Benign Orbital Neurilemmoma - An Uncommon Cause of Unilateral Proptosis

Bangal SV*, Bhandari AJ**, Badhe KP***, Bankar MS****

Abstract

Neurilemmomas are benign, localized, encapsulated tumors arising from the Schwann's cells of the peripheral, cranial and sympathetic nerves as well as from spinal nerve roots. Neurilemmoma occurs at any age, more commonly seen in females than males. It is a rare orbital tumor comprising only 1.1% of all orbital tumors. 70 year old female presented with protrusion of left eye since 9 years which gradually increased in size accompanied by mass over the lateral aspect of left eye since 9 years. On examination non-axial type of proptosis of 23 mm was found in left eye. MR imaging showed 3.25x3.04x3.23 cm enhancing mass with intra as well as extra-conal component on superolateral part of the left orbit with no intracranial extension. The orbital mass was removed in toto under general anesthesia via lateral orbitotomy approach. Histopathological examination revealed spindle shaped Schwann cells with elongated nuclei showing characteristic palisading, suggestive of neurilemmoma.

Keywords: *Neurilemmomas, Proptosis, Lateral orbitotomy*

Introduction

The occurrence of neurilemmoma in general is not infrequent, but it is a relatively uncommon orbital neoplasm. Neurilemmomas are benign, localized, encapsulated tumors arising from the Schwann's cells of the peripheral, cranial and sympathetic nerves as well as from spinal nerve roots.[1,2,3] They involve auditory nerve, trigeminal nerve and also the ciliary nerves of the orbit.[3,4] Rarely, orbital bones are also involved.[3] Neurilemmoma occurs at any age, more commonly seen in females than males.[5] It is a rare orbital tumor comprising only 1.1% of all orbital tumors.[6] Neurilemmoma of the orbit grows very slowly and it causes, exophthalmos, restriction of ocular movements and blurring of vision. Neurilemmoma are commonly benign and less than 1% becomes malignant, degenerating into a form of cancer known as neurofibrosarcoma.

*Professor and Head, **Asst. Professor, ***Professor,
**** Postgraduate Student, Department of Ophthalmology

Corresponding Author
Dr. Akshay J. Bhandari,
Department of Ophthalmology, Rural Medical College, Pravara Institute of Medical Sciences, Loni, Maharashtra.
E-mail: kiwis143@gmail.com

Case report

A 70 years old female presented with gradual protrusion of left eye since 9 years, which gradually increased in size accompanied by mass over the lateral aspect of left eye since 9 years. There was no history of systemic disease or trauma. On general examination, patient was averagely built and well nourished. Systemic examination was normal.

Right eye examination was within normal limits except for the presence of immature senile cataract. In left eye ocular movements were normal except in elevation. Best corrected visual acuity in right eye was 6/12 and left eye it was 6/24. In Left eye, mild conjunctival congestion was noticed with normal pupillary light reactions. Lens showed grade II nuclear sclerosis with cortical cataract, rest of the anterior segment findings of left eye was normal. Colour vision of both the eyes was normal. On examination non-axial type of proptosis of 23 mm was found in left eye. Fundus examination of both eyes were normal. Local examination of left eye, revealed a firm, non-tender, non-compressible, non pulsatile mass on superolateral aspect of left eyeball. (Fig. 1) Posterior extent of mass was not accessible on clinical examination. The mass was free from the surrounding structures.

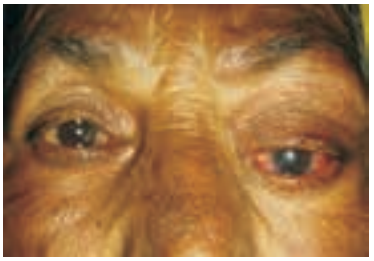

Figure 1: Nonaxial proptosis in left eye

Laboratory investigations and X-ray orbit were normal. MR imaging showed 3.25x3.04x3.23 cm enhancing mass with intra as well as extra conal component on superolateral part of the left orbit with no intracranial extension. (Fig. 2) The central part revealed intermediate signal intensity on T1- and T2-weighted images, having signal characteristics similar to those of gray matter. The periphery of the tumor appeared as low signal intensity on the T1-weighted image and hyper intensity on the T2-weighted image. After intravenous administration of gadopentetate dimeglumine, the central part of the tumor showed clear, uniform contrast enhancement, whereas the periphery of the tumor showed no enhancement.

