Epibulbar schwannoma in a 12-year-old boy: A case report and review of literature

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Schwannomas are benign, encapsulated, primary neurilemmal tumors composed of proliferating Schwann cells. Schwannomas are commonly seen in the orbit, but are rare on the epibulbar surface. Herein, we report a case of a 12-year-old boy who presented to us with a slow-growing painless subconjunctival mass in the left eye. There was no intraocular extension of the mass and intra-operatively, the mass could be clearly delineated and was excised off the underlying sclera. Histopathological examination of the mass showed typical features of schwannoma and immunohistochemistry helped to confirm the diagnosis. There was no recurrence of the lesion observed at follow-up 26 months after surgery. Here, we describe this uncommon tumor and review the available literature. Although rare, an epibulbar

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schwannoma should be considered in the differential diagnosis of an amelanotic, painless subconjunctival nodular mass. Excision of the lesion is the recommended treatment.

Key words: Conjunctiva, eye, neurilemmoma, schwannoma, tumor

Schwannomas are benign, encapsulated, primary neurilemmal tumors composed of proliferating Schwann cells. The most common location for schwannomas is the head and neck region followed by the flexor surfaces of the limbs and the trunk.^[1] Schwannomas can occur in isolation or in association with neurofibromatosis type I or type II.^[2] In the ophthalmic region, schwannomas are most commonly seen in the orbit. Other reported locations include choroid, caruncle, sclera, and conjunctiva.^[3] Epibulbar schwannomas are very rare with less than ten cases being described in the literature. We report a case of a 12-year-old boy who presented to us with a slow-growing painless subconjunctival mass in the left eye. There was no intraocular extension of the mass. Intra-operatively, the mass could be clearly delineated and was excised off the underlying sclera. Histopathological examination of the mass showed typical features of schwannoma. There were no systemic features suggestive of neurofibromatosis and no subsequent recurrence of the lesion was noted. In this communication, we report this uncommon entity and review the typical clinical features of an epibulbar schwannoma. Although rare, an epibulbar schwannoma should be considered in the differential diagnosis of an amelanotic, painless subconjunctival nodular mass. Excision of the lesion is curative, and recurrence is uncommon.

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Case Report

A healthy 12-year-old boy presented with complaints of a slow growing painless conjunctival mass of 3 years' duration in the left eye. On examination, his uncorrected visual acuity in both eyes was 20/20 N6. There was a subconjunctival, nontender, well-defined, multi-lobed, immobile, firm mass with scleral fixity at 5'O-clock position of the left eye. The mass was prominent on dextroelevation [Fig. 1a and b]. There was no restriction of motility and the anterior and posterior segments in both eyes were within normal limits. Ultrasound biomicroscopy of the lesion showed uniform internal low reflectivity. Computed tomography of the orbits confirmed a well-defined mass in close relation to the globe with homogenous consistency and the extraocular muscles were



Figure 1: (a) Clinical photograph is demonstrating the nodular lesion seen as an elevation under the left lower eyelid. (b) The multilobed lesion is seen on dextroelevation. The mass is subconjunctival in location. (c) Intraoperative photograph showing the lesion, visible after incising the overlying conjunctiva. The mass was adherent to the sclera underneath but could be easily delineated and excised in-toto. (d) The excised mass was relatively avascular and firm in consistency

seen distinctly separate from the mass. Differential diagnosis based on clinical examination included atypical nodular scleritis, scleral nodule, hemangioma, parasitic cyst, or a myxoma. The patient subsequently underwent excision of the mass. Intraoperatively, the mass was found to be entirely subconjunctival and adherent to the sclera [Fig. 1c]. However, it could be separated from the underlying sclera and excised in-toto [Fig. 1d]. Double freeze-thaw cryotherapy was done at its location over the sclera, and the conjunctiva was closed with 8-0 vicryl suture. The histopathological examination of the hematoxylin-eosin stained slides showed tumor consisting of spindle cells arranged in whorls and twisted configuration, which were the Antoni-A areas as well as the irregularly arranged, paucicellular Antoni-B areas [Fig. 2a]. The cells were elongated with thin cytoplasmic processes, and the nuclei were arranged in parallel lines which assumed a typical "picket fence" pattern; features that were typical of



