Corneal Juvenile Xanthogranuloma in a 4-Month-Old Child

Juvenile xanthogranuloma (JXG) is a benign histiocytic disease of uncertain pathogenesis that usually appears as a cutaneous lesion(s) in the head and neck region of young children. Diagnosis is based on histological examination that shows a nodular, dense infiltrate of polygonal or spindled mononuclear cells with moderate amounts of cytoplasm and scattered Touton giant cells. Juvenile xanthogranulomas are typically positive for CD68 and factor XIIIa immunostains and are negative for CD1a and S100 protein; this information is helpful in differentiating JXG from other histiocytic proliferations. We present the case of a 4-month-old child with corneal JXG.

Report of a Case. The mother of a 4-month-old infant boy noted an enlarging, raised mass in the medial corner of his right eye. A biopsy performed by a local ophthalmologist was interpreted as noncaseating granulomatous inflammation. Despite normal findings on systemic evaluation, sarcoidosis was diagnosed. He received topical steroid therapy and was referred to us.

Examination of the patient’s right eye at 11 months revealed an elevated 12 × 8-mm yellow epibulbar mass involving the temporal conjunctiva, overlying the medial rectus muscle, extending onto the cornea to the nasal pupillary margin, and invading the corneal stroma (Figure 1). No prominent blood supply was present. His vision was centered, steadied, and maintained. Cycloplegic refraction was plano +2.50 × 90 OD and +0.75 sphere OS. A dilated fundus examination was normal. No refractive error was present. A review of original biopsy material showed a JXG, with a dense nodular infiltrate beneath the corneal epithelium comprising polygonal cells with moderate amounts of eosinophilic cytoplasm and rare Touton giant cells (Figure 2). Immunohistochemical studies showed strong positivity for vimentin and CD68, weak staining for factor XIIIa, and negative staining for S100 protein and CD1a. No evidence of necrosis was noted. The patient was treated with a topical steroid regimen of loteprednol etabonate every 2 hours, and fluorometholone ointment nightly with no complications. Follow-up examinations showed regression of the mass during 12 months, with associated scarring of the cornea.

Comment. A benign proliferation of cells of the monocyte/macrophage or dendritic cell lineage, JXG represents one of several “non-Langerhans cell”–type histiocytoses. In 80% of cases, JXG appears in the first 6 to 9 months of life as solitary or multiple red or brown cutaneous nodules. The lesions most commonly occur in the head and neck region, the upper part of the trunk, or the proximal limbs. The natural history is usually one of spontaneous involution after several months. Ocular involvement occurs in 10% of cases of JXG. The iris is the most common location, followed by the eyelid, choroid, optic nerve, retina, conjunctiva, limbus, and cornea. Thirteen cases involving the corneoscleral limbus are documented, 6 exhibiting extension onto the cornea in patients aged 18 months through 30 years. Although topical steroid therapy has proven successful in managing lesions of the iris, its usefulness for JXG involving the cornea has not been established. Of the 6 cases with corneal involvement, 4 were treated with excision and graft placement, one with excision followed by irradiation, and the last with simple excision. In deciding on a treatment strategy, we weighed the risks of topical steroids against those of surgery. Because this lesion did not produce a significant refractive error, we...
Conjunctival Keratoacanthoma in an Asian

Keratoacanthoma, a benign epithelial tumor that grows rapidly and shows spontaneous regression, has a characteristic central crater filled with keratin. Keratoacanthomas arise most commonly in sun-exposed skin and only rarely in mucosa. The few reported cases of conjunctival keratoacanthoma have occurred mostly in whites. Here we describe what we believe to be the second reported Asian case of conjunctival keratoacanthoma.

Report of a Case. On January 3, 2001, a 39-year-old Japanese man noted hyperemia of the right bulbar conjunctiva associated with a foreign-body sensation. One week later he consulted an ophthalmologist because the hyperemic bulbar lesion had become elevated. He was diagnosed as having phlyctenular conjunctivitis and was treated with topical 0.1% dexamethasone and antibiotic eye-drops. This limbal lesion continued to grow and on February 5, 2001, the patient was referred to the Department of Ophthalmology at Kyushu University, Fukuoka, Japan.

His corrected visual acuity was 20/20 OU. Slitlamp examination of the right eye revealed a white, firm, and dome-shaped mass at the nasal limbus. The lesion had a central crater filled with white material. The mass was approximately 4 mm in diameter and was surrounded by hyperemia (Figure 1). A delle was noted on the cornea adjacent to the mass. We curetted the white material plugging the crater; however, 1 week later the crater had refilled with similar material. We then totally excised the tumor.

Results. Findings from histopathologic examination showed a central crater containing a keratotic plug surrounded by acanthotic conjunctival epithelium that included horn pearls (Figure 2). The tumor cells had abundant, glassy, eosinophilic cytoplasm and small nuclei. In the deeper regions, tumor cells showed cellular atypia and infiltrative growth associated with an inflammatory reaction. Based on the overall shape of the lesion, a crater filled with keratin and surrounded by epidermis extending in a liplike manner over its sides, we diagnosed the mass as a keratoacanthoma. One year after resection no recurrence has been observed.

Comment. Keratoacanthoma ordinarily arises in skin containing hair follicles, and rarely subungually, in the palms, or in buccal or conjunctival mucosa. Keratoacanthoma of the conjunctiva was first reported by Freeman et al in 1961. Only 12 cases have been reported in the 40 years since the first account, mainly in whites; one patient was a mulatto, and another was black. To our knowledge, only 1 Asian case of conjunctival keratoacanthoma has been reported previously in the English-language literature, and this case occurred in a Thai individual.