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Bilateral Conjunctival Nevi in a 8 year old boy

Conjunctival nevi are the most common benign tumors of the ocular surface. They arise from a proliferation of melanocytes in the basal conjunctival epithelium. These lesions in children often present a diagnostic dilemma; as they can be amelanotic, enlarge or become inflamed. We report a case of bilateral conjunctival nevi in an 8 year old boy confirmed by histopathology. The bilaterality of the lesions in a young patient was highly atypical and prompted excision to confirm the clinical diagnosis of conjunctival nevus.

Case Presentation

An 8 year-old boy was referred to our practice for evaluation of a conjunctival lesion in the left eye (September 2017). On examination, vision was 20/20 in both eyes. The left eye demonstrated a flat pinkish 2mm by 4mm amelanotic lesion at the limbus (see figure 1). A similar sized lesion was also seen in the right eye with a small amount of pigment noted and an adjacent cyst. (figure 2). The eyelids, cornea and anterior chamber and fundus evaluation were all unremarkable bilaterally

A discussion was had with the patient's parents and it was decided to monitor the lesions. Serial slit-lamp photos were taken to monitor the lesions. At the 8 months follow up, the mother had noted intermittent inflammation and enlargement of the lesion in the left eye (May 2018). Slit lamp photos confirmed an increase in size of the lesion in the left eye (figure 3,4). After discussion, the decision was made to proceed with a bilateral excisional biopsy.

Histopathologic examination of the lesions demonstrated bilateral compound melanocytic nevi. The excised lesions disclosed prominent proliferation of nevus cells at the epithelial/ sub-epithelial junction in both eyes (Fig. 5). Conspicuous nests were frequently seen within the epithelium. There was a noticeable lessening of the nests at the edges of the lesion. (Fig. 6). The majority of the subsurface nevus cells were in junctional nests situated within squamous clusters with focal central clear spaces. The lesions in the left eye were similar except for a more prominent lymphocytic reaction and increased vascularity.

Discussion

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Conjunctival melanocytic lesions are typically classified into the following categories: nevus, benign acquired melanosis and primary acquired melanosis. The vast majority of conjunctival melanoma arises from primary acquired melanosis, though it may also arise de novo or from existing nevi [1]. Conjunctival melanoma in children is rare [2]. Among melanocytic lesions, conjunctival nevi are the most common tumors, constituting more than 50% of all conjunctival tumors [1,3]. They typically occur before the age of 20. Although changes in size and pigmentation are common, transformation to malignancy is very rare [3].

Clinically, conjunctival nevi can be identified by a number of characteristics. They normally present during adolescence (versus melanoma, which presents most commonly in the 6th and 7th decade of life) [4].They are normally solitary and move freely over the sclera. Pigmentation can be patchy and often contains cystic components. Conjunctival nevi generally do not extend onto the cornea.

Shields et al described clinical features and natural course of conjunctival nevi in a cohort of 410 patients.[5] In their study, 89% of patients were caucasian and 16% of the nevi were nonpigmented. The most common location was the bulbar conjunctiva, followed by the caruncle and plica semilunaris. Over the course of observation (mean 11 years), 5% of lesions became darker and 7% increased in size. Interestingly, 43% of patients reported that the lesion enlarged in size prior to initial examination.

Histologically, conjunctival nevi demonstrate proliferations of melanocytes in the conjunctival epithelium and stroma. The amount of melanin varies from case to case. Very often they contain inclusion cysts with abundant goblet cells. Conjunctival nevi have historically been classified as junctional or compound. Though this classification has no impact on management, junctional nevi are typically only seen in children.

Inflamed nevi can contain eosinophils and lymphocytic infiltrate consisting primarily of plasma cells. They are most commonly seen in patients with chronic allergic conjunctivitis [6]. One retrospective study examined excised conjunctival nevi in patients under 20 years of age who had been referred for growth of the lesion [6]. They found that 75% of the lesions could be histologically defined as "inflamed." On histology, the dense inflammatory infiltrate can cause confusion for lymphoma. Additionally, the subepithelial melanocytes can show reverse maturation (larger nuclei and more abundant cytoplasm), which can appear concerning [7].

Amelanotic conjunctival nevi can resemble granuloma, ocular surface neoplasia, lymphoid hyperplasia, leukemia, sarcoid, JXG, Rosai-Dorfman disease, Rheumatoid and Pseudorheumatoid nodules, colloid milium and amyloidosis. Amelanotic nevi should be differentiated from conjunctival lymphoma. When inflamed they may also appear similar to episcleritis, nodular scleritis, and allergic conjunctivitis,

The vast majority of melanocytic lesions of the conjunctiva can be observed with serial slit lamp photographs. If major changes are noted these lesions can be excised. Shields et al proposed that the increase in size of conjunctival nevi is likely due to enlargement of cysts, increased pigmentation of amelanotic areas and inflammation within the nevus. Indications for removal of conjunctival lesions in children include suspicion of malignancy (detail) or other pathology listed previously, cosmesis and risk for amblyopia (induced astigmatism).

Figures

1. Clinical Photo showing lesion Left eye at presentation
2. Clinicla Photo showing lesion Right eye at presentation
3. Clinical Photo of the left eye prior to excision showing enlargement.
4. Clinical Photo of left eye higher power prior to excision demonstrating thickening and increased vascularity.
5. H & E 20X Left eye lesion of conjunctiva with areas of infiltrate and vascular engorgement.
6. H & E 100X Characteristic junctional nevus with focal infiltration of small lymphocytes.
7. H & E 20X Right eye showing junctional nevus with cyst..

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