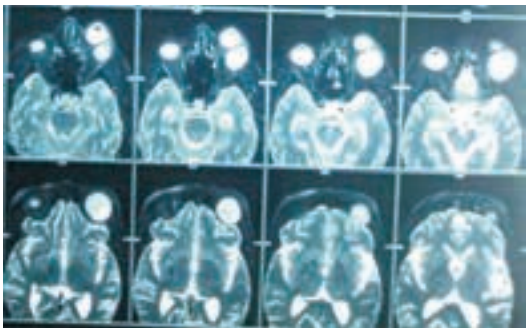

Figure 2: MRI showing tumour mass

The orbital mass was removed in toto under general anesthesia via lateral orbitotomy approach. Histopathological examination revealed spindle shaped Schwann cells with elongated nuclei showing characteristic palisading. Between the cells there was an abundance of long, slender, straight or serpentine reticulum fibres of Antoni type A, suggestive of neurilemmoma. (Fig. 3)

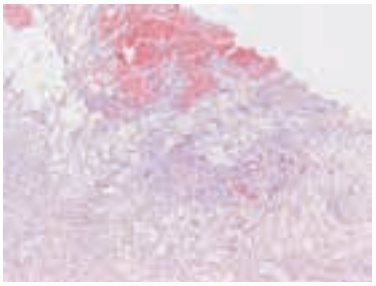

Figure 3: Schwann cells with palisading of nuclei in type A pattern of neurilemmoma

Postoperatively, visual acuity of patient was maintained, proptosis disappeared and extraocular movements became normal even in elevation. Patient was followed up to 6 months and there was no recurrence of mass. (Fig. 4)

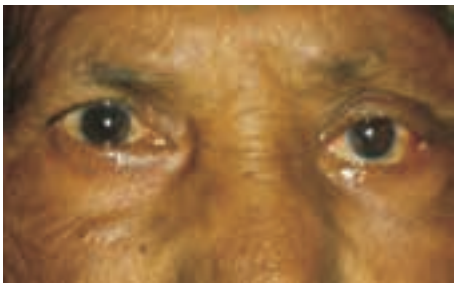

Figure 4: Proptosis disappeared postoperatively

Discussion

Neurilemmoma is a term introduced by Stout in 1935 for tumors consisting of proliferation of Schwann cells within the sheath of a nerve.[7] It has also been termed schwannoma by Masson in 1932, perineural fibroblastoma by Penfield in 1927 and neurinoma by Verocay in 1910.[8] Neurilemmoma, also known as Neurolemmoma, Schwann-cell tumour or Schwannoma, is a benign encapsulated tumour arising from the neuro-ectodermal sheath of Schwann and occurs in subjects of all ages without predilection of any ethnic group.[9] Neurilemmoma has got no distinct clinical feature. So the diagnosis depends solely on the characteristic histological findings. It may result from a family history of neurofibromatosis, a genetic disorder. The presentation is usually seen between 2- 70 years of age seen mainly in females.[10] However, a small proportion of cases are associated with von Recklinghausen's neurofibromatosis. The tumour can arise from any peripheral, autonomic or cranial nerves, with the exception of the optic and olfactory nerves, since these do not have Schwann cells.[9] For this reason, it is uncommonly seen in the orbit. Henderson and Reese estimated the incidence of orbital neurilemmomas to be approximately 1.5% of orbital neoplasm.

Most orbital neurilemmomas are believed to arise from the oculomotor nerve because it is widely distributed in the orbit. Sensory changes are absent, as would be expected if the trigeminal nerve is involved. In the orbit, as in most other sites, it may be a round, ovoid, fusiform or occasionally bosselated growth. The cut surface is usually pinkish grey and may show irregular yellow areas and cysts, surrounded by a fibrous capsule. The tumour tissue characteristically exhibits two contrasting tissue

patterns, designated as Antoni type A and B. The type-A pattern is compact with its spindle cells arranged in wavy, flowing, interlacing cords or whorled bundles. The long oval nuclei are characteristically orientated as in palisades (the nuclear rows of Verocay), wherein they exhibit a polar disposition. The type B pattern is composed of cells that are widely separated and haphazardly disposed in a loose reticular stroma. There is nonnuclear palisading or fibrillar regimentation in the type B pattern. The Antoni A part of the neurilemmoma showed intermediate signal intensity in both T1- and T2- weighted MR images and exhibited post contrast enhancement. The Antoni B part revealed hypointensity on T1-weighted images, hyper intensity in T2-weighted images, and showed no contrast enhancement. Orbital neurilemmoma are usually solitary [1] and generally grow slowly. The typical presentation is proptosis, restriction of ocular movements and impaired vision due to compression of optic nerve.

In our patient the visual acuity was not hampered because of slow progression of tumor and location of the mass was away from optic nerve. A long interval between the onset of proptosis and surgical removal is common. The superior quadrant is the commonest site for the tumour. This was true in our patient and the affected eye was displaced downwards. Neurilemmoma is essentially a benign lesion which rarely undergoes malignant transformation.[6] Neurilemmoma is usually treated with surgery by complete excision. Recurrence usually follows incomplete removal of the initial tumour whereas complete excision carries an excellent prognosis.

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