

Orbital Myxoma - An Unusual Cause of Inferoorbital Mass

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SUMMARY

To describe a case of isolated infraorbital mass which had been present for the past 9 years in a young woman. Despite the size, the mass was successfully excised in total. Method: Case report. Result: Histologically, the tumour was found to be an isolated orbital myxoma. Conclusion: Isolated orbital myxoma is an exceedingly rare tumour of the orbit. The case shows the lesion could be excised in its entirety with good cosmetic result.

KEY WORDS:

Infraorbital mass, Orbital myxoma, Orbital tumour

INTRODUCTION

Orbital myxoma is an extremely rare benign tumour of mesenchymal origin and only a handful of cases have ever been reported in the literature¹. We report a case of myxoma of the orbit in a young woman.

CASE REPORTS

A 23 year-old Dayak woman living in a remote part of Sarawak was referred by a charity organisation because of a large painless mass in the lower orbit for the past 9 years. She was seen once three years ago in a local hospital where the doctor attempted to aspirate the mass on a few occasions but without success. She could not visit the tertiary referral centre because of financial constraints. Her past medical history was only significant for the presence of a ventricular septal defect which was confirmed later on echocardiogram on admission.

At the time of referral, the mass was confined to the lower eyelid causing a significant non-axial proptosis of the globe. The eye was displaced nasally and the corneal reflex from the midline measured about 8mm less as compared to the left. Vertical displacement was more and measured about 15mm higher than the fellow eye (Figure 1a). The left extraocular movements were restricted in all directions especially inferiorly. There was diplopia worst on looking to the left and also down. The mass was non-pulsatile, firm to touch and not reducible on retropulsion. Otherwise, the examination of the eye structures including the anterior segment, optic nerve, retina and intraocular pressure was normal. The presenting visual acuity was 6/6 in both eyes and there was no relative afferent pupillary defect.

The orbital CT scan showed a well circumscribed encapsulated homogenous mass occupying the inferotemporal quadrant with extension to the orbital apex. (Figure 1b and c)

A planned anterior orbitotomy under general anaesthesia was carried out. The orbitotomy was performed via a subciliary skin incision with a split skin approach. The mass was dissected from the surrounding tissue and removed en-bloc (Figure 1d). The lesion was pale-yellow, 9 cm long and worm-like with a large anterior end which tapered off towards the posterior end (Figure 1d). It was firm and rubbery in consistency and when the lesion was bisected the cut surfaces had a shiny gelatinous appearance (Figure 2a). The orbital floor was thinned due to chronic compression from the mass. The wound was closed after excising the excess skin.

Histopathological examination revealed hypocellular myxoid stroma containing scattered spindle cells. The cells contained hyperchromatic oval nuclei. There were no mitosis, pleomorphism or large atypical cells noted (Figure 2b). The features are suggestive of myxoma.

Post-operatively, we followed up the patient over 1 year duration and there was no recurrence of the growth. Postoperative visual acuity remained the same but both the proptosis and diplopia improved. The corneal light reflex in the left eye from the midline of the nasal bridge now matched that of the fellow eye and vertical displacement came down to 5mm from the contra lateral eye's corneal reflex (Figure 2c).

The patient also had no trouble with diplopia post surgery.

DISCUSSION

Myxoma is an uncommon benign tumour of mesenchymal origin and only a few cases have ever been reported in the medical literature¹⁻⁴. It is characterised by abundant mucoid material with a relatively small number of spindle and stellate-shaped cells. Most cases occur in the left atrium of the heart. In the head, most myxomas are reported in the mandible or the maxillary sinus. Its occurrence in the latter can cause secondary orbital invasion. Isolated orbital myxoma is extremely rare and only a few cases are reported and most do not grow to the size as in our patient.

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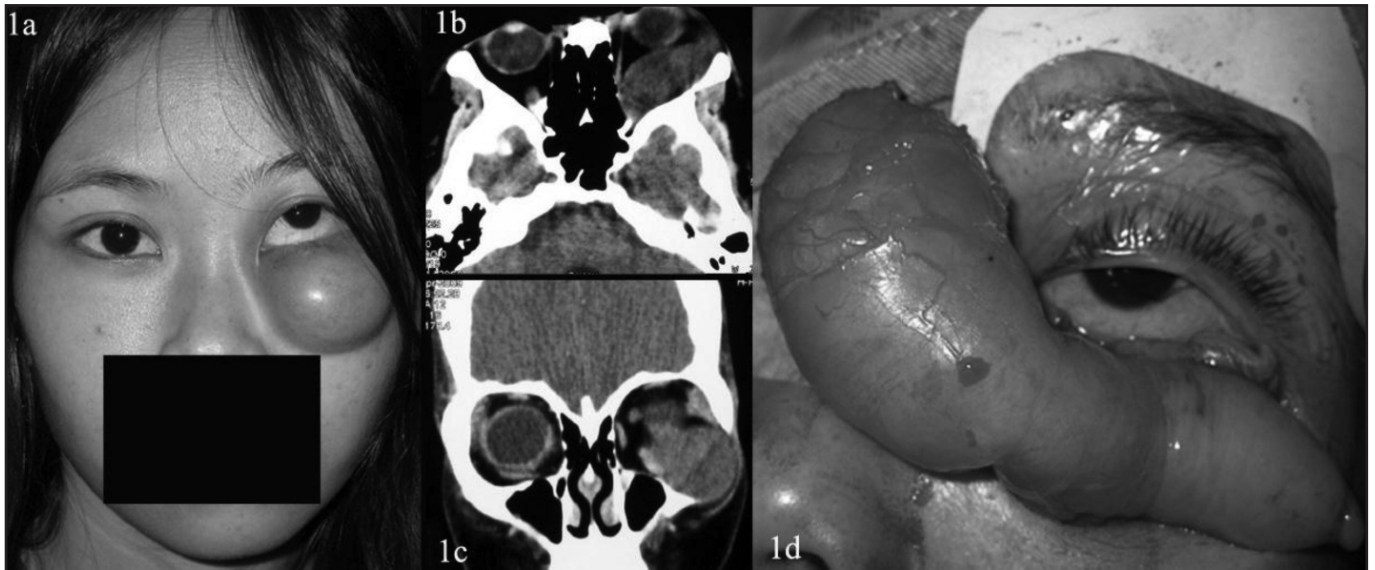


