Case Report

Giant Schwannoma of upper eye lid in a male child of 13 years – A rare case report

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Abstract

Isolated Schwannoma of the eyelid is an extremely rare benign neoplasm. Although it is believed to be present from birth, it show accelerated growth during the childhood or later in adults. These are generally asymptomatic or present as slowly enlarging, painless mass with progressive ptosis of the eyelid. I hereby report a case of giant schwannoma of size 3cmx3cmx2cm in the left upper eyelid of a 13 year old boy who developed progressive ptosis.

Key words

Schwannoma, Benign Eyelid tumors, Histopathology.

Introduction

Schwannoma is a benign peripheral nerve sheath tumour composed of a proliferation of schwann cells [1]. It is a slow growing encapsulated tumour that seldom affects ocular tissues. These are asymptomatic [2]. Schwammoma can be associated with neurofibromatosis but when isolated they not related to this condition [2]. In ophthalmic region, it has been reported in relation to orbit and infrequently to conjuntiva, uveal tract, and sclera [1, 3, 4]. Eyelid schwannomas are extremely rare. Till date less than 20 cases have been reported so far.

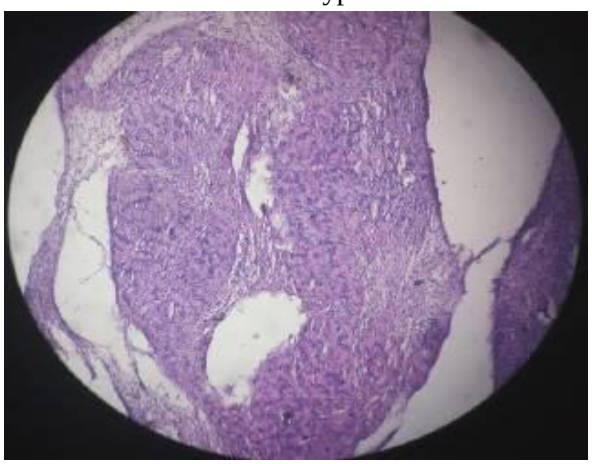
Case report

A male child of 13 years of came with the complaint of slowly progressive painless mass in his left upper eyelid for the last one year. He had developed ptosis of the left eye lid. On examination, there was a firm non-tender nodule of size 3x3x2 cm on the left upper eyelid. The growth was not adherent to the skin and was mobile on palpation. There were no clinical features of neurofibromatosis. An excision biopsy of the mass was performed under local anesthesia. A well-defined encapsulated non-pigmented tumor was completely dissected out and lid was reconstructed and sent for

histopathology. It was not attached to any nerve. Gross specimen of the tumor was pearly white. Cut section shown no haemorrhage or necrosis. It was 3x3x2cm and firm in consistency (**Figure – 1**). Histopathological section of the tumor showed compact groups of spindle shaped or fusiform cells arranged around ovoid mass of strongly eosinophilic cytoplasm called as Verocay bodies and tend to arrange them in palisades called as Antoni A areas which predominate the slide. Few areas were of Antoni B type which are Schwann cells arranged haphazardly in very loose vacuolated reticular stromal tissue. No histopathological features of malignancy were present (**Figure – 2**).

Figure – 1: Gross specimen of eye lid schwannoma. Cut section shows no hemorrhage or necrosis.

Figure – 2: Histological section of schwannoma at 10 X. Antoni A areas are highly cellular showing verocay bodies and Antoni B areas which is very loose vacuolated myxomatous stromal tissues which are hypocellular.



Discussion

Schwannoma is a bengin neoplasm which originates from the schwann cells of peripheral nerves. Isolated schwannoma of eyelid is an extremely rare benign tumor [1] multiple schwannoma usually indicate neurofibromatosis - 2. Our patient had isolated eyelid schwannoma with no family history or clinical features of neurofibromatosis 1 or 2. It is believed to be present from birth but show accelerated growth during childhood and later on in adult life [5]. When associated with neurofibromatosis, there is plexiform tumour of lid with ptosis, thickened corneal nerves, pulsating proptosis, glioma of the optic nerve and congenital glaucoma [6]. Schwannoma can be associated with neurofibromatosis but when isolated they are not related to this condition [2]. Generally, these are asymptomatic [2]. It is usually seen in adults but only 3 cases have been reported in children [7, 8]. Schwannoma of eyelid originate from the branches of the supraorbital supratrochlear and infraorbital nerves [8]. It manifest clinically as solid, painless, slow growing masses. The various types of schwannomas are conventional, cellular and melanotic schwannoma [9]. In eye, it is reported in relation to orbit and infrequently to conjunctiva [1, 3], uveal tract selera [4]. Grossly it is well circumscribed, on cut section white to yellow and partially surrounded cut section white to yellow and partially surrounded by skin. On microscopy it shows 2 types of areas as Antoni A areas which show compact groups of spindle shaped cells with spindle shaped basophilic nuclei surrounding an ovoid mass of strongly eosinophilic cytoplasm called as Verocay bodies and tend to arrange them in palisades. Antoni B areas which show schwann cells arranged haphazardly in very loose vacuolated reticular stromal tissue and very little collagen. Hyalinization of the blood vessels is a prominent feature [10]. Immunohistochemically it shows strong reactivity for S-100 protein. The differential diagnosis of isolated schwannoma of eyelid are chalazion, sweat gland tumor, sebaceous gland carcinoma, hair follicle tumor and other neurogenic and soft tissue tumors [11].

Its management is done by complete excision with clear margins to establish the histopathological diagnosis and prevent recurrence. Incomplete removal is associated with recurrence and aggressive behavior [12].

Conclusion

Isolated schwannoma of eyelid are although rare it should always be considered as differential diagnosis of benign eyelid tumors. When associated with neurofibromatosis it causes marked asymmetry of the face due to diffuse hypertrophy and pendulousness of the facial tissues on one side. Its diagnosis is suggested by clinical examination, imaging studies, and its macroscopic appearance. The diagnosis is confirmed by histopathology and immunohistochemistry which shown strong reactvity to S-100 protein predominantly is Antoni A areas.

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