Figure 2: (a) Tumor consisting of spindle cells arranged in whorls and twisted configuration, which were the Antoni-A are as (H and E stain, \times 10). (b) The slender schwannoma cells showing strong immunopositivity to S100 stain (S100 stain, \times 10)

Authors/year of publication	Age/sex	Eye	Location on conjunctiva	Site of lesion	Treatment	Recurrence	Immunohistochemistry features	
Dabezies and Penner/1961	50/female	OS	Bulbar	Superotemporal	Excision	Yes	NA	
Vincent and Cleasby/1968	12/female	OD	Bulbar	Nasal (limbus)	Excision	No	NA	
Marc'hadour <i>et al.</i> /1996	37/male	OS	Bulbar	Nasal (limbus)	Excision	No	Positive for S100, negative for HMB-45	
Charles et al./1997	19/female	OS	Fornix	Inferior (fornix)	Excision	No	Positive for S100	
Charles et al./1997	26/female	OS	Bulbar	Superior (limbus)	Excision	No	Positive for S100	
Charles et al./1997	72/female	OD	Palpebral	Superior (tarsal)	Excision	No	Positive for S100	
Andreoli <i>et al.</i> /2004	68/male	OS	Bulbar	Nasal (limbus)	Excision	No	Positive for S100, negative for HMB-45	
Ohshima <i>et al.</i> /2007	10/female	OS	Palpebral	Temporal	Excision	No	Positive for S100, negative for SMA	
Demirci <i>et al.</i> /2010	17/male	OS	Bulbar	Superonasal	Excision	No	Positive for S100, negative for CD34, CD68, SMA	
Present case	12/male	OS	Bulbar	Inferior	Excision	No	Positive for S100	

The details of the solitary case described by Grossniklaus *et al.* are not available and hence the case is not included in this table.^[6] OS: Left eye, OD: Right eye, NA: Not available, SMA: Smooth muscle actin

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a benign schwannoma. The tumor cells also stained strongly positive for S100, a highly sensitive marker for melanocytes and neural crest-derived cells; thus confirming the diagnosis of epibulbar schwannoma [Fig. 2b]. The patient did not have features of neurofibromatosis. At a follow-up of 26 months, there was no recurrence of the lesion. He has been advised annual follow-up and is being monitored for clinical signs suggestive of neurofibromatosis.

Discussion

Conjunctival/epibulbar schwannoma is an uncommon tumor. In a retrospective, non-interventional case series of 1643 consecutive patients with conjunctival tumors by Shields *et al.*, there were no cases of conjunctival/epibulbar schwannoma.^[4] In a large review of 2455 conjunctival tumors by Grossniklaus *et al.*, a single case of conjunctival schwannoma was reported in a 61-year-old male.^[5]

Le Marc'hadour *et al.* described the electron microscopic features of conjunctival schwannoma in a 37-year-old male patient.^[3] The tumor almost entirely consisted of Schwann cells that also enclosed myelinated fibers and were coated by a basal lamina. In between cell bodies, there were numerous Schwann cell processes that were seen admixed with non-myelinated fibers. Also, neuritic processes were seen and were encircled by Schwann cell plasma membrane, leading to the formation of mesaxons.^[3]