Fig. 1a : Left eye showing upward displacement by a large inferior orbital mass.
Fig. 1b and 1c : CT scans showing a well-defined orbital mass extending to the orbital apex.
Fig. 1d : Intraoperative picture showing worm-like tumour removed from the inferior orbit

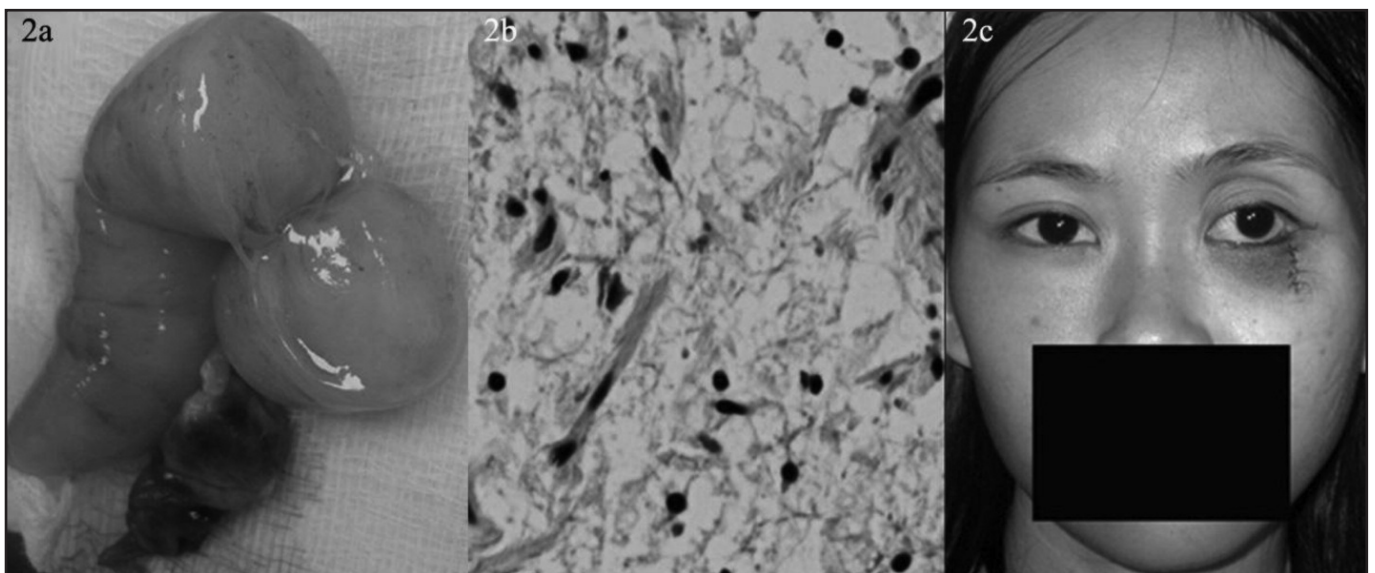


Fig. 2 : The cut surfaces of the tumour showing a shiny gelatinous surface.
Fig. 2b : Histology showing abundant myxoid extracellular stroma with sparse spindle-shaped cells.
Fig. 2c : Improved cosmetic appearance following removal of the tumour.

Myxomas grow slowly and may be present several years before consultation is sought, occasionally attaining sufficient size to produce considerable facial deformity. Although benign, these tumours are locally destructive and aggressive and may extend into the surrounding structures such as the nasopharynx, nose or paranasal sinuses. The treatment of choice is excision of the lesion en bloc to avoid future growth or recurrence. Most myxomas are well-defined and not diffusely infiltrative making it easy to remove in its entirety without damage to the surrounding vital structures such as the optic nerve.

Although most reported cases of orbital myxomas are isolated, it is essential to look for signs of Carney syndrome⁵. This syndrome has multi-system involvement and is inherited in an autosomal dominant fashion with variable expressivity. It is characterized by spotted pigmentation of the skin and mucous membrane, myxoma, endocrine overactivity and schwannoma. The spotted pigmentation is often the first sign of the syndrome and so any patient with myxoma should have the skin examined. The diagnosis of this condition is important as the patient may have associated atrial myxoma which can be detected early before it causes a fatal embolic event. However, this syndrome is not present in our patient.

This case shows that despite the significant disfigurement that an orbital myxoma may give rise to, it is possible to achieve good cosmetic result if the lesion is carefully dissected and removed completely.

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