarded as radioresistant, but favorable responses have been reported, and it probably has a role to play in the salvage situation. The role of chemotherapy and immunotherapy with interferon was discussed earlier in this chapter and applies equally to mucosal disease and its disseminated metastases.

The ultimate prognosis for mucosal melanomas is extremely poor, although prolonged periods of palliation can be obtained.