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A Rare Case of Optic Nerve Schwannoma in Neurofibromatosis

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ABSTRACT

A highly unusual patient of neurofibromatosis with optic nerve schwannoma is reported. Orbital mass with visual loss were presenting features. Complete excision of mass with debulking was done. Diagnosis of optic nerve schwannoma was confirmed by histopathological examination. Optic nerve glioma and meningioma are common in neurofibromatosis while optic nerve schwannoma rarely occurs in this disease. This patient enlarges the spectrum of clinical presentations that can be encountered at young age in patients with neurofibromatosis.

Key words: *neurofibromatosis, schwannoma, optic nerve , histopathology*

INTRODUCTION:

Neurofibromatosis is a genetically transmitted disease which affects all neural crest cells [schwann cells, melanocytes, endoneural fibroblasts][1]. Cellular elements from these cell types proliferate excessively throughout the body forming tumors[2]. Schwannoma are benign tumors that arise from schwann cells in the peripheral nervous system. The vestibular division of the 8th cranial nerve is the most commonly affected, followed by the trigeminal nerve root[3]. Although schwannoma of other cranial nerves are very rare, these nerves do

have myelinated sheaths composed of schwann cells and are thus potential sites for intracranial schwannoma. However optic nerve proposed to be an exception to this rule, as there are no schwann cells in optic nerve .As optic nerve myelin is produced by oligodendrocytes rather than Schwann cells, these tumors probably arise from the Schwann cells that accompany the sympathetic nerves that are tightly adherent to the optic nerve sheath[4]. A rare case of optic nerve schwannoma in neurofibromatosis diagnosed on histopathology is being described Case report-A 15 year's old male presented with swelling of right eye since 3 years. According to the patient he noticed gradually progressing bulge in right eye over one year. This was associated with dimension of vision .In next six months patient lost his vision completely for which he consulted an ophthalmologist. As per records his right eye was enucleated. However he had no histopathology reports. Few months after surgery he again noticed swelling of right upper lid for which he consulted us. Patient did not give any history of pain, weight loss, hearing loss, tinnitus, vertigo and headache. On general examination we noticed bilateral posterior cervical lymph nodes enlargement [1.5 cm size, firm, and smooth surface, mobile]. Multiple café-au-lait spots > 1.5 cm size were present over chest, abdomen and back of neck (Fig 1). On ocular examination the socket of right side showed a mass underneath the upper lid and it was adherent to lid causing bulge and enlargement of the lid (Fig 2). Mass was approximately 6x5 cms in size, globular in shape occupying the whole socket .Overlying skin was smooth with visible dilated veins .The swelling was not expansile with no secondary changes or visible pulsations. On palpation it was firm ,non tender ,non pulsatile, non reducible, adherent to lids, with negative cough impulse and the temperature of swelling was not raised,. No bruit was heard on auscultation. Conjunctiva was normal with shallow fornices. Left eye showed 8 lisch nodules over iris from 2-4 o'clock & 8-9 o'clock (Fig 3).

Fig 1

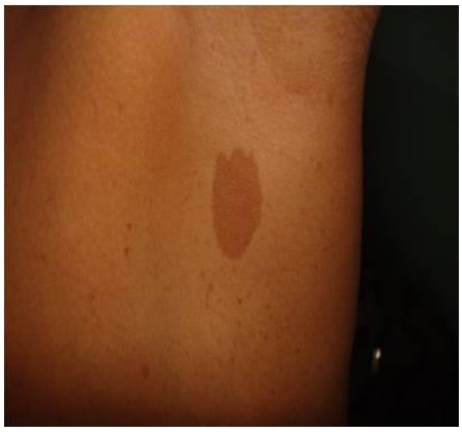
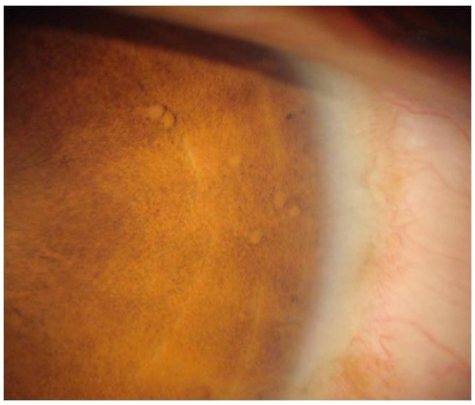


Fig 2

Fig 3



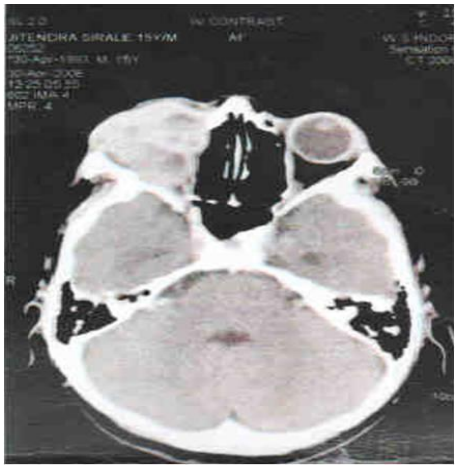
Investigations

All routine investigations were within normal limit.

CT scan Orbits

A large heterogeneous enhancing hyper vascular extraconal soft tissue lesion in superior part 4.3(AP)x 3.9(RL)x4(SI) cms, almost occupying entire orbit. Superior rectus inseparable from lesion. Optic nerve & other recti are severely compressed & displaced inferiorly with mild expansion of bony orbit (Fig 4).

