Schwannoma Of The Eyelid

Gözkapağı schwannoması

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Abstract

A 21-year-old woman presented with a nodular lesion, a history of a slowly enlarging, mobile, 2 cm in diameter, located on the nasal region of the left upper eyelid. She reported no history of previous trauma or surgery regarding the lesion. The mass had a cystic appearance and cartilaginous consistence; it was non-adherent to the skin or the underlying tissue. Excisional biopsy revealed the characteristic histopathologic features of a schwannoma. After complete excision, there was no recurrence at 6-months follow-up. Schwannomas usually occur as single and slow growing benign tumours thought to arise from the Schwann cells of the peripheral nerves involving commonly the cranial and spinal nerve roots. These tumors preferentially involve sympathetic, cervical, and vagus nerves. Other sites involved include the face, neck, scalp and hands. Examples have been recorded in the tongue, palate and larynx. In most cases, while Schwannoma is sporadically manifested as a single benign neoplasm, the presence of multiple Schwannoma is usually indicative of neurofibromatosis–2. Our patient had isolated eyelid Schwannoma with no family history or clinical findings of neurofibromatosis–1 or neurofibromatosis–2. Schwannomas of ophthalmic interest are rare, although they have been reported in relation to the orbit (1% of orbital tumors), and infrequently to the uveal tract, conjunctiva and sclera. Eyelid schwannoma is a rare, slowly growing, benign neoplasm. This case suggest that schwannomas should be included in the differential diagnosis of any eyelid mass lesions and that accurate histological diagnosis and complete excision should be carried out in these cases.

Key words: Eyelid, schwannoma, tumour.

Özet

Yirmibir yaşında olgu sol üst göz kapağı nazal kısmında yerleşmiş son 1 yıl içerisinde yavaş bir şekilde büyüyen, 2 cm çapında hareketli bir kitle şikayeti ile başvurdu. Olgunun hikayesinden travma ve kitle ile ilgili bir cerrahi girişimin olmadığı anlaşıldı. Fizik muayenede kitle kistik görünümde olup etrafındaki dokulara yapışık değildi. Gerçekleştirilen eksizyonel biyopside schwannomaya özgü karakteristik histopatolojik bulgular saptandı. Kitlenin tam olarak çıkarılmasından sonraki 6 aylık takipte herhangi bir nüks ile karşılaşılmadı. Schwannomalar genellikle tek ve yavaş olarak büyüyen sıklıkla kraniyal ve spinal sinir köklerinin dahil olduğu periferal sinirlerin schwan hücrelerinden kaynaklandığı düşünülen iyi huylu tümörlerdir. Bu tümörler daha çok sempatik, servikal ve vagus sinirlerini tercih ederler. Diğer yerleşim yerleri yüz, boyun, kafa derisi ve eller olup dil damak ve larenks kaynaklı schwannomalar da bildirilmiştir. Çoğu vakada schwannomalar iyi huylu, tek tümör olarak görülürken birden fazla schwannoma varlığı genellikle nörofibromatozis-2 varlığının göstergesidir. Olgumuz da herhangi bir aile hikayesi veya nörofibromatozis 1 ve nörofibromatozis 2'e ait klinik bulgu mevcut olmadığı için izole gözkapağı schwannoması kabul edildi. Schwannomaların göz tutulumu nadir olmakla birlikte orbita (orbita tümörlerinin %1'i), uvea, konjonktiva ve sklera tutulumu bildirilmiştir. Gözkapağı schwannomaları nadir, yavaş büyüyen iyi huylu tümörlerdir. Bu olguda olduğu gibi gözkapağı kitlesel lezyonlarının ayırıcı tanısında schwannomalar akılda tutulmalı ve tam bir eksizyon ile birlikte histopatolojik değerlendirme gerçekleştirilmelidir.

Anahtar kelimeler: Gözkapağı, schwannoma, tümör.

