 Orbital Ganglioneuroma in a Young Healthy Person

Ganglioneuroma is considered to be the benign counterpart of neuroblastoma. Orbital involvement of ganglioneuroma is extremely rare, and to our knowledge only 2 cases have been reported. One was a case of direct extension to the orbit from an adjacent sinus, and the other was in a patient who had a history of stage IV neuroblastoma. This is the first reported case of orbital ganglioneuroma in a young healthy person.

Report of a Case. A 15-year-old Korean boy had progressive proptosis of the right eye during a 4-year period (Figure 1). The proptosis had advanced at an increasing rate in the 6 months leading up to the patient’s initial visit. His general medical history was unremarkable. Eye examination disclosed 2 mm of proptosis in the right eye. His best-corrected visual acuity was 20/20 OU. Intraocular pressures were 16 mm Hg OD and 14 mm Hg OS, and the pupils were reactive to light bilaterally with no afferent pupillary defect. Funduscopic examination revealed no abnormal findings. Magnetic resonance imaging and computed tomography of the orbit obtained before biopsy showed an irregularly enhancing mass (2.7 × 1.7 × 2.7 cm) in the right, infratemporal, extracranial space along with bony hypertrophy of the zygoma (Figure 2 and Figure 3). The neoplasm appeared homogeneous in composition and there was no evidence of bony metastasis. During incisional biopsy, a lesion with a gray-tan surface was found to be adherent to the adjacent bony structures. Histopathologic examination showed mature ganglion cells in a neurofibrillary matrix with no neuroblastic component. Immunoreactivity for S-100 protein and synaptophysin was positive in the ganglion cells (Figure 4). These findings were consistent with ganglioneuroma. A secondary operation for debulking the mass and reshaping.
ing the zygoma was performed. Our patient has been followed up for more than 6 months, with no evidence of recurrence. He does not have diplopia or proptosis, and visual acuity is 20/20 OU.

Comment. Tumors of the sympathetic nervous system include neuroblastomas, ganglioneuroblastomas, and ganglioneuromas. All are derived from neural crest cells and are considered to represent different maturational steps of a unique neoplasm.

Ganglioneuromas are well-differentiated, benign tumors composed of mature Schwann and sympathetic ganglion cells. They are associated with neurofibromatosis type I and multiple endocrine neoplasia. There are 2 subtypes of ganglioneuroma: maturing and mature. The maturing subtype is composed predominantly of ganglioneuromatous stroma with scattered collections of differentiating neuroblasts and/or maturing ganglion cells. The mature subtype is composed of mature Schwannian stroma and ganglion cells. It is located most frequently in the posterior mediastinum and the retroperi-
toneum; other sites of origin are much less common. A case of a ganglioneuroma arising in the orbit has been reported, but the patient had a history of neuroblastoma. Ours is the first reported case of an orbital ganglioneuroma in a healthy person.

Ultrasonography, computed tomography, and magnetic resonance imaging provide only an unspecified diagnosis that has to be confirmed using pathologic studies.

Ganglioneuromas are benign neoplasms, so neither extensive surgical resection nor chemotherapy is usually necessary, provided the surgical specimen is sufficient to allow histopathologic study and to assure no malignant components are present. Excision may be considered when the pathologic diagnosis is vague or visual function is impaired by the mass. In our case, progressive proptosis and corneal complications prompted an excisional biopsy and a secondary debulking operation.

Ganglioneuromas have a tendency to remain silent for an extended period of time, and they are often associated with long-term disease-free survival. Some au-
thors have reported malignant transformation, either spontaneously or after radiotherapy. Therefore, considering the potential growth of ganglioneuromas in patients with incomplete resection, regular follow-up examinations and imaging are necessary.6

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COMMENTS AND OPINIONS

Misleading Titles Cause Confusion

As much as I respect the opinion of my erudite colleague, Dr Sommer, I believe he may have misread the study by Solomon and colleagues1 in his recent editorial.2

Sommer reports that the study of Solomon “found that a single dose of (azithromycin) virtually eliminated infection for 2 years.” While it is true that a single dose of azithromycin was given and the title of the article reemphasizes this misconception, the authors do state that “for ethical reasons at 6, 12 and 18 months, we gave 2 tubes of tetracycline to subjects with active disease.”1

This article, with its misleading title, has been frequently misquoted as reflecting the effect of a single dose of azithromycin where, in fact, it truly represents a hybrid treatment model with initial azithromycin and 6 monthly retreatments with tetracycline ointment. A now somewhat outdated Cochrane review considered topical tetracycline and oral antibiotics to be equally effective.3

Programmatically, it is of particular importance that policy makers clearly understand that a single dose of azithromycin is highly unlikely to eliminate trachoma in any area. Six or 12 monthly retreatments with high coverage may be required for a number of years, and programs need to be prepared to commit the resources to achieve this.

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In reply

Dr Taylor is, of course, quite correct. Interpretation of the results reported by Solomon et al1 is complicated by their having treated subjects with clinical disease at 6, 12, and 18 months with topical tetracycline ointment. The effect this might have had on final infection rates at 24 months is hard to judge. By the 6-month examination, before any additional topical treatment was applied, the infection load had been reduced by more than 90% and the prevalence of infection by more than 80%.2 While the effect of topical and systemic antibiotic treatment is judged to be similar, this depends to a large degree on the level of compliance with the treatment regimen, which can be problematic in the case of topical tetracycline ointment.

A number of other caveats listed in my original review of the article by Solomon et al1 are relevant to interpreting and extrapolating results to other populations and settings.

As my more recent commentary2 noted, the ideal frequency and duration of communitywide azithromycin treatment for the eradication and control of trachoma remains to be determined and will likely vary with the particular conditions of the community in which it is used. I thank Dr Taylor for pointing out yet another complexity in interpreting the results of this important clinical trial.

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How Was Histologic Analysis of the Descemet Membrane Done After DSAEK?

We read with interest the article by Heindl et al regarding histologic and transmission electron microscopy analysis of 20 Descemet membrane specimens obtained after Descemet


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