



Case Report

A rare case report of orbital malignant solitary fibrous tumor

Priyam Gupta¹, Somanath N², Chinmayee Thrishulamurthy^{1,*}, Dayananda S Biligi²¹Dept. of Ophthalmology, Minto Eye Hospital, Bengaluru, Karnataka, India²Dept. of Pathology, BMCRI- Bangalore Medical College and Research Institute, Bengaluru, Karnataka, India

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ABSTRACT

Purpose: Orbital Solitary Fibrous Tumor (SFT) is a rare spindle cell neoplasm of mesenchymal origin. We describe the clinical presentation, radiological features, pathological features of a 23yr old female patient with orbital malignant SFT.

Observation: We report a case of Malignant Orbital Solitary Fibrous Tumor of the orbit in a 23yr old female who presented with painless unilateral proptosis of the left globe noticed by her in the last 1 month. Computed Tomography scan showed a well-defined homogeneous enhancing soft tissue density in superior and extraconal compartment, with no evidence of bony erosion or calcification. En bloc tumor resection confirmed SFT based on histopathological and immunohistochemistry studies. Microscopic examination showed a highly cellular tumor, moderate amount of pleomorphism and increased mitosis ($>4/10$). Immunohistochemistry studies confirmed diagnosis as tumor was diffuse positivity with CD34, Bcl2 positive, STAT6 positive with ki 67 high i.e. a 9-10% proliferative index in hot spot areas. Desmin, CD99, S100 and EMA was negative. Patient subsequently was also given External Beam Radiotherapy.

Conclusion: Solitary Fibrous Tumors are rare tumor, which should be included in differential diagnosis of soft tissue tumor of the orbit. Combination of Computed Tomography scan, histological examination and immunohistochemical markers provide an accurate diagnosis. Complete surgical resection along with long term follow up is essential.

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1. Introduction

Solitary fibrous tumor is a distinct rare spindle cell neoplasm^{1,2} of mesenchymal origin^{3,4} it arises frequently in the pleura.¹⁻⁶ Only recently has it been recognized in extraserosal locations such as lung, mediastinum, liver, thyroid¹ nasal and paranasal sinus, parotid and salivary glands, spine and orbit.⁷ Solitary fibrous tumors arising in the orbit are relatively rare.¹ Benign and malignant forms are reported however, benign variants appear to be more common.²

We report a case of Malignant Orbital SFT in a 23yr old female and discuss the clinical presentation, radiological characteristics, histological and immunohistochemical features, treatment of this rare tumor with a review of literature.

* Corresponding author.

E-mail address: chinmayee.jt@gmail.com (C. Thrishulamurthy).

2. Case Report

A 23yr old female patient presented with history of painless swelling and outward protrusion of the left eye which she noticed since last 1 month (Figure 1). However, photographs more than 6 months old showed presence of left eye protrusion. There were no visual symptoms. Ocular examination revealed a 2mm downward and inward displacement of the left eye with fullness in outer aspect of the left upper eyelid. There was no palpable mass, fingers could be insinuated all around the orbital margin. Visual acuity, Pupillary reaction, anterior segment examination, extraocular movements, fundus examination and visual fields were within normal limits.

Computed tomography (CT) showed a well-defined homogeneous enhancing soft tissue density in superior and extraconal compartment measuring 2x1.4cms. Lesion

extended posteriorly causing proptosis and was seen displacing the lateral rectus muscle. The left lacrimal gland was not seen separately. There was no evidence of bony erosion or calcification. Optic tracts were normal bilaterally. (Figures 2 and 3)

En-bloc excision of the mass was done by lateral orbitotomy via lateral canthal incision (Figure 4).

Histopathological examination of the 25mmx20mmx15mm well encapsulated grey white to grey brown mass of tissue which showed a highly cellular tumor, arranged in form of sheets with short fascicles, numerous dilated and congested vessels in stroma. Intracytoplasmic lumina formation with RBCS in the lumina at periphery of the tumor. Moderate amount of pleomorphism and increased mitosis ($>4/10$). No areas of necrosis were noted. (Figure 5)

Immunohistochemistry studies showed diffuse positivity with CD34, Bcl2 positivity, STAT6 positive (Figure 6), ki-67 high with 9-10% proliferative index in hot spot areas (Figure 7). Desmin, CD99, S100 and EMA negative.

Patient was then subjected to radiotherapy from 11/1/2019 to 23/2/2019. She completed a course of External Beam Radiotherapy via Volumetric modulated arc therapy (VMAT). The doses were as follows: Optic nerve left eye: 53Gy, right eye: 10Gy. Chiasma: 29Gy, Lens left 45Gy, Right 3.2Gy, eye: left 56Gy and right 76Gy and retina 54Gy, 6Gy right and left respectively.