Conjunctival/epibulbar schwannomas have an indolent clinical course. They usually present as a slow growing painless nodule on the conjunctiva/epibulbar surface. Based on published literature, the age at presentation varies from 10 to 72 years^[1,2,6-10] [Table 1]. There is no obvious sex predilection with 6 of the 10 cases (60%) reported being female. As elucidated by Demirci et al., since the bulbar conjunctiva is the most common site of this tumor, "epibulbar schwannoma" is probably the most apt description.^[2] Nils Antoni, a Swedish neurologist, described two distinct patterns of cellular architecture in the peripheral nerve sheath tumors. The first was what appeared to be a "fibrillary, intensely polar, elongated appearing tissue type" which was called "tissue type A." These highly cellular regions were eventually referred to as Antoni-A regions. He also described seemingly distinct, loose microcystic tissue adjacent to the Antoni-A regions, and these came to be known as Antoni-B regions.[11] These tumors eventually were referred as schwannomas. The conjunctiva receives its sensory nerve supply from the nasociliary nerve; in addition there are autonomic nerves that are subconjunctival in location. It has been hypothesized that such epibulbar schwannomas possibly arise from any of these nerves.^[10]

The treatment of epibulbar schwannomas is complete excision. While excising, one must be careful to excise it in-toto and prevent damage to the underlying sclera. Scleral involvement from the overlying lesion has been noted in two cases. Additional double freeze cryotherapy to the base, namely the underlying sclera, in addition to our case; was done in only one of the reported cases.^[2] However, it is unlikely that cryotherapy has any role in preventing recurrence, given the benign pathology of the tumor. Furthermore, no malignant transformation was noted in any of the other cases. Epibulbar or conjunctival schwannoma can be best described as a common tumor at an uncommon site. It has a benign course with excision being curative in most cases.

Literature search

A systematic Medline search was performed on PubMed using the key words: Schwannoma, neurilemmoma, conjunctiva, conjunctival, and epibulbar. There was no restriction on the date of publication. While this review relied on English articles, non-English language articles that had abstracts translated into English were also reviewed.

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Conflicts of interest

There are no conflicts of interest.

References

- Enzinger FM, Weiss SW, editors. Benign tumors of peripheral nerves. In: Soft Tissue Tumors. 3rd ed. St. Louis, Mo: Mosby – Year Book; 1995. p. 821-88.
- 2. Demirci H, Shields CL, Eagle RC Jr, Shields JA. Epibulbar schwannoma in a 17-year-old boy and review of the literature. Ophthal Plast Reconstr Surg 2010;26:48-50.
- Le Marc'hadour F, Romanet JP, Fdili A, Pèoc'h M, Pinel N. Schwannoma of the bulbar conjunctiva. Arch Ophthalmol 1996;114:1258-60.
- Shields CL, Demirci H, Karatza E, Shields JA. Clinical survey of 1643 melanocytic and nonmelanocytic conjunctival tumors. Ophthalmology 2004;111:1747-54.
- Grossniklaus HE, Green WR, Luckenbach M, Chan CC. Conjunctival lesions in adults. A clinical and histopathologic review. Cornea 1987;6:78-116.
- Dabezies OH Jr, Penner R. Neurofibroma or neurilemmoma of the bulbar conjunctiva. Arch Ophthalmol 1961;66:73-5.
- Vincent NJ, Cleasby GW. Schwannoma of the bulbar conjunctiva. Arch Ophthalmol 1968;80:641-2.
- Charles NC, Fox DM, Avendaño JA, Marroquín LS, Appleman W. Conjunctival neurilemoma. Report of 3 cases. Arch Ophthalmol 1997;115:547-9.
- Andreoli CM, Hatton M, Semple JP, Soukiasian SH, Fay AM. Perilimbal conjunctival schwannoma. Arch Ophthalmol 2004;122:388-9.
- Ohshima K, Kitada M, Yamadori I. Neurilemoma of the bulbar conjunctiva. Jpn J Ophthalmol 2007;51:68-9.
- Wippold FJ 2nd, Lubner M, Perrin RJ, Lämmle M, Perry A. Neuropathology for the neuroradiologist: Antoni A and Antoni B tissue patterns. AJNR Am J Neuroradiol 2007;28:1633-8.