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Solitary fibrous tumor of orbit: A rare entity

Sir,

Solitary fibrous tumor is a rare spindle cell neoplasm. It generally occurs in serosal surfaces and pleura are the commonest site. It may occur in head and neck sites like orbit, sinonasal cavity, salivary gland, thyroid, infratemporal fossa and parapharyngeal space. About less than 50 cases of orbital SFT have been reported in the world literature till date.

A 44-year-old male presented with a history of painless swelling in the left eye since 2 years. He gave a past history of enucleation of the left eyeball due to penetrating trauma to his left eye 13 years back. On examination, there was a swelling in the left orbit with soft cystic component in the superolateral aspect [Figure 1], with no palpable neck nodes.

CECT scan revealed a homogeneously enhancing intraorbital mass with marked widening of the left orbital cavity [Figures 2 and 3].

The patient underwent orbital exenteration (left) and the defect was covered by split skin graft.

Post-operative histopathology revealed alternate hypo- and hyper-cellular spindle cellular areas. The cells were round and oval and arranged around blood vessels [Figure 4]. Muscles and fatty tissue were involved. Immunohistochemistry revealed strong CD34 positivity [Figure 5], vimentin positivity and focally Bcl_2 positive, but negative for CK, S 100, SMA, EMA, Desmin, Keratin and CD 68 supporting the histological diagnosis of Solitary fibrous tumor. The patient is on regularfollow-up since last 2 year and the patient is disease free till the time of reporting.

Orbital SFT was first reported by Dorfman *et al*. and Westa *et al*. in 1994.[1]

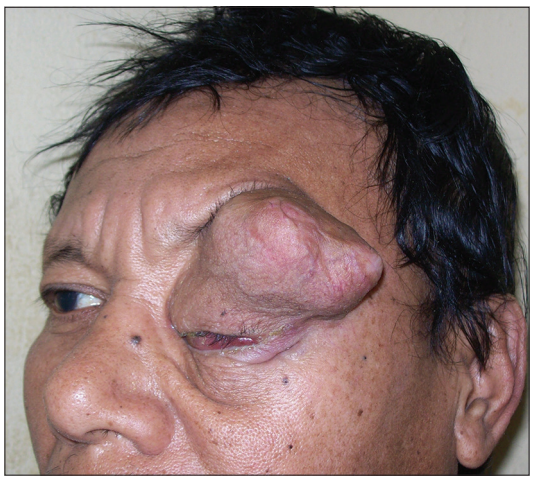

Figure 1: Clinical photograph of the patient with a swelling in the superolateral aspect of left eye with proptosis

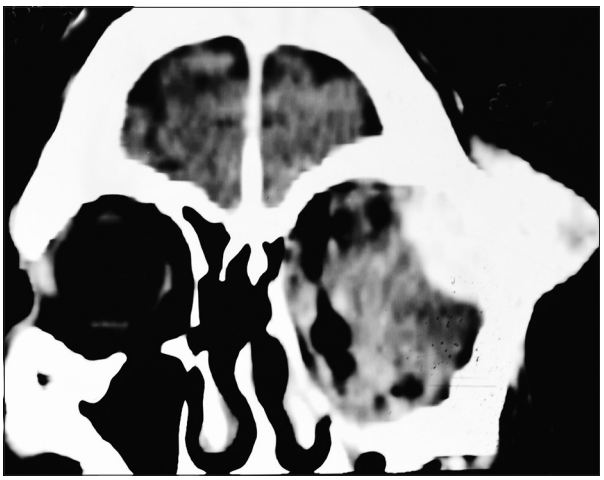

Figure 2: Coronal CECT of the intraorbital mass with marked widening of left orbital cavity

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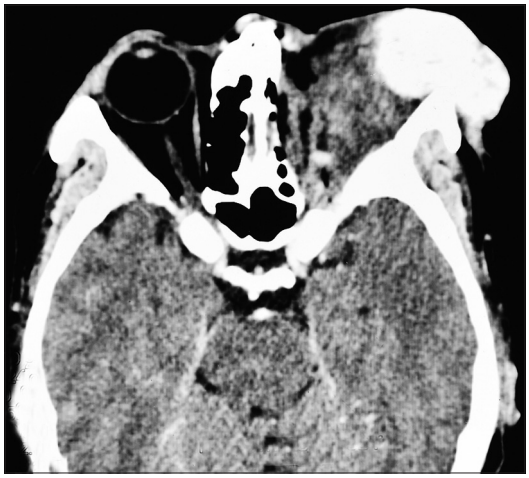

