Case report

A complex choristoma presenting as a salmon patch lesion in the bulbar conjunctiva

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ABSTRACT

Purpose: We describe a rare case of a complex choristoma appearing as a salmon-patch lesion in the nasal conjunctiva. While benign, complex choristomas are grossly indistinguishable from malignant lesions, and an excisional biopsy is warranted to confirm the diagnosis.

Observations: A 31-year-old man presented with an elevated glistening pink mass on the conjunctiva. An excisional biopsy with a "no-touch" technique was performed, followed by placement of an amniotic membrane graft without postoperative complications. No subsequent medical treatment was pursued after the biopsy and histopathologic evaluation. Histopathology confirmed a diagnosis of a complex choristoma with sections consisting of fibroadipose tissue with cartilage, smooth muscle, and nerves. Histopathology was negative for malignant cells without morphologic evidence of a lymphoma.

Conclusions and Importance: Conjunctival tumors are difficult to distinguish clinically, and a differential diagnosis often includes the possibility of a malignancy. Histopathologic diagnosis may be required to distinguish between various entities. In our case, a salmon-patch conjunctival tumor was biopsied and confirmed to be a benign complex choristoma.

1. Introduction

Choristomas of the eye are characterized by normal tissue derived from germ cell layers that are foreign to the site of the lesion. Prior case reports and case series have described choristomas with adipose, cartilaginous, bony, smooth muscle, or glandular tissues. Histopathologic evaluation and analysis is often necessary to confirm the tissue of origin, as the types of tissue comprising the choristoma are not always apparent on gross examination. The lesions themselves may be difficult to appreciate and may be found incidentally on biopsy. One study reported several cases of subpterygial cartilaginous choristomas in a cohort of patients undergoing pterygium removal. On histopathologic evaluation, these choristomas were embedded in Tenon’s capsule, deep in the caruncle with an overlying pterygium.

Complex ocular choristomas are exceedingly rare. These tumors contain heterotopic tissues and are characterized by the presence of at least two germ cell layers within the lesion. Many reported complex choristomas appear as limbal dermoids or dermolipomas. Few have been reported to involve the eye, and even fewer to involve the epibulbar surface. Such complex choristomas are usually asymptomatic but can involve both the conjunctiva and cornea, leading to amblyopia in children. Although these lesions do not typically undergo significant growth, large lesions can protrude through the eyelid affecting eyelid closure. Complex choristomas are also associated with systemic syndromes. For example, Goldenhar syndrome is associated with limbal dermoids and lipodermoids. Linear nevus sebaceous syndrome (LNSS), a rare sporadic phakomatosis, is characterized by multiple cutaneous nevi, sebaceous lesions, and systemic abnormalities. Nevus sebaceus of Jadassohn is a syndrome of facial sebaceous nevi, seizures, mental retardation, arachnoid cysts, and cerebral atrophy.

Case Report:

The patient was a 31-year-old man referred for evaluation of a left nasal conjunctival lesion that had been present for two years. According to the patient, the lesion had been stable without growth or significant change. He denied ocular irritation or changes in his vision from the...
conjunctival lesion. He denied fevers, night sweats, and weight loss. His ocular history was notable for refractive error, and his past medical and surgical history was notable for seasonal allergies and a jaw fracture repair. He did not use medications and had no known drug allergies.

His best-corrected visual acuity was 20/25 OD and 20/20 OS. Extraocular motility, confrontation visual fields, and intraocular pressures were normal. On external exam, small, non-tender submandibular lymph nodes were palpable. The slit lamp examination showed an elevated, glistening, well-circumscribed salmon-colored patch, measuring 9.5 × 7.5mm, in the nasal bulbar conjunctiva with overlying superficial vasculature (Fig. 1). There was concern for enlarged tortuous feeder vessels leading into the mass nasally. The dilated fundus examination was unremarkable in both eyes.

An excisional biopsy was performed utilizing a "no-touch technique," given the possibility of a malignant lesion, with removal of the conjunctival tumor and placement of an amniotic membrane graft. Histopathology revealed fibroadipose tissue with cartilage, smooth muscle, and nerves (Fig. 2). This confirmed the rare diagnosis of a complex choristoma with no morphologic evidence of a lymphoma or malignant cells. In the contralateral eye, the conjunctival tissue showed benign surface keratinization and was also negative for malignant cells.

Postoperatively, the patient underwent a repeat slit lamp examination showing stable resection of the bulbar conjunctiva without recurrence of the lesion (Fig. 3). Repeat examinations up to 19 months later continued to show a well-healed excision site with no evidence of recurrence.

2. Discussion

The differential diagnosis for an elevated salmon-colored conjunctival patch most commonly includes benign lymphoid hyperplasia and conjunctival lymphoma. Benign diagnoses that may be considered include a non-pigmented nevus, dermoid or lipodermoid, pyogenic granuloma, lymphangioma, amyloid deposition, papilloma, fibroma, or as in our case, a complex choristoma. Malignant diagnoses that should be considered include ocular surface squamous neoplasia, amelanotic melanoma, sebaceous carcinoma, or metastatic lesions.

General principles of managing conjunctival tumors differ depending on the location (limbal versus extralimbal) and the layer of the tumor involvement (epithelial versus stromal). Histopathology of benign and malignant lymphoproliferative lesions on gross examination are difficult to distinguish. Both may appear smooth, mobile, and salmon-pink. Given this challenge, we pursued a biopsy due to the suspicious appearance of the lesion and the patient’s possible lymphadenopathy.

One report recommends that if there are no signs of systemic lymphoma, as in our patient, a small lymphoid-appearing mass can be excised completely; however, a larger mass may require an incisional biopsy. Lesions should be treated as malignant when doing a biopsy, and a “no-touch” technique should be considered. The “no-touch” technique is widely accepted as a standard technique for such ocular surface tumors, avoiding direct manipulation of the lesion to prevent tumor seeding.

Interestingly, our patient noticed the mass two years prior to presentation at the age of 29. While choristomas are congenital, they are known to grow slowly over time if not excised. While we do not have information regarding the choristoma prior to his presentation, it is likely that he did not notice the presence of the choristoma for many years given that even on his initial presentation, he noted no ocular irritation or visual symptoms. He has recovered well and continues to be followed periodically.

Patient consent: Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

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Conflicts of interest

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Authorship

All authors attest that they meet the current ICMJE criteria for
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