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Case report

Isolated eyelid Schwannoma: A rare differential diagnosis of lid tumor

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Abstract

Primary Schwannomas of the eyelid are extremely uncommon. It accounts for one percent of orbital tumors. We present a case of isolated eyelid Schwannoma in the lateral canthus of the left eye with no systemic diseases associated. Surgical excisional biopsy was done. In two years follow up, no recurrence or malignant conversion was detected.

Keywords: Benign tumors, Eyelid, Histopathology, Schwannoma

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Introduction

Schwannoma, neurilemmoma, is a benign tumor that originates from the Schwann cells that form the neural sheath of the sensory nerves. Although schwannomas can arise from any nerve in the body, the most common areas include the nerves of head and neck, but not the ophthalmic ones. It is almost always benign in nature and while malignant forms have been documented in other areas of the body, primary schwannomas are very rarely reported in ophthalmology.[1–14] We report here an extremely uncommon lid schwannoma and its successful long term management.

Case report

A 17-year-old Saudi male medically free presented to ophthalmology clinic complaining of a painless and slowly progressing lesion of the lateral angle of the left eye for nine years. The patient did not complain of any systemic disease and was not on any medications. On ocular examination, his visual acuity (V/A) was 20/20 OU. Slit lamp examination and fundoscopy of both eyes were within normal limits. The swelling was located at the lateral angle of the left eye and it was firm, non-tender, and mobile, with a smooth surface measuring 12×7 mm, not adherent to underlying tissues or the skin. The mass did not affect the movement of the lid and did not cause ptosis. There was no neurological complaint associated with the lid lesion. The lesion was completely excised under local anesthesia through upper lid crease approach. The lesion was well-defined, capsulated, easily dissected and sent to histopathology lab in our hospital. Postoperatively, the patient was completely free of symptoms for two years with an excellent wound healing.

Histopathologically, the lesion has a well-defined capsule and consists of a polypoidal cyst measuring 1.0×0.6 cm. The microscopic examination revealed highly cellular areas of non-pigmented spindle cells containing elongated nuclei. Alternation of solid cellular areas (Antoni A pattern) with areas of looser myxoid tissues having ovoid cells suspended in a mucinous background (Antoni B pattern) (Fig. 1). No mitotic figures were seen. Blood vessels were

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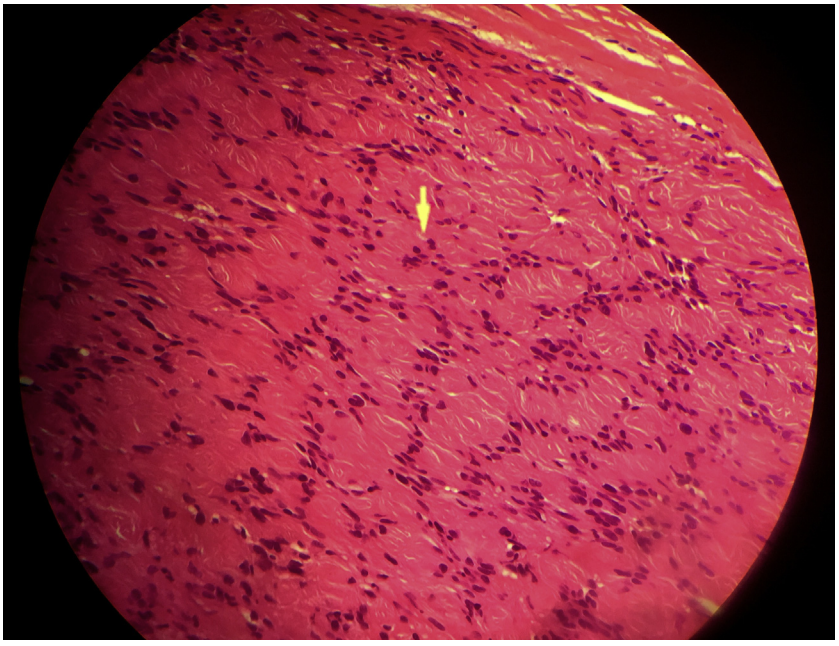

Fig. 1. Microscopic view showing fusiform cells arranged in intertwined bundles along with alternating Antoni A and Antoni B patterns (magnification 100).

