Surgical Management of Conjunctival Melanomas

It has been proposed that ocular surface melanomas should be treated by simple local excision and cryotherapy to the conjunctival borders. Based on what is known of the biology of both cutaneous and mucosal melanomas, however, we consider it unlikely that such a technique, without deep scleral clearance, will reliably eliminate ocular surface melanomas. Indeed, the documented behavior of melanomas on other areas of the body is contradictory to this possibility. As with other melanomas, avoiding local recurrence of conjunctival melanomas by excising the tumor with adequately wide margins is clearly of critical importance because it has previously been shown by the same authors that local recurrence of conjunctival melanoma is associated with a markedly increased rate of distant metastasis.

We are very concerned that the results of the conservative excision techniques described by Shields et al have not been reported and believe that more radical excision techniques should not be abandoned unless these results are made available and demonstrate at least equivalent efficacy. Until we understand more about the biology of melanoma in general and ocular surface melanoma in particular, we consider that hard data are required to show that simple local methods of excision are satisfactory for preventing local recurrence and curing the patient as well as for preserving vision.

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In reply

Drs Francis, Coroneo, and Thompson seem to be critical of the technique that we advocate for removal of conjunctival melanoma, stating that our methods are too conservative. In the past, patients with conjunctival melanomas were often treated with simple excision only. That method is associated with increased local recurrence. The method that we have successfully employed for years for limbal melanoma is alcohol corneal epithelectomy, wide excision of the tumor by partial lamellar sclerokeratitic conjunctivectomy using a “no touch” technique, and supplemental double freeze–thaw cryotherapy to the adjacent normal conjunctival margins. This method usually ensures complete tumor removal but leaves the conjunctiva intact so that the patient is comfortable. We have found that this technique is successful in complete tumor removal when histopathologic sections of the specimens are examined and patient follow-up is obtained.

The most peculiar thing about the letter is that the authors state that radical excision techniques should not be abandoned. It is unfortunate, however, that they do not define what they mean by radical techniques. Because the method that we advocate involves a wide excision with tumor-free margins but spares the globe, one must assume that they can only be advocating orbital exenteration for all cases of conjunctival melanoma. If that is the case, we disagree with them. Indeed, the surgical approach to conjunctival melanoma is important in determining the patient’s prognosis. However, complete tumor control can usually be achieved without resorting to orbital exenteration.

In most cases, a circumscribed conjunctival melanoma (arising from a pre-existing nevus or de novo) becomes clinically apparent as a distinct pigmented lesion near the limbus that can be removed successfully using the techniques that we recommend. There is no need for radical surgery, such as orbital exenteration, in such instances. The patient shown in the Figure is just one example of many patients whom we have treated and followed up for several years. These patients have had complete tumor removal with retention of the eye and excellent vision and no evidence of local recurrence or systemic metastasis. They are certainly grateful that we did not subject them to radical orbital exenteration.

In the case of melanoma arising from primary acquired melanosis, the melanoma component can be removed intact by a modification of our standard technique. The flat melanosis in the adjacent conjunctiva can then be treated with double freeze–thaw cryotherapy. We have also had considerable success with that approach.

It is quite possible that Drs Francis, Coroneo, and Thompson have had experience with cutaneous melanoma and they could be correct in advocating wide excision for that disease. However, we believe that there is little place for radical surgery (orbital exenteration) in the initial treatment of most conjunctival melanomas. Other authorities have supported that viewpoint, indicating that exenteration should be reserved as a palliative procedure for advanced stages of the disease. Drs Francis, Coroneo, and Thompson should provide convincing proof that primary orbital exenteration is beneficial with regard to patient survival before their radical views are accepted. At this time there is no such proof and we continue to recommend the methods described in our article.

Parry-Romberg Syndrome

Miedziak et al report a case of Parry-Romberg syndrome and evidence of intracranial vascular malformations. They cite no previous reports of a similar association. The authors acknowledge that the cause of this syndrome is unknown, but they favor the concept that Parry-Romberg syndrome may be the result of an arrested angiogenic process affecting the central nervous system during growth and development. Lending some support to their concept, and yet not mentioned by them, is the occurrence of retinal telangiectasis, Coats disease, and exudative neuroretinopathy in some patients with Parry-Romberg syndrome.

None of the 4 cases reported with retinal and optic nerve involvement had clinical evidence of intracranial vascular malformations. Only 1 of the 4 cases, however, was studied with modern scanning techniques (computed tomography). The authors’ use of the term “malformation” implies a focal dysgenesis of the vascular system of the central nervous system, and yet the vascular changes in the brain and eye may be acquired secondary to some other pathological process underlying the Parry-Romberg syndrome.