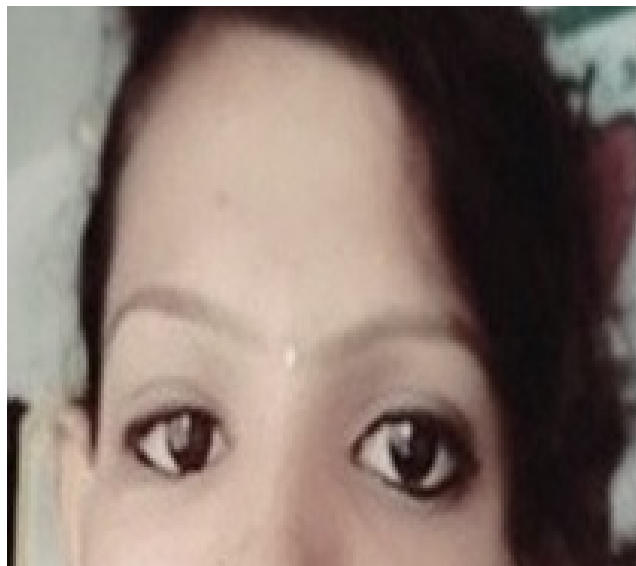


Fig. 1: Initial presentation with Left eye proptosis

3. Discussion

Solitary fibrous tumor is a rare, spindle shaped neoplasm that has been described on the mesothelial surface of pleura and the mediastinum.⁷ The neoplasm was originally described by Klemperer and Rabin in 1931.¹ More recently



Fig. 2: Computed Tomography of orbit

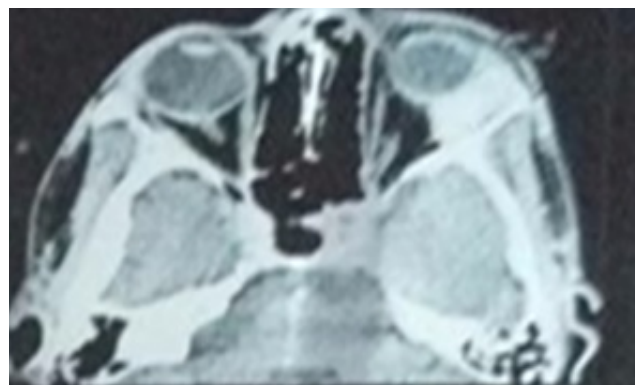


Fig. 3: Computed Tomography of orbit



Fig. 4: Status post en bloc tumor excision via lateral Orbitotomy via Lateral Canthal Approach

the tumor has been described in extraserosal sites like lung, liver, paranasal sinuses, and the orbit, suggesting a mesenchymal- fibroblastic origin^{4,7}

The first SFT presenting in the orbit was independently reported in 1994 by Dorfman et al and Westra et al respectively.¹ Orbital SFT can affect any orbital space including lacrimal gland fossa and intraconal or extraconal space.⁷ In vast majority of cases, they have a benign and indolent course with no tendency of distant metastasis.⁷

