

Intraorbital Tumours, Schwannoma, Neurofibroma

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INTRODUCTION

Schwannomas are rare benign tumours arising from neuroectodermal Schwann cells. They can arise from cranial, intraspinal, peripheral and autonomic nerve sheaths. Schwannomas represent 1 - 8 % of head and neck tumours. They constitute 25 – 40 % of extracranial tumours in head and neck region.¹ Among the schwannomas arising in head and neck region those arising intraorbitally are very rare. The most common benign intra orbital tumours are haemangiomas. They can arise as localised forms or in association with Neurofibromatosis-1 (NF-1). In patients with neurofibromatosis-type 1 or in patients with family history of NF, the risk of developing orbital schwannoma is 1.5 %. Schwannomas constitute 1 - 6.5 % of intra orbital tumours.²⁻⁶ Of these tumours those undergoing cystic changes are still rare. The most common cystic lesions in the orbit are dermoid cysts or mucoceles.

PRESENTATION OF CASE

A 62-year-old lady presented with a painless swelling which was insidious in onset and was gradually progressing over a period of 10 years and had reached the present size. She was having diplopia. On examination, there was 6 x 5 cms smooth swelling located supraocularly with eyeball pushed down and laterally. Pupillary size was normal and reacting to light normally. Eye ball movement was mildly restricted superiorly. There was no loss of sensation around the orbit or there were no other skin lesions else were in the body. There was no cervical lymphadenopathy or conjunctival congestion. Magnetic Resonance Imaging (MRI) was performed which showed isointense lesion in T1 weighted image and T2-weighted image showed a hyper intense lesion. Patient was taken up for surgery, with patient in supine position, a supraorbital incision was placed and flap was dissected. The lesion being cystic and extraconal in location, it was carefully dissected from the surrounding structures and tumour excised en masse. Haemostasis was achieved and wound closed in layers.

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CLINICAL DIAGNOSIS

With the long standing history and location of tumour, a benign tumour was suspected. The history did not show any change in visual acuity, ruling out lesions arising from optic nerve or intraocular tumours. With restriction of movement of the eyeball we thought in favour of extra ocular lesion occupying space and restricting movements of eye ball. As the lesion was cystic we thought of dermoid cyst or cystic nerve sheath tumour. But dermoid cyst usually occurs in middle age and haemangiomas usually occur in early 1st and 2nd decades.

DIFFERENTIAL DIAGNOSIS

The various differential diagnoses sought were a dermoid cyst, schwannoma, neurofibroma, vascular lesions and other tumours. Various vascular lesions include AVM (Arterio-Venous Malformations), cavernous sinus thrombosis, orbital venous varix, etc. Though neurofibroma was thought for differential diagnosis; with no existing lesions elsewhere in the body and no skin lesions it was thought as a rare possibility. The various tumours include epithelial neoplasms of lacrimal apparatus, benign and malignant connective tissue tumours and other metastatic tumours of orbit. Dermatofibroma protuberans is a soft tissue tumour usually less circumscribed and occurs in 2nd decade. Magnetic resonance imaging helps to identify anatomical location of tumour, extent of the tumour, vascularity of the lesion. Schwannomas in MR are smooth lobulated lesions with long axis in anteroposterior direction. They may be cystic and usually enhance with contrast.

PATHOLOGICAL DISCUSSION

A classical schwannoma is a well encapsulated lesion. It contains various areas with various densities. There are Antoni A (more cellular areas) and Antoni B (loose, less cellular areas) with loose oedematous and mucin containing stroma with fibrillar collagen. Typical Verocay bodies may be seen and atypical cells are rare. They are positive for S 100 in immunohistochemistry. A neurofibroma is an unencapsulated lesion which lacks Antoni A and B pattern. A peri neuroma is EMA (Endo-Mysial Antibodies) positive and S100 negative and dermatofibroma protuberans are CD34 positive and S 100 negative.

DISCUSSION OF MANAGEMENT

She was planned for surgical excision with a supraorbital approach. The lesion was found arising from supraorbital or supratrochlear nerve which was excised successfully. Postoperative recovery was uneventful and there was sensory loss in supraorbital area.

Schwannomas are benign tumours arising from hyperplasia of the myelin producing Schwann cells. They are

rare tumours contributing to 1 - 6.5 % of orbital tumours. They usually develop in 2nd to 5th decade and are very rare in children. Von Recklinghausen neurofibromatosis has been reported to be present in 2 – 18 % of cases.⁷ They usually present with insidious onset, gradual progressing non-pulsating lesions with proptosis or swelling. It is a non-invasive tumour without invading other structures. With increasing swelling they may develop diplopia, paraesthesia or loss of sensation in the region of distribution of the nerve, pain and swelling of eyelid. Most commonly it originates from superior orbit nearly in 61.9 % cases.⁶ In long standing cases bone changes may occur due to pressure exerted by the mass. Cystic degeneration can occur in 32 – 41 % of cases depending on various studies.^{3,7,8} There are no definite imaging criteria to differentiate schwannomas from neurofibromas. Various sonographic findings like lobulated appearance, cystic portion, hypovascularity, maximum to minimum diameter ratio, nerve tumour position are used to differentiate schwannomas from neurofibromas but none can definitely differentiate them.⁸

Surgical excision is the main stay of therapy for orbital schwannoma or neurofibroma. Various surgical approaches anterior, lateral, combined lateral orbitotomy or frontal craniotomy are described depending on the location of the tumour in the orbit. But the definitive diagnosis will be made only after histopathology. Schwannomas show two distinct patterns on histopathology. Antoni A which consists of well differentiated spindle cells and Antoni B which shows bipolar or multipolar cells interspersed in myxoid matrix.

FINAL DIAGNOSIS



Figure 1. MRI Images Showing Superiorly Located Intraorbital Lesion in Right Eye, Post-Surgical Specimen, Pre- and Post-Operative Images

Intra orbital cystic schwannoma is a very rare tumour with surgery being the main modality of treatment. Surgery leads to cure. After excision, the globe returns to normal position and normal visual acuity is regained. Few patients may develop loss of sensation over the area of distribution

of nerve. Here we describe a rare case of intraorbital cystic schwannoma.

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