Recurrent neurofibroma of the orbit

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CASE REPORT

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Abstract

A 55-year-old male patient presented with gradual progressive outward and downward deviation of right eye since last two years, with history of a similar complaint 10 years ago when he was diagnosed as having neurofibroma of the orbit. Computed Tomography imaging revealed a large, multilobulated, heterogeneous, soft tissue density mass lesion in the retro bulbar region on the medial side of right orbit suggestive of a neurofibroma. Excision and histopathology confirmed it to be a recurrence of neurofibroma of the orbit.

Key Words
Recurrent, neurofibroma, orbit

Implications for Practice

1. **What is known about such cases?** Recurrent solitary neurofibromas of orbit are very rare with very few cases reported in literature.
2. **What are the implications for future practice?** Neurofibromas should be considered in the differential diagnosis of orbital tumours.
3. **Follow-up?** Long-term follow-up of surgically treated cases of neurofibroma of orbit should be done for early detection of recurrence.

Background

Neurofibromas are tumours consisting of a proliferation of peripheral nerves that can affect almost any organ, soft tissue, bone or anatomical site including, quite commonly, the skin. The systemic manifestation of this disease is called neurofibromatosis, which is a widespread and variable disease. This abnormal growth of the peripheral nerve Schwann cells, endoneural fibroblasts and collagen can lead to a distortion of the bony anatomy in the eye socket and soft tissue of the eyelids. Localised neurofibromas of the orbit are relatively uncommon. The symptoms and signs of localised orbital neurofibromas depend on their location in the orbit. Their clinical presentation is similar to the orbital schwannomas and present as a localised orbital mass with slow progressing painless or mildly painful proptosis in young to middle aged adults. We describe an unusual case of recurrent localised orbital neurofibroma arising in superomedial orbit, diagnosed by histopathology.

Case details

A 55-year-old male patient presented with gradual progressive outward and downward deviation of right eye since last two years. It was painless initially but had become painful since the last three months (Figure 1). There was no history of trauma or complaint of diplopia. He had a similar complaint around 10 years ago, for which he had undergone medial transconjunctival orbitotomy and the excised mass was histopathologically diagnosed as neurofibroma.

Figure 1: Outward and downward deviation of right eye

On examination, visual acuity in the right eye was 2/60. A solid slightly tender mass in medial side of the orbit was seen in the right eye; it became more prominent on applying pressure on the lower part of the globe. The size of the mass did not vary with valsalva manoeuvre, ocular movement or posture. The mass was non-pulsatile and

trans-illumination was negative. There was downward and outward globe proptosis of about 18 mm (Figure 2) and ocular motility was restricted in superior, superomedial and medial direction. Intra ocular pressure was 14 mm of Hg in both eyes. Corneal sensations were intact. All the cranial nerves were normal and neurological examination were within normal limits. Slit lamp and fundus examination revealed normal anterior and posterior segments. The left eye examination and general physical examination were within normal limits. Computed Tomography imaging revealed a large 4.9 cms x 2 cms sized, multilobulated, heterogeneous, slightly enhancing, mildly hyper-dense, soft tissue density mass lesion in the retro bulbar region on the medial side of right orbit, suggestive of a neurofibroma (Figure 3).

Figure 2: Proptosis of right eye

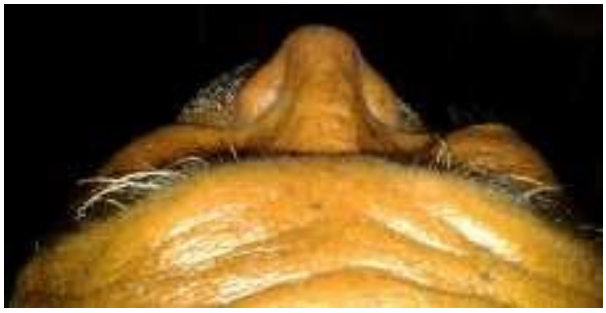
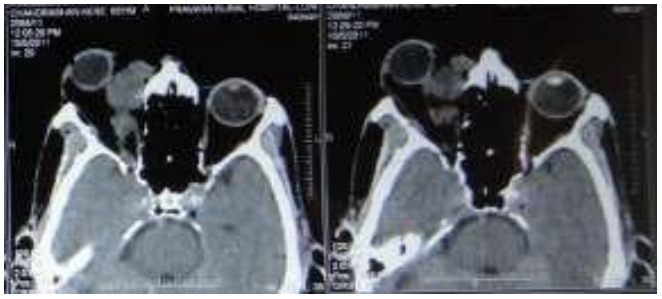


