ORBITAL SOLITARY FIBROUS TUMOR: CASE REPORT WITH NUCLEAR STAT6 EXPRESSION AND CD34+ WITH LITERATURE REVIEW

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ABSTRACT Background: solitary fibrous tumour (SFT) has been documented in many other organs after its original pleural description. Orbital SFT is very rare and only 263 cases from the English literature are reported since 2021. The use of immunohistochemical markers is very helpful in the diagnosis especially CD34 positivity and STAT6 immunoreactivity.

Case report: A 50-year-old man presented with a 15 months history of progressive left exophthalmos. MRI showed a well-circumscribed, expansible soft tissue mass located above the right globe extending posterolaterally. The lesion was isointense to the extraocular muscles and cerebral cortex with strong enhancement with gadolinium. The patient underwent a left lateral orbitotomy. Total removal was achieved under a microscope and cryotherapy tool.

Results: By using an immunohistochemistry panel; the tumour was stained intensely for vimentin, CD34, and STAT6 and it was negative immunoreactivity to pancytokeratin (AE1/AE3), S100 protein, epithelial membrane antigen, glial fibrillary acidic protein, and FVIII. With 12 months follow-up, no recurrence was seen.

Conclusion: Orbit SFT is very rare. The generally accepted treatment for primary and recurrent orbital SFT is complete excision. The use of immunohistochemical markers is very helpful in the diagnosis. orbital SFTs are rare but can be reliably diagnosed based on the presence of characteristic morphologic features and STAT6 and CD34 immunohistochemistry. Nuclear staining of STAT6 is a useful diagnostic adjunct in conjunction with CD34 positivity to help delineate SFT from histologic mimics. Orbital SFTs are generally accepted as benign tumours with a favourable clinical course. However long term clinical follow-up is recommended.

KEYWORDS Exophthalmos, Orbital neoplasm, lateral orbitotomy, Immunohistochemistry CD34+, STAT6, Solitary fibrous tumor

Copyright © 2021 by the Bulgarian Association of Young Surgeons
DOI: 10.5455/IJMRCR.OrbitalSolitaryFibrousTumor.
First Received: October 28, 2021
Accepted: December 22, 2021
Associate Editor: Ivan Inkov (BG);

Introduction

First described in 1931 [1], solitary fibrous tumour (SFT) has been documented in many other organs after its original pleural description. It is much more common around body cavities, such as pleura, peritoneum, and meninges, and thus orbit tumours may be related to the meninges. [2,3,4]

Orbital involvement was first reported in 1942, and since then, approximately 263 cases originating in the orbital region have been described in English literature. Recently Lester D. R. Thompson and al. 2021 [4] can report 263 cases from the English literature.
The use of immunohistochemical markers is very helpful in the diagnosis. Nuclear staining of STAT6 is a useful diagnostic adjunct in conjunction with CD34 positivity to help delineate SFT from histologic mimics. A case of orbital SFT confirmed by CD34+, and STAT6 immunoreactivity is reported.

Case report
A 50-year-old immunocompetent Moroccan man presented a 15 months history of progressive left exophthalmos. Recently he was accused of a decrease in his visual acuity, with some intermittent diplopia. His medical history was unremarkable. He was good healthy. On examination, he had no history of trauma, and his overall medical history was unremarkable. On ophthalmologic examination, normal visual acuity in the left eye and light perception on the right. His physical examination was notable for moderate exophthalmos of the left eye. No neurological deficits were noted. Somatic examination showed no abnormalities.

Magnetic resonance imaging (MRI) showed a well-circumscribed, expansible soft tissue mass located above the right globe extending posterolaterally close to the optic nerve that was medially deviated. Compression on the globe with exophthalmos was evident; however, no obvious invasion of either the optic nerve or the adjacent ocular muscles or bone was noted. The lesion was isointense to the extraocular muscles and cerebral cortex. The same lesion enhances avidly with gadolinium (Fig 1). The patient underwent a left lateral orbitotomy. Total removal was achieved under a microscope and cryotherapy tool. Intraoperatively, the tumour had a soft, capsular texture and a vascular nature. The perioperative diagnosis was a meningioma-like appearance with extensive adhesions to surrounding muscular structures. We use cryotherapy in surgical orbital hemangioma. This tool was very helpful in our experience in 10 orbital hemangiomas according to Cryo-assisted minimally invasive surgery to treat orbital cavernous hemangiomas. The Postoperative was unremarkable. He reported some immediate postoperative amelioration in his left eye. He was referred to the ophthalmological department for follow-up. Postoperative orbital CT scan showed total removal of the lesion with good cosmetic reconstruction with the disappearance of exophthalmos.

Histological and immunohistochemical studies showed a Proliferation with spindle-shaped cells arranged in short intersecting bundles. There was no evidence of haemorrhage, necrosis, or infiltrative borders nor architectural signs of malignancy. By using an immunohistochemistry panel; The tumour was stained intensely for vimentin, CD34, and STAT6, and it was negative immunoreactivity to pancytokeratin (AE1/AE3), S100 protein, epithelial membrane antigen, glial fibrillary acidic protein, and FVIII. With 12 months follow-up. No recurrence was seen.

Discussion
First described in 1931 [1], solitary fibrous tumour (SFT) has been documented in many other organs after its original pleural description. Although initially described in the pleura, SFTs have subsequently been encountered in extrapleural locations, including the pericardium, mediastinum, peritoneum, paranasal sinuses, and orbit [3-5]. Orbital localization is very rare. In general, orbital SFTs affect both sexes equally and present in a younger age group (median 42 years) than the 5th to 7th decades for other anatomic sites.

