Epibulbar conjunctival Schwannoma in the differential diagnosis of subconjunctival masses

Subkonjonktival kitlelerin ayırıcı tanısında epibulber konjonktival Schwannoma

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Abstract
Here, in this study a case of a 33-year-old man presented with a 5-year history of subconjunctival mass is reported. During the immunohistochemical analysis, the tumor cells were strongly positive for S-100 protein. Herein, presenting this rare case of ours, we imply that ophthalmologists should consider schwannoma in the differential diagnosis of subconjuntival masses.


Key words: Subconjunctival mass, schwannoma, S-100 protein

Özet
Bu makalede 33 yaşında, 5 yıllık bir subkonjonktival kitle öyküsü olan erkek hastayı sunduk. İmmünohistokimyasal analizde tümör hücreleri S-100 proteine pozitif yanıt verdi. Bu nadir görülen vakayı sunarak oftalmologların subkonjonktival kitlelerin tanısında schwannomayı da düşünmelerini istedik.

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Anahtar sözcükler: Subkonjonktival kitle, schwannoma, S-100 protein

Introduction
Schwannoma is a tumor of Schwann cells. It arises from peripheral or cranial nerves. It can be solitary or associated with neurofibromatosis (NF) type 1 and 2 or schwannomatosis [1]. Although it is mostly benign, it may rarely undergo malignant transformation, especially in NF [2,3]. Schwannomas form only 1% of orbital tumors and they rarely involve conjunctiva [3-6]. Herein, presenting this rare case of ours, we draw attention of ophthalmologists towards schwannomas in the differential diagnosis of subconjuntival masses.

Case Presentation
A 33-year-old man presented to our clinic with a 5-year history of subconjunctival mass on his left eye. The mass had been growing in the last 3 months. His visual acuity was 20/20 bilaterally. On slit-lamp examination, a yellowish, well-circumscribed, round and translucent mass was present under the mobile medial bulbar conjunctiva. Ophthalmic and systemic evaluation of the patient was normal. The family history was unremarkable.

During surgery, the lesion was observed to be not adherent to the conjunctival epithelium and sclera. A 5×3×2 mm mass was excised from the superficial sclera. Histopathologic examination revealed an encapsulated tumor composed of slender or spindle-shaped cells with eosinophilic cytoplasm and oval nuclei, showing a fascicular or palisade arrangement (Figure 1). During the immunohistochemical analysis, the tumor...
cells were strongly positive for S-100 protein (Figure 2). Accordingly, the patient was diagnosed with epibulbar schwannoma.

Figure 1. Fascicular or palisade arrangement of proliferated tumor cells (H&E staining, x200).

Figure 2. Diffuse and strong S-100 positivity in the tumor cells (Immunoperoxidase, x400).

Systemic evaluation including brain magnetic resonance imaging (MRI) was performed but no other lesions like neurogenic tumor, neurofibromatosis, or schwannomatosis could be found.

Discussion

Conjunctival schwannoma can be seen in any age group, ranging from 12 to 72 years [3,7]. It can be seen in any location under the tarsal and bulbar conjunctiva [5]. In the bulbar conjunctiva, it has been found horizontally at the 3 and 9 o’clock meridians, inferiorly, and superotemporally [3,5,7]. In our patient, the lesion was at the 9 o’clock meridian and the patient was 33-year-old. Scleral involvement has been reported [3,7]. However, in our patient there was no scleral involvement.

The differential diagnosis of these masses includes similar lesions like dermoid, dermolipoma, hemangioma, ectopic lacrimal gland, melanoma, conjunctival intraepithelial neoplasia, and squamous cell carcinoma [3,6]. Conjunctival schwannoma is usually seen as a subconjunctival, amelanotic, well-circumscribed solid mass. Epibulbar dermoids are usually seen as well-circumscribed oval masses of the ocular surface that arise from the bulbar conjunctiva and almost always protrude across the limbus onto the cornea. Often, hair follicles are seen on the dermoid. Dermolipom is a congenital lesion of the bulbar conjunctiva, generally occurring towards the lateral canthi between the superior and lateral recti muscles. It is covered by a thick epidermal epithelium, in which there may be hair or evidences of glandular appendages with the secretions [8]. Hemangioma is examined as a flat arrangement of intertwining, mildly dilated blood vessels, usually on the bulbar conjunctiva [9]. Ectopic lacrimal gland tissue is another mass which may be found in the caruncle or bulbar conjunctiva [10]. Conjunctival melanoma is usually observed as a brown to tan elevated mass on the bulbar conjunctiva surrounded by a bed of flat primary acquired melanosis in middle-aged patients [11]. Conjunctival intraepithelial neoplasia appears as a fleshy, sessile or minimally elevated lesion usually at limbus in the interpalpebral fissure and less commonly in the fornicial or tarsal conjunctiva. A white plaque may occur on the surface of the lesion due to secondary hyperkeratosis [12]. Conjunctival squamous cell carcinoma is clinically similar to conjunctival intraepithelial neoplasia. However, it may be larger and more elevated than intraepithelial neoplasia.

It is sometimes difficult to make the correct diagnosis of subconjunctival masses by slit-lamp examination. The diagnosis of conjunctival schwannoma is made histopathologically showing 2 patterns, including Antoni A pattern (composed of sheets of palisading spindle cells with spindle-shaped nuclei) and Antoni B pattern (composed of haphazardly arranged spindle and multipolar cells in myxoid stroma). The most important finding for the diagnosis is the strong reactivity to S100 protein in the background of a characteristic histology.

In conclusion, the reason why we reported our patient was two-fold. First, presenting this rare case, we aimed to highlight that schwannoma must be kept in mind in the differential diagnosis of epibulbar conjunctival lesions. Second, due to the possibility of malignant transformation and/or association with NF or schwannomatosis, we
would like to bring to the attention of physicians to screen the whole body once they are faced with a solitary schwannoma.

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References