Giant Solitary, Encapsulated, Venous-lymphatic Malformations (Cavernous Haemangioma) - A Case Report


Abstract

Solitary encapsulated venous-lymphatic malformation (SELVM) is the most common benign orbital tumour in adults. A 55-year-old woman residing in a tribal area of Thane district presented with severe non-axial proptosis of the right eye of 20 years duration. Computed tomography scans of the orbit showed a well defined, oval solid mass occupying the whole right orbital cavity measuring 7.3 x 5 x 4 cm. The patient underwent surgical excision of the tumour through a temporal transconjunctival approach combined with lateral canthotomy. To the best of our knowledge, in the available literature, among SELVM (cavernous haemangioma), present case is the largest.

Introduction

Solitary, encapsulated, venous-lymphatic malformations (SELVM) is the most common benign orbital tumour in adults1-3. It is generally considered developmental hamartoma but does not become symptomatic until the third to fifth decade of life.1 It is usually found within the muscle cone and rarely produces proptosis more than 5 mm. We report a case of a giant SELVM of the orbit in a 55-year-old woman causing non-axial proptosis.

Case Report

A 55-year-old woman residing in a tribal area of Thane district presented with history of gradual progressive painless protrusion of the right eye of 20 years duration. It was associated with decrease in vision over the last 10 years.

On examination, the best corrected visual acuity in the right eye was no perception of light and left eye was 6/9. The right eye had severe non-axial proptosis with inferonasal dystopia, pushing the eye ball completely outside the orbit (Fig. 1). A firm mass was palpable on supero-temporal aspect of the orbit measuring approximately 5 x 4.5 cm. The conjunctiva over the mass had bluish discolouration and the exposed conjunctiva was markedly keratinised. There was lagophthalmos and linear leucomatous corneal opacity crossing pupillary area. The fundal glow was absent due to complicated cataract.

Computed tomography scans of the orbit showed a well defined, oval solid mass occupying the whole right orbital cavity measuring 7.3 x 5 x 4 cm (Fig. 2). Chronic widening and erosion of orbital walls, floor and roof was noted. Delayed cuts revealed ‘filling in’ of the lesion with smooth homogeneous appearance. B-scan ultrasonography corroborated the solid nature of the mass. It also showed very low flow pattern consistent with venous flow pattern.

The patient underwent surgical excision of the tumour through a temporal trans-conjunctival approach combined with lateral canthotomy under general anaesthesia. The tumour mass was enucleated after sorting out the plane between periorbita and tumour. The post-operative period was uneventful.

Gross examination showed the violet coloured tumour which was well capsulated, soft and slightly
nodular measuring 7 x 5 x 4 cm.

Histopathology examination showed large dilated vascular channels, at places having papillary infolding and blood in the lumen (Fig. 3).

There was no recurrence of the tumour after a follow up of one and half years (Fig. 4).

**Discussion**

SELVM are benign, non-infiltrative, slowly progressive tumours of large endothelial channels.\(^4\) For years, SELVM have been called cavernous haemangioma. According to modified ISSVA (International Society for Study of Vascular Anomalies) classification, cavernous haemangiomas are now classified as SELVM under vascular malformations.\(^1\)

They are generally considered developmental hamartomas or choriostomas. The mass is solitary, unilateral, well tolerated, encapsulated and commonly located in the retrobulbar space leading to slowly developing axial proptosis in the 3\(^{rd}\) decade of life.

In the present case, the mass was solitary, extraconal and unusually large (7.3 x 5 x 4 cm). To the best of our knowledge, in the available literature, among the cavernous haemangiomas, present case is the largest. The maximum size recorded is 2.3 x 3 x 3.7 cm in the retrobulbar space.\(^5\)

There was unusual delay in seeking medical help by the patient inspite of cosmetic disfigurement along with complete loss of vision in the affected eye. CT scan revealed chronic widening and pressure erosion of the orbital walls, roof and floor suggesting...
chronicity of the process.

In SELVM, most often the surgical approach is lateral orbitotomy due to intraconal location of the mass. In the present case, during surgical excision, transconjunctival approach was taken as the tumour was extraconal and protruding outside the orbit.

References

STORM OVER STATINS - THE CONTROVERSY SURROUNDING PHARMACOLOGIC TREATMENT OF CHILDREN

There are limited, short-term data showing that statins appear safe in children, though long-term follow-up is completely lacking. At 8 years of age, a child’s brain and other organ systems remain in dynamic stages of growth and development, raising concern that long-term pharmacotherapy initiated at this age may adversely affect the central nervous system, immune function, hormones, energy metabolism, or other systems in unanticipated ways.

Regardless of how many additional children may receive statin treatment under these new guidelines, the broader, more important question is whether we intend to treat paediatric obesity with an ever-increasing array of powerful adult drugs - beta-blockers and diuretics for hypertension, aspirin for coagulopathy, insulin sensitizers for the metabolic syndrome, and of course insulin for diabetes.