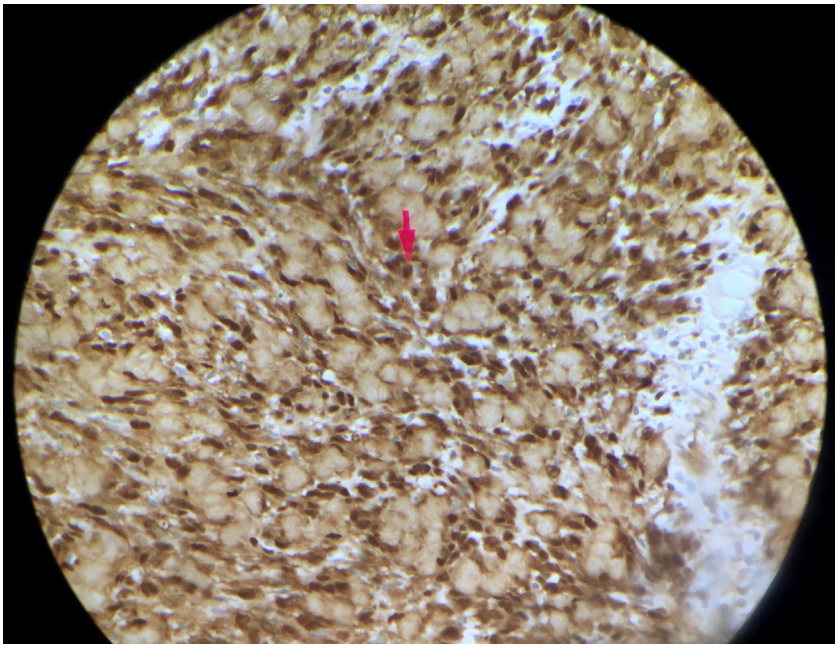

Fig. 2. Microscopic view showing positive nuclear reactivity to S100 protein (magnification 100).

prominent. S100 immunization showed diffuse strong nuclear activity (Fig. 2).

Discussion

Schwannoma (Neurilemmoma) is a tumor that originates from the proliferation of Schwann cells of peripheral nerve sheaths. It is a neoplasm which occurs wherever Schwann cells are present in any myelinated peripheral nerves.[1,9] In most cases, schwannoma usually manifests as a single neoplasm. The presence of multiple schwannomas is usually indicative of neurofibromatosis 1 or 2. Our case was an isolated lesion with no systemic disease.

Schwannoma arises from the sensory nerves of the orbit, including the infraorbital nerve. Schwannoma, in the eyelid, is very rare and just a few cases were published.[1–14] It was first reported in 2007 by Lopez-Tizon et al. and represents only 0.1–0.7% of the eyelid neoplasm.[1,9] The age range in the reported cases were between 19 and 63 while our patient is 17-year-old with an early appearance of the mass. The size

of the tumor in our case was 12 × 7 mm which matches with Sudhir et al. who reported that the size of the tumor ranges from few millimetres to 3.5 cm.[15]

Clinically, the lesion in our case was firm, slowly progressing and non-tender. There was no ptosis in our case which goes with previous reports.[1–3,5–14] Some cases reported ptosis as the presenting symptom.[4,15] Only one case associated with an ulceration have been reported.[4] What is unique in our case, is the presence of the lesion in the lateral canthus which has not been reported in the literature. Previous reports had showed the tumor involving upper or lower lid of either eyes.[1–15] In differential diagnosis we have dermoid cyst, inclusion cyst and chalazion. Only with excisional biopsy we could get the final diagnosis. Histopathologically, schwannoma is a capsulated lesion and classically shows a mixture of two patterns, the Antoni A (dense cellular pattern) and the Antoni B (edematous disorganized pattern), but the most important feature for diagnosis is the strong reactivity to S100 protein in immunochemistry and rare mitotic figures.[1–14] Poor prognosis has been described in some cases if the cells are fusiform, contain melanin granules, or if epithelioid cells are present.[5] No features of malignancy were seen in our patient. Nevertheless, malignant transformation has not been reported in eyelid schwannoma.[5]

Management of schwannoma of the eyelid is complete excision with clear margins to establish the histopathological diagnosis and to prevent recurrence, as we decided to do in our case. Incomplete excision is associated with recurrence and more aggressive behavior. There have been reports of malignant changes in a previously incomplete removal of benign schwannoma[5].

In conclusion, isolated eyelid schwannoma is extremely rare and the presence of the lesion in the lateral canthus of a young patient is not reported in the literature. To the best of our knowledge, this is the first case to be reported in Saudi Arabia. It is highly recommended to be considered in the differential diagnosis of eyelid tumor. Accurate histopathological diagnosis and early complete excision of the tumor should be the objective in the management of eyelid schwannomas.

Conflict of interest

The authors declared that there is no conflict of interest.

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