Primary Orbital Melanoma With Poliosis and a Palpable Mass

Poliosis, defined as the depigmentation of hair, has been linked with several inflammatory conditions such as blepharitis, herpes zoster, idiopathic uveitis, Vogt-Koyanagi-Harada syndrome, sarcoidosis, vitiligo, Marfan syndrome, and tuberous sclerosis. To our knowledge, we report the first case in the English-language ophthalmic literature of eyelash poliosis as one of the manifesting signs of primary orbital melanoma.

Report of a Case. A healthy 60-year-old white man had swelling above the left eyeball and whitening of the left upper eyelid lashes for 3 months. Visual acuity was 20/20 OD and 20/20 OS. No afferent pupillary defect was detected. He had eyelash poliosis of the left upper eyelid with a mobile, nontender, firm mass in the left anterior orbit superonasally (Figure 1). There was no proptosis, and he had normal ocular surface examination, anterior segment examination, and fundus examination results. Examination results of his right eye were unremarkable. Magnetic resonance imaging with contrast revealed a well-circumscribed, diffusely enhancing mass in the superonasal region of the left anterior orbit (Figure 2). Excisional biopsy of the mass was achieved through an eyelid crease incision. A histopathologic report revealed solid sheets of spindle cell neoplasm. Melan-A immunohistochemical staining showed cytoplasmic immunoreactivity in the neoplastic cells. The histopathologic findings were consistent with the diagnosis of orbital melanoma. No inflammatory cells were seen in the mass.

An extensive systemic evaluation showed no evidence of distant metastasis or primary tumor elsewhere. The mass was diagnosed as primary orbital melanoma, although primary tumor elsewhere cannot be totally ex-
cluded. The patient refused to have an exenteration of the left orbit performed and instead preferred to undergo close observation. Three months later, the patient had local recurrence at the nasal part of the superior fornix but no evidence of distant metastasis. The patient underwent exenteration of the left orbit with a split-thickness skin graft covering the orbital bones. No inflammatory cells were seen in histopathologic examination following exenteration around the eyelashes in the area involving poliosis. The eyelashes had only absence of melanin without disruption of the anatomy of the eyelashes.

At his last follow-up examination (6 months following exenteration), the patient was doing fine and exhibited no signs of local recurrence or distant metastasis.

Comment. Primary orbital melanomas, which account for 1% of all orbital tumors, can occur de novo or arise from congenital ocular melanocytosis or hypercellular blue nevus. Patients with primary orbital melanoma generally have a slow onset of painless proptosis. Primary orbital melanomas arising from a blue nevus show related congenital, thickened, periorbital, pigmented nevus of slate gray–blue color, whereas those arising from melanocytosis display flat blue–gray pigmentation as well as episcleral pigmentation. Our patient is the first to our knowledge to manifest poliosis of the eyelashes with primary orbital melanoma.

Poliosis of the eyelashes has been previously reported in a patient with conjunctival melanoma. Although the pathogenesis of acquired poliosis is not known, several possible explanations have been put forward, including an inflammatory destruction of the melanocytes in the hair follicle, apoptosis of the follicular melanocytes, or a targeted autoimmune response with the molecular mimicry. The development of poliosis in patients with melanoma could be related to an immune response triggered by the cross-reaction of malignant melanoma cells with normal follicular melanocytes.

In summary, acquired poliosis of the eyelashes is usually a sign of benign conditions such as blepharitis, sarcoidosis, herpes zoster, Vogt-Koyanagi-Harada syndrome, vitiligo, and tuberous sclerosis. Based on our study, the presence of acquired poliosis in patients with an orbital mass needs to raise the suspicion of primary orbital melanoma.

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COMMENTS AND OPINIONS

Estimating Cataract Surgery Rates

It was with much interest I read the insightful and careful study by Lewallen and colleagues in which they used data from a number of population-based studies in Africa to assess what might be the appropriate cataract surgery rate (CSR). They show, not unexpectedly, that the CSR changes with the level of visual acuity used as an indicator for cataract surgery, whether one or both eyes are operated on, and the proportion of older people in the population.

I was interested to compare the rates found by Lewallen and colleagues with those that we reported for Aus-