Figure 3: Axial CECT of the intraorbital mass showing intense homogenous enhancement

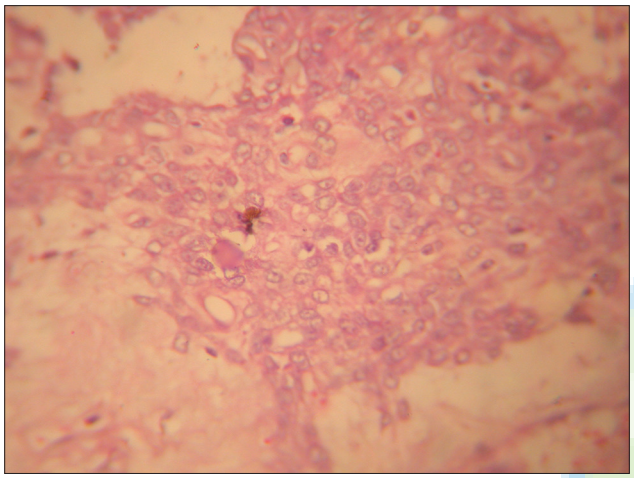

Figure 4: H and E stain showing alternate hypo and hyper cellular spindle cell areas arranged around blood vessels

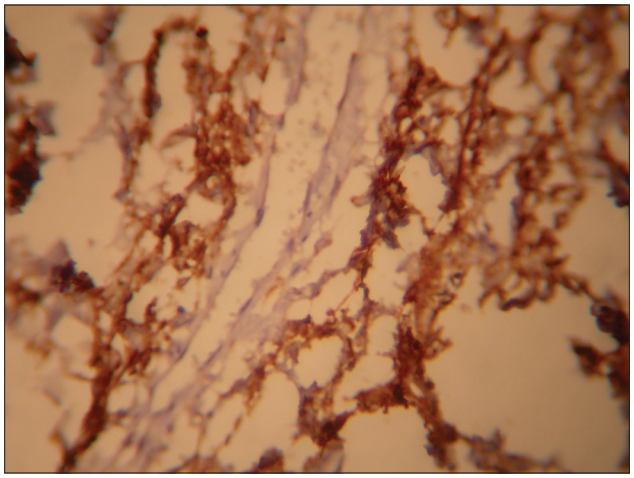

Figure 5: Immunohistochemistry to show CD 34 Positivity in tumor cells

Clinically SFT of the orbit usually presents as a slow growing, unilateral painless proptosis (60%). The reported radiological head and neck SFT are rather non-specific. Although not pathognomic, homogenous and heterogenous attenuated enhancement is reported to be the most prominent feature of SFT, revealed on CT and MR Images.[2]

The diagnosis of SFT can be confirmed by Immunohistochemistry analysis of CD34 which is the prime marker. Diffuse and strong immunoreactivity has been demonstrated in 79 -100% cases.[3] Histologic differential diagnosis includes other mesenchymal tumors, like fibrous hitiocytoma (CD68 positive), hemangiopericytoma, fibrous meningioma (EMA positive), and leiomyoma(Actin positive). SFT may also be immunoreactive to vimentin and BCL 2.

The long-term prognosis of head and neck sites of SFT is still uncertain due to the limited number of reported case. Treatment is complete surgical excision. Resectability is the most important prognostic factor. Local recurrences of SFT are usually due to an incomplete resection. Recurrent tumors of the orbit tend to infiltrate surrounding tissue and bone, thus complicates secondary excision.[4] Radiotherapy is advocated for incomplete resection. Chemotherapy is reserved for histological aggressive tumor. Although enblok resection is the definitive treatment but residual tumor may be stable for several years, so close long-term follow-up is necessary.[5] Although extremely rare, the Head and Neck Surgeon and Pathologist should be aware of this entity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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