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Introduciton

Schwannomas are benign tumors of neuroectodermic origin derived from the cells of schwann that form the neural sheath. These tumors preferentially involve spinal nerve roots, and the sympathetic, cervical, and vagus nerves. They may be associated with neurofibromatosis, but solitary schwannoma at any site is not usually associated with this entity. Solitary schwannoma of orbital region is rare and have been reported uvea, conjunctiva and eyelid in literature [1–7].

Case

A 21-year-old woman presented with a nodular lesion, a history of a slowly enlarging, mobile, left upper eyelid mass of 1 year duration, 2 cm in diameter, located on the nasal region of the left upper eyelid(Fig. 1).

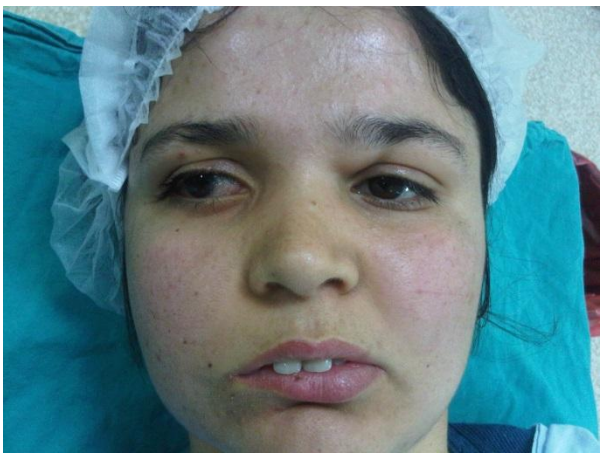

Figure 1. 21-year-old woman with mass involving left upper eyelid.

She reported no history of previous trauma or surgery regarding the lesion. The mass had a cystic appearance and cartilaginous consistence; it was non-adherent to the skin or the underlying tissue. Physical examination was unremarkable and no signs of neurofibromatosis were present. The results of anterior segment, lens and fundus examinations of both eyes were unremarkable.

The lesion was initially thought to be an dermoid cyst and it was surgically excised by means of an anterior approach through the skin. The specimen was an apparently encapsulated smooth surfaced nodule measuring 2 x 1.4 x 1 cm. Microscopically, a lesion composed of fusiform cells arranged in intertwined bundles was revealed. The nuclei were oval and formed palisades. There was no mitotic activity(Fig. 2).

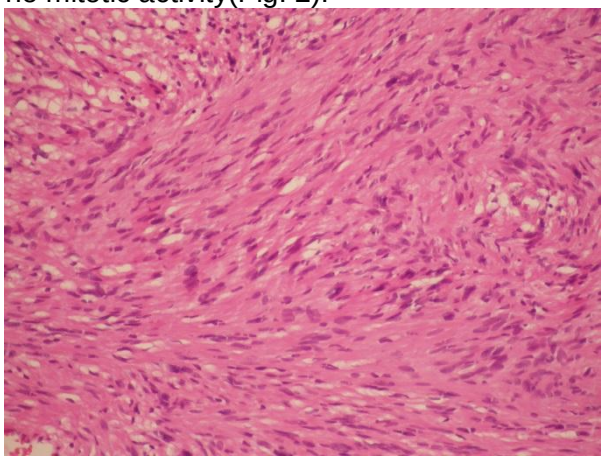

Figure 2. HxE, x10; Histopathology of the tumour.

Immunocytohistochemical analysis for S–100 protein showed a strongly positive reaction(Fig. 3). These findings established the diagnosis of schwannoma. After 6 months of follow-up, no evidence exists of local recurrence.

