

CASE REPORT

A RARE CASE OF ISOLATED NEUROFIBROMA PRESENTED AS BENIGN SUBCONJUNCTIVAL GROWTH

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ABSTRACT: BACKGROUND: Neurofibroma is usually associated with systemic neurofibromatosis, but can occur as isolated lesions. Subconjunctival neurofibroma without any associated features of neurofibromatosis I is of rare occurrence and hasn't been reported in literature till date. **CASE:** We report a case of an atypical presentation of neurofibroma, diagnosed on histopathologically, without any associated features of neurofibromatosis I. 60 year old male presented with painful swelling in inner aspect of right eye since 2 years. Systemic Examination was normal. Patient didn't have neurofibromas on or in the body anywhere else. Patient did not have Café-au-lait spots or Leisch nodules on iris. Patient didn't have hypertrophy or destruction of orbital bone/exophthalmos. CT Scan Orbit showed intensely enhancing oval mass of size 19x19x21mm in intraorbital extraconal compartment anteroinferior to eyeball s/o Nerve Sheath Tumour. Excision Biopsy of lesion was done and sent for histopathological examination which confirmed the diagnosis of Neurofibroma. **CONCLUSION:** Isolated Neurofibromatosis should not be excluded in absence of systemic features of Neurofibromatosis I.

KEYWORDS: Isolated Neurofibroma, Subconjunctival benign growth.

KEYMESSAGE: Any mass presenting in subconjunctival region gradually progressing in size, possibility of isolated Neurofibromatosis should be kept in mind in absence of systemic features of Neurofibromatosis I.

INTRODUCTION: Orbital neurofibromas are peripheral nerve sheath neoplasms derived from Schwann cells, perineural cells and fibroblasts. They can manifest within the orbit, and they may or may not be associated with systemic neurofibromatosis.¹ Neurofibromas, relatively uncommon orbital lesions, represent approximately 2-4% of orbital tumors. Plexiform neurofibromas account for 2%, and localized neurofibromas account for 1%.² This is a rare case which presented as subconjunctival growth and turned out to be Neurofibroma. Three features of this case are unusual. Firstly, isolated subconjunctival neurofibromas are rare. Secondly, neurofibromas are usually associated with systemic neurofibromatosis. A thorough systemic examination in this patient showed no signs of neurofibromatosis. Thirdly, patients with these lesions often present at an earlier age than our patient in the sixth decade of life.

CASE: A 60 year old male, resident of Shilai, Distt Sirmour, labourer by occupation presented with a 2 year history of mass in the inner aspect of right eye (Fig 1). The onset of the swelling was spontaneous and gradually progressive. It was painless in the beginning and became painful at the time of presentation. No h/o Trauma, redness or diminution of vision. No h/o brown macules over the body. There is no similar family history. Patient has no systemic illness.

O/E: Visual Acuity was 6/60 OD and 6/36 OS.

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Right Eye Examination showed a large firm to hard mobile non tender mass in the inner aspect of the eye, not adherent to underlying structures. The conjunctiva was freely mobile over the growth. The posterior extent could not be reached. Conjunctiva, Cornea, AC, Iris appeared normal. Pupil was circular reacting to light. Lens was cataractous.

Left Eye Examination was normal.

Extra-ocular movement was full range. Sac syringing showed patency. Systemic examination does not reveal any abnormality.

Neurosurgery opinion was taken to r/o neurofibromatosis (to r/o neurofibromatosis) was normal. Skin reference was also made to r/o any cutaneous lesions which was also normal. Orthopedic opinion taken to r/o any bony abnormality (to r/o neurofibromatosis) was normal.

INVESTIGATIONS:

Hb-11.3gm%	WBC- 7.24 m/mm ³
FBS- 90 mg%	
Serum Urea – 25 mg%	Serum Creatinine – 07mg%
Total Proteins-6.1 mg%	Albumin- 4.1g/dL
SGOT- 7 IU/L	SGPT- 5 IU/L
Serum Na- 139 mEq/L	Serum Cl- 97 mEq/L
ECG- within normal limits	X-Ray Orbit- within normal limits.

CT Scan Orbit- showed intensely enhancing oval mass of size 19(transverse) x 19 (craniocaudal) x 21mm (anteroposterior) in intraorbital extraconal compartment anteroinferior to eyeball. The medial recti and superior oblique muscles are abutted by this mass and are thickened (Fig 2 & 3). CT findings were s/o Nerve Sheath Tumour? Schwannoma.

Excision Biopsy of the lesion was done and sent for Histopathology examination. It was a homogenous globular grey white mass of size approximately 18x18x7 mm. The posterior & posterolateral surface of the lesion were smooth, anterior & medial surface were rough. Histopathology examination showed interlacing fascicles of spindle shaped cells with uniform serpentine nucleus, inconspicuous nucleoli & moderate amount of eosinophilic fibrillary cytoplasm with occasional blood vessels, lymphocytes & mast cells. There is absence of cytological atypia or mitotic activity. The findings confirmed the diagnosis of Neurofibroma (Fig. 5.)

DISCUSSION: Three types of neurofibroma occur in the orbit: solitary, diffuse, or plexiform (the latter is considered pathognomonic of neurofibromatosis). Ocular involvement may include the eyebrow, eyelids, conjunctiva, iris, choroid, optic nerve, and orbit. Kalina et al³ reviewed the literature for isolated conjunctival neurofibromas and documented a detailed description of 13 cases in the literature. Of the 13 cases, 10 had systemic neurofibromatosis. The lesions were located at the limbus in over half of the cases, and the remainder was located on the upper tarsal conjunctiva and on the temporal bulbar conjunctiva. At this location, they can be mistaken for a dermoid. None were documented at the subconjunctival region. Simple excision was curative in these cases, similar to ours, as the growth of these lesions is characteristically uniformly slow.

TREATMENT: Surgical Excision/ Biopsy of the tumour is done because of cosmetic concern. Risk of malignant transformation from solitary neurofibroma is extremely small.⁴

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CONCLUSION: Isolated Neurofibromatosis should not be excluded in the absence of systemic features of Neurofibromatosis I.

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Fig. 1: Showing mass in the right eye.



Fig. 1

Fig. 2: Showing the rough medial surface of the mass after excision.



Fig. 2

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Fig. 3: CT scan (Coronal view) showing intensely enhancing oval mass in intraorbital extraconal compartment anteroinferior to eyeball abutting superior oblique muscle.



Fig. 3

Fig. 4: CT scan (Axial view) showing intensely enhancing oval mass in intraorbital extraconal compartment medial to eyeball.



Fig. 4

Fig. 5: Neurofibroma: interlacing fascicles of spindle shaped cells with uniform serpentine nucleus, inconspicuous nucleoli & moderate amount of eosinophilic fibrillary cytoplasm.

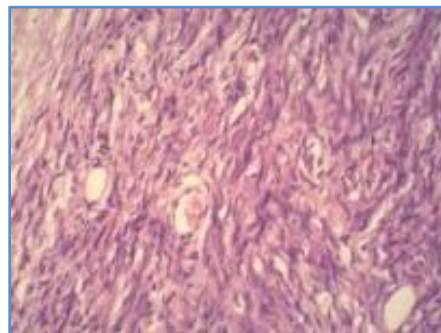


Fig. 5

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