The first orbital and orbito-facial locations have been reported since 1994. The prevalence of the SFT among orbital tumours varies from 0.71 to 1.5% [5-8]. This frequency is probably underestimated because the extra-pleural locations still have problems of differential diagnosis with other fusiform cell neoplasms. The literature review reveals an accelerating rate of reports of orbital SFTs. This is most likely due to increased awareness and widespread use of immunohistochemistry for definitive diagnosis [9-14].

SFT of the orbit presents as a well-circumscribed solitary lesion that ranges in size from 1 to 3 centimetres and may arise anywhere in orbit, including intraconal, extraconal, both intraconal and extraconal, and in any quadrant.

The most common ophthalmic manifestation is unilateral exophthalmos, and it can be accompanied by eyelid swelling, vision disturbances, a palpable mass, tearing, or ptosis [2,3,4,14]. The majority of patients experienced unilateral exophthalmos with visual changes and headaches. Visual changes included double vision, blurred vision, and/or flashing lights. Hyde RA and al.[9] reported a case of SFT to the orbit with growth during pregnancy. The tumour was strongly positive for the progesterone receptor, consistent with its clinical growth during the antenatal and postnatal periods.

Our patient presented progressive exophthalmos. In his literature review, Lester D. R et al. [4] reported size highlights that 95.6% of all cases are ≤ 5 cm. As such, tumours in this anatomic location are smaller than other sites. Bouazza and al.[5] reported a neglected and historical case of orbito-facial SFT with huge dimensions: 28 cm vertical diameter, 8 cm horizontal diameter, and 9 cm in the anteroposterior diameter. The diagnostic was confirmed by immunoreactivity for CD34. This tumour sits in the right orbital region, and it extends to the upper and lower right eyelids and the lower half of the right hemiface. This historical case testifies to these tumours’ aggressive and progressive
SFT imaging characteristics have been described using the cerebral cortex and extraocular muscles for reference. They include isodensity on CT isointense on T1-weighted MRI and heterogeneous isointense to hyperintensity on T2-weighted scans. In both modalities, SFT enhances robustly with contrast with inconsistent homogeneity. The pattern of contrast enhancement and washout on dynamic imaging may help differentiate between cavernous hemangioma, SFT, and schwannoma. Occasionally demonstrating internal cystic areas. Depending on the extent of collagen, the tumours may have a low T2-weighted MRI finding. Some authors performed angiography to guide presurgical embolization in some reported cases.

Nuclear staining of STAT6 is a useful diagnostic adjunct in conjunction with CD34 positivity to help delineate SFT from histologic mimics. SFTs are consistently, strongly immunoreactive for CD34 (90–100%) [2,10,11]. The CD34 protein is a member of a family of single-pass transmembrane sialomucin proteins that are expressed in early hematopoietic and vascular-associated tissue. Therefore, CD34 is expressed early in the cells and embryonic fibroblasts. The high reactivity of CD34 is helpful for the diagnosis of SFT. The molecular driver aberration in the solitary fibrous tumour is a gene fusion between NGFI-A binding protein 2 (NAB2) with signal transducer and activator of transcription 6 (STAT6). The protein STAT6 is detectable by immunohistochemistry, is a reliable surrogate marker for the NAB2-STAT6 gene fusion, and is a highly sensitive and specific diagnostic marker for this tumour. [10] The generally accepted treatment for primary and recurrent orbital SFT is complete microsurgical excision. Many orbitotomy and cranio-orbital approaches were used according to the localization and the volume of the tumour in orbit. The tumour can be removed in toto or a piecemeal fashion. We used in our case cryotherapy device according to Cryo-assisted minimally invasive surgery to treat orbital cavernous hemangiomas. This tool was very helpful in our experience in 10 previous operated orbital hemangiomas.

In his cohort, blessing NW and al. [10] found that 24% of the tumors were removed in a piecemeal fashion. This increases the risk of leaving behind residual microscopic disease with a concomitant risk of local recurrence. He found no clinicopathologic correlates in his study that predicted recurrence, including fragmentation at the time of removal, and no patients had demonstrable metastases or disease-attributable death. However, this must be weighed against the risk of blindness, diplopia, and ptosis for persistent or recurrent disease in the orbital apex. Orbit SFTs are uncommon tumours displaying a generally benign clinical behaviour, but with a small subset of cases showing local recurrence and/or distant metastatic disease. These tumours have been shown to have a characteristic histologic appearance of a fibroblastic population set within a variably collagenized stroma and associated with branching, patulous, slit-like, or staghorn type vessels. [10] The case reported by Bouazza and al.[5] significantly demonstrates potential aggressive behaviour if the tumour was neglected and not operated on. Therefore, complete surgical resection is this tumour’s most important prognosis factor.

Conclusion

Orbit SFT is very rare. The generally accepted treatment for primary and recurrent orbital SFT is complete excision. The use of immunohistochemical markers is very helpful in the diagnosis. Orbital SFTs are rare but can be reliably diagnosed based on the presence of characteristic morphologic features and STAT6 immunohistochemistry. Nuclear staining of STAT6 is a useful diagnostic adjunct in conjunction with CD34 positivity to help delineate SFT from histologic mimics. Orbital SFTs are generally accepted as benign tumours with a favourable clinical course. However, some cases are aggressive and require long-term follow-up for recurrence. Therefore, long term clinical follow-up is recommended.

Funding

This work did not receive any grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflict of interest

There are no conflicts of interest to declare by any of the authors of this study.

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