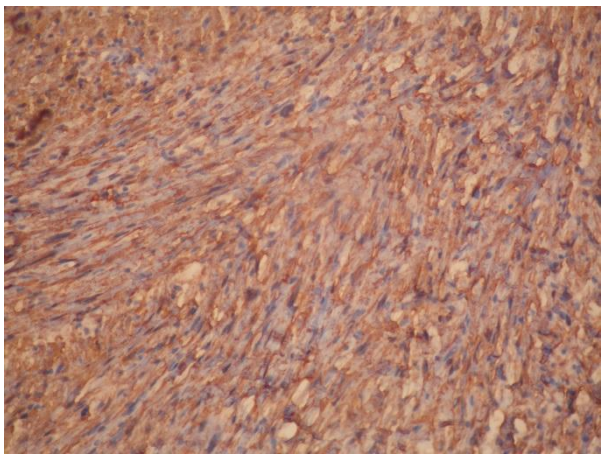

Figure 3. S–100, x20; Immunochemistry for S–100 protein is strongly positive.

Discussion

Schwannomas usually occur as single and slow growing benign tumours thought to arise from the Schwann cells of the peripheral nerves involving commonly the cranial and spinal nerve roots. Other sites involved include the face, neck, scalp and hands. Examples have been recorded in the tongue, palate and larynx [1]. In most cases, while Schwannoma is sporadically manifested as a single benign neoplasm, the presence of multiple Schwannoma is usually indicative of neurofibromatosis–2. Our patient had isolated eyelid Schwannoma with no family history or

clinical findings of neurofibromatosis–1 or neurofibromatosis–2. Eyelid schwannoma is a rare, slowly growing, benign neoplasm. They are relatively common, occurring in many sites throughout the body, making up to 1% of orbital tumors [8]. However they are rarely found on eyelid, so they are not usually considered in the differantial diagnosis on eyelid mass lessions and are easily misdiagnosed as dermoid cysts, epidermal inclusion cysts and chalasions [9]. On gross examination, schwannomas appear as well encapsulated gelatinous or cystic masses. They usually grow very slowly and are asymptomatic.

Microscopically, they may demonstrate a biphasic pattern with areas of highly cellular (Antoni type A) and myxoid matrix (Antoni type B) [10]. The most important feature in its diagnosis remains the strong reactivity to S–100 protein by immunohistochemistry, particularly in Antoni type A areas. Despite sometimes striking cytologic atypia, mitotic figures are rare. It is postulated that degenerative changes occur due to the long period of time over which large schwannomas develop [11]. Because of their rarity and location, eyelid schwannomas have been confused with dermoid and epidermal inclusion cysts. Their management is complete excision with clear margins to establish the histopathologic diagnosis and prevent recurrence. Incomplete removal is associated with eventual recurrence and more aggressive behavior [9,12].

An attempt to preserve the continuity of the nerve should be made, but this is not always possible and does not appear to have any major consequences in this site. Histopathologically, the neoplasm appears to be well-circumscribed and composed of bundles of elongated cells with thin, attenuated nuclei. These cells tend to align themselves into compact parallel rows, with intermittent dense anucleate zones. In other locations, a poor prognosis has been described if the cells are fusiform, contain melanin granules, or if epithelioid cells are present [29]. Nevertheless, malignant transformation has not been reported in eyelid schwannomas.
This case is the first case in our Department of Ophthalmology. The age range in the adult group of published cases was between 19 and 63. The female to male proportion was 3:2. Our case was 21 years old and no association with neurofibromatosis was detected. In case, the mass was located on the nasal region of the upper left eyelid and was subcutaneous. The tumor probably arose from branches of the supraorbital nerve. Schwannomas of ophthalmic interest are rare, although they have been reported in relation to the orbit (1% of orbital tumors)[8], and infrequently to the uveal tract [13], conjunctiva [14] and sclera [15]. The occurrence of schwannoma in eyelid is extremely uncommon. Although very rare, schwannoma should be taken into consideration in the differential diagnosis of any eyelid neoplasm, especially dermoid and epidermal inclusion cysts.

In conclusion, this case suggest that schwannomas should be included in the differential diagnosis of any eyelid mass lesions and that accurate histological diagnosis and complete excision should be carried out in these cases.

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