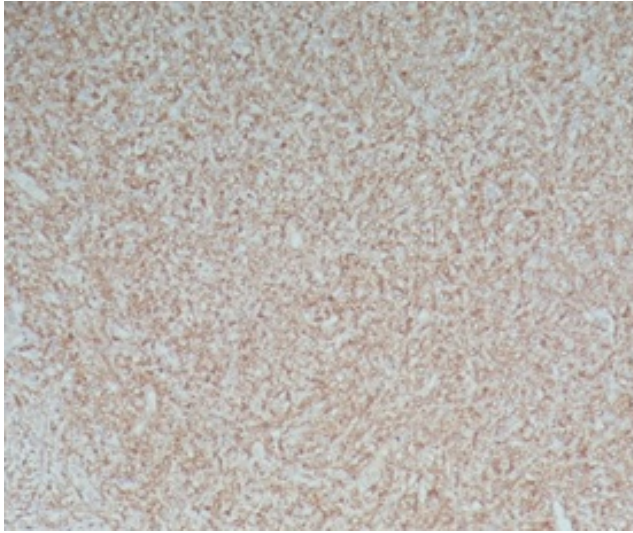


Fig. 5: CD 34 positive slide

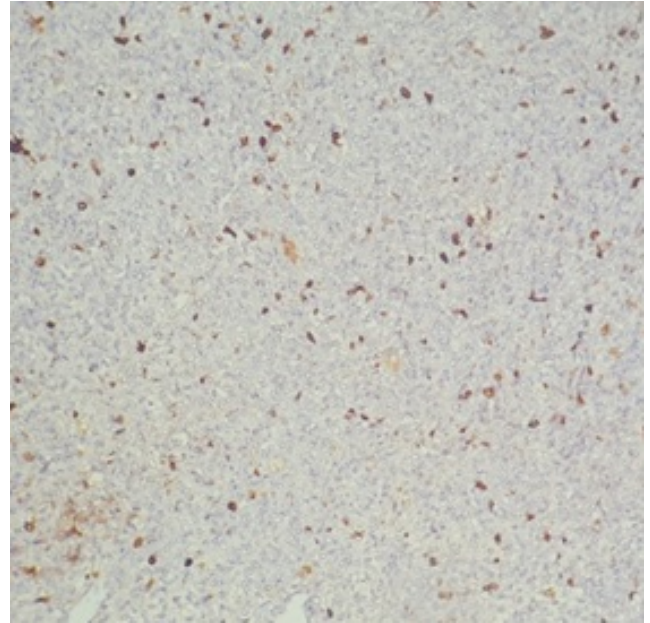


Fig. 7: Ki 67 positivity

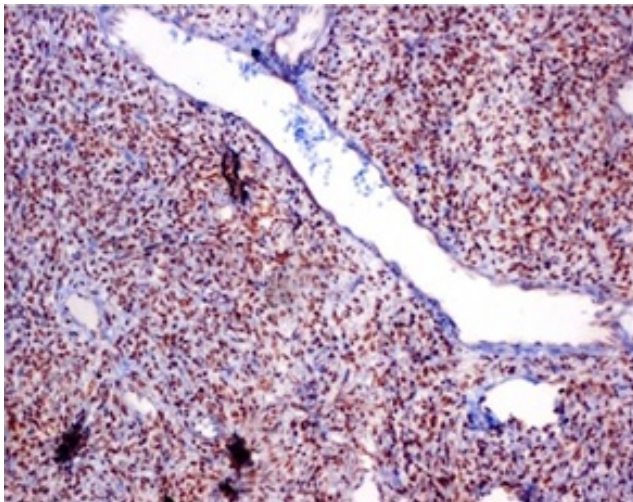


Fig. 6: STAT 6 positivity

Orbital SFT is unilateral and occur both in children and adults.⁸ A wide age range has been reported from 9-76yrs without predilection to sex.^{4,9} Clinically it usually presents as a slow growing, unilateral, painless proptosis⁶ with an insidious onset over a mean duration of 6-8 months.¹

On imaging Solitary Fibrous Tumor is generally seen as a well-defined soft tissue mass with homogeneously or heterogeneously strong enhancement on CT and MR. Differential diagnosis include capillary haemangiomas, cavernous haemangiomas, haemangiopericytoma and giant cell angiofibroma. Giant cell angiofibroma display enhancing characteristics similar to the orbital SFTs.⁴

Histopathological diagnosis of SFT shows tumor matrix is composed of spindle shaped cells with scant cytoplasm and indistinct nucleoli. Matrix consists of distinctive thick ropey type of collagen between the tumor cells. Tumor

cells are randomly oriented and have a so called patternless pattern.² It is important to differentiate SFT of the orbit from other spindle cell tumors¹ like Haemangiopericytoma (staghorn pattern) and fibrous histiocytoma (storiform architecture).² Immunohistochemical analysis continues to be the gold standard for diagnosis.³ Immunoreactivity with marker CD34 in 79-100% cases, positive for mesenchymal markers such as vimentin and Bcl2. Negative for desmin, cytokeratin, vascular markers like cytokeratin, neural markers(S-100) and muscle specific actin and smooth muscle actin.¹ STAT6 is a highly specific marker for SFTs. Recent biomolecular studies have suggested the potential role of NGFI-A binding protein 2 (NAB2) and nuclear signal transducer and activator of transcription 6 gene (STAT6) as the SFT specific fusion gene. The prognostic role for determining disease free survival remain clear.³

Solitary fibrous tumor, fibrous histiocytoma, haemangiopericytoma all show positivity for BCL2, vimentin and CD34, but to varying degree. Fibrous Histiocytoma shows focal positivity for Bcl2 or CD34. Haemangiopericytoma has similar histological features but weak and patchy CD34 positivity in contrast to solitary fibrous tumor which has strong and generalized positivity to CD34.⁷ Criteria for malignancy include the presence of more than 4 mitosis per high power field (or presence of abnormal mitotic figures), cellular pleomorphism.¹

Treatment of an SFT of the orbit consists of complete surgical excision with long term follow up. Various surgical approaches are available: fronto orbital approach, pterional approach, lateral orbitotomy and medial orbitotomy. Intracranial extension are addressed via transcranial-

fronto-orbital approach.⁵ Single most important factor in predicting recurrences is initial incomplete excision.^{4,7}

4. Conclusion

SFT should be considered in the differential diagnosis when a patient presents with a well circumscribed lesion causing unilateral proptosis which enhances on CT scan evaluation. Histopathological examination and immunohistochemical markers are a gold standard for diagnosis. Complete surgical excision and long term follow up of patient is important to monitor for recurrences.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Author biography

Priyam Gupta Junior Resident

Somanath N Junior Resident

Chinmayee Thrishulamurthy Assistant Professor

Dayananda S Biligi Professor and Head

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