Figure 3: Computed tomography soft tissue density mass lesion in peribulbar region on medial side of right orbit suggestive of a neurofibroma

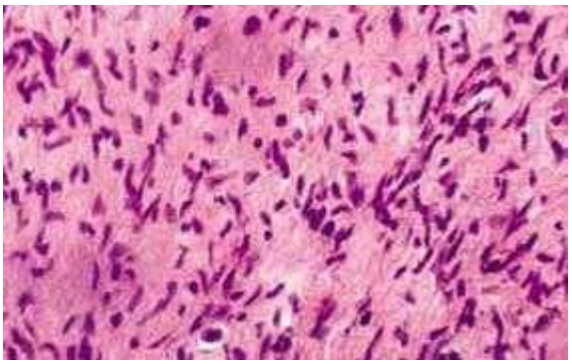


The lesion was excised using an anterior superior transseptal orbitotomy approach. Special care was taken to preserve the muscles and the globe. The mass was removed in 3 parts and a drain was left in situ after the surgery (Figure 4). Patient had a smooth post-operative recovery, regional sensory function was normal and no neuro-ophthalmic sequelae were present. On histopathological examination, the specimen consisted of a three nodular, whitish grey, solid mass measuring 2cm each in the greatest diameter of pure gum rubber consistency (Figure 5). Its cut surface was homogeneous, smooth and grey-white. Microscopically, it was diagnosed as a case of a localised neurofibroma (Figure 6).

Figure 4: Post-operative day 1 showing drain in place

Figure 5: Excised nodular masses of around 2cm each, of rubbery consistency

Figure 6: Histology suggestive of neurofibroma (HE, x20)



Discussion

Neurofibroma is a benign, peripheral nerve sheath tumour. Peripheral nerve sheath tumours are composed of variable combinations of Schwann cells, perineural cells, and fibroblasts. They account for approximately 4% of orbital tumours and comprise plexiform neurofibromas (2%), localised neurofibromas (1%), and neurilemmomas (1%).[1] Neurofibromas of the orbit are rare and account for 0.6 - 2.4% of all orbital tumours.[2,3] Neurofibromas of the orbit may be of three subtypes, namely, plexiform, diffuse and localised neurofibromas. Plexiform neurofibromas, the most common orbital subtype, occur exclusively in neurofibromatosis type 1, become manifest during the first decade of life, and diffusely infiltrate the eyelid and orbital

soft tissue.[1] Diffuse neurofibromas are usually the dermal variants; they rarely affect the orbit and are clinically indistinguishable from the plexiform subtype. Histologically, diffuse neurofibromas show greater cellularity, less collagen deposition, and lack the cellular perineural sheathing characteristic of plexiform neurofibromas. Both the plexiform and diffuse subtypes lack clear margination and tend to be highly vascular.[1,4] These neoplasms do not respond to medical therapy, and subtotal resection and haemorrhage frequently complicate their surgical management. Localised neurofibromas of the orbit are uncommon. Although they may arise from either sensory or motor nerves, branches of the frontal nerve are most commonly affected. Presenting typically in the second to fifth decades of life as slowly progressive, orbital soft tissue masses, they induce axial and nonaxial globe dystopia.[1,5,6,7] Visual acuity impairment is typically minimal. Ocular dysmotility secondary to the mass effect may exist.[5,6,7] Localised neurofibroma is relatively well circumscribed and much less vascular, unlike plexiform and diffuse neurofibroma which are more vascular and diffusely involve the orbital tissues.[2,5,8] The clinical presentation of localised neurofibroma usually depends on the origin and location of the tumour in the orbit. The most common site of localised neurofibromas is the superior orbit but they may also be present in the inferior orbit.[8,9]

Recurrence and re-growth is common with plexiform orbital neurofibroma but not with solitary localised neurofibroma. Recurrence may be due to missing of a very small tumour in the excision or transaction of involved nerve during surgery producing an amputation tumour. In our case, there was recurrence after about 10 years of excision of primary tumour. This case highlights the fact that neurofibromas should be considered in the differential diagnosis of orbital tumours. Complete, meticulous surgical excision preserving the ocular nerves is the treatment of choice in neurofibromas. Also, long term follow up is necessary for early detection of possible recurrence.

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PEER REVIEW
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CONFLICTS OF INTEREST
The authors declare that they have no competing interests.

PATIENT CONSENT
The authors, Somen Misra, Pratik Gogri, Neeta Misra and Akshay Bhandari declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient in this report.
2. All possible steps have been taken to safeguard the identity of the patient.
3. This submission is compliant with the requirements of local research ethics committees.