Fig 4



MRI-

A large lobulated heterogeneously enhancing mass lesion in right orbit 4.9x4.5x3.7 cms size with intraregional haemorrhagic & necrotic areas limited to orbit. Multiple air fluid levels seen in lesion. No intracranial or intrusions extension. Features s/o recurrent neurofibromatosis in right orbit (Fig 5 & 6)

Fig 5

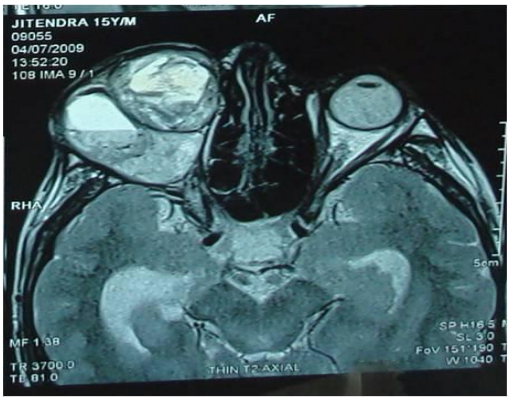
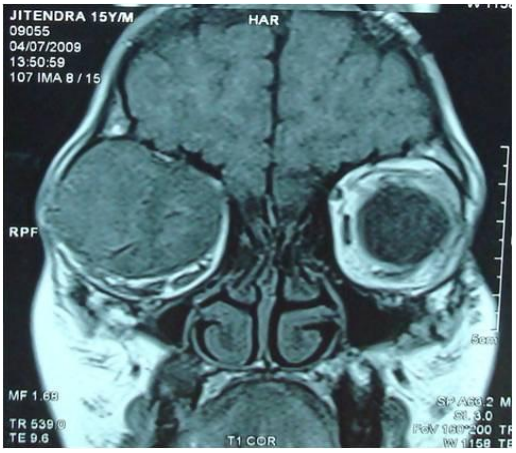


Fig 6



TREATMENT

Complete excision of mass with debunking of adjacent tissue was done (Fig 7) Excised tissue sent for histopathological examination. Histopathological Examination showed neural

tissue with areas of hypercellularity and hypocellularity. The hypercellular areas showed spindle shaped cells with elongated oval to spindloid basophillic nuclei. The cells were arranged at the periphery around eosinophillic mass. The hypocellular area showed schwann cells dispersed in between loose vacuolated reticular tissue.Adjoining areas showed fibrocollagenous tissue with congested capillaries. These features were suggestive of schwannoma (Fig 8).

Fig 7

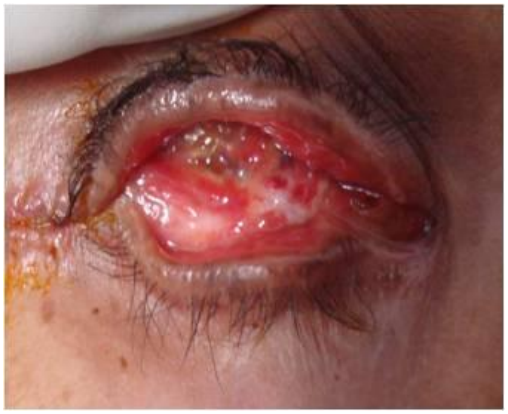
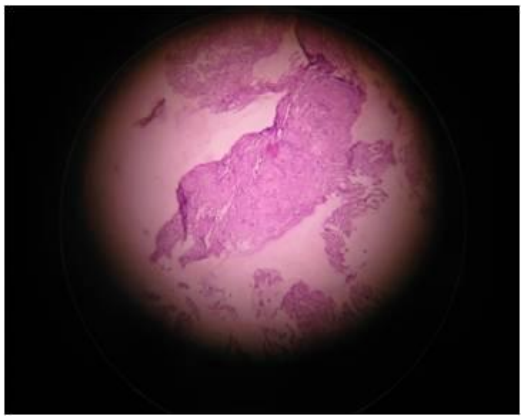


Fig 8



DISCUSSION

The schwannoma may be associated with neurofibromatosis type 1 and neurofibromatosis type 2[5]. Benign tumours arise from Schwann cells in the peripheral nervous system. Although the most common sites are the vestibular division of 8 cranial nerve and trigeminal nerve root, Schwannoma occasionally involve the optic nerve[6]. The microscopic appearance of schwannoma of optic nerve is same as all schwannoma of PNS origin i,e presence of antoni type A tissue and antoni type B tissue. Thus distinguishing them from similar appearing lesion such as optic nerve gliomas or meningiomas, However clinical presentation of these lesion is non specific[7].They have been reported in both children and adult's .They typically follow a benign course and produce symptoms via mass effect. The tumour typically present as slowly enlarging tumours resulting in proptosis, extra ocular muscle dysfunction,occasionally pain and progressive visual loss associated with optic neuropathy[8].Optic nerve sheath schwannomas probably can't be diagnosed on clinical grounds alone, and their neuroimaging appearance mimics that of the more common optic nerve glioma[9].Instead,in reported cases ,the diagnosis has been made at surgery same as with our case. To date surgery has been the treatment of choice, but in view of increasing tendency to treat many intracranial schwannomas with stereotactic radio surgery, this treatment option perhaps should be considered for schwannomas of the optic nerve.

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