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CASE REPORT

Solitary Fibrous Tumor of Orbit Mimicking Sarcoma in 14 Years Old Boy: A Rare Case Report

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ABSTRACT

A solitary fibrous tumor (SFT) is a rare spindle cell neoplasm representing <2% of all soft tissue tumors. SFT was first described in the pleura and previously thought to be arising from mesothelium but recent studies have shown that any part of the body can be affected. Here, we report a case of SFT arising from the extraconal compartment of the right eye of a 14-year-old boy who presented with painless, slowly progressive swelling of the right eye for two years. On the basis of clinical details, a diagnosis of sarcoma was made. MRI findings revealed a well-circumscribed heterogeneously enhancing extraconal right orbital lesion suggestive of low-grade sarcoma. The patient underwent wide local excision. Grossly, it was a grey-white firm tumor. Histopathological features and immunohistochemistry were consistent with solitary fibrous tumor.

Keywords: Solitary fibrous tumor, Spindle cell neoplasm, Immunohistochemistry, CD34.

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INTRODUCTION

The solitary fibrous tumor is a rare spindle cell mesenchymal tumor representing < 2% of all soft tissue tumors.[1] The solitary fibrous tumor was first described in the pleura and previously thought to be arising from the mesothelium,[2] but recent studies have shown that any part of the body can be affected by the SFTs,[3] like the skin nervous system, liver, lung, kidney, thyroid, etc.[4] In head and neck most common sites in descending order are sinonasal tract, orbit followed by oral cavity and salivary gland.[2,5] Here, we report a case of SFT arising from the extraconal compartment of the right eye of 14-year-old boy.

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CASE REPORT

About 14 years old boy presented with painless, slowly progressive swelling of the right eye for two years. Ophthalmologic examination revealed normal right eye function while eye movement was diminished in the inferior and lateral sides. MRI findings revealed a well-circumscribed heterogeneously enhancing extraconal right orbital lesion suggestive of low-grade sarcoma. Fine needle aspiration cytology was suspicious for malignancy. On the basis of clinical history, physical examination and imaging, it was thought to be a case of sarcoma. The patient underwent wide local excision, grossly it was a grey firm white tumor measuring approximately 4 x 3.5 x 3 cms. Microscopic examination showed spindle to ovoid cells arranged haphazardly and around the dilated blood vessels, forming a haemangiopericytomatous pattern (Fig. 1). immunohistochemistry revealed diffuse and strong positivity to Vimentin (Fig. 2) and CD 34 (Fig. 3), while tumor cells are negative for PanCK, Desmin, EMA, myogenin and CD45. Ki67 proliferative index was 3%. On the basis of histopathological examination and immunohistochemistry, we came to a diagnosis of benign solitary fibrous tumor.

DISCUSSION

SFT is a rare, benign, gradually progressive mesenchymal tumor, the histogenesis of which is poorly understood. It is first described in pleura and thought to be mesothelial in origin.[2] Recently, many cases have been observed occurring in the extrapleural site like the upper airway tract, salivary gland, thyroid gland, lung, mediastinum, pericardium, peritoneum, liver, spine and orbit.[6] The orbital involvement was first described by Dorfman *et al*.,[7] and Westra *et al*.,[8] in 1994. The most common clinical presentation is unilateral, slowly progressive, painless proptosis with well-defined, strongly enhancing mass lesions on CT and MRI.[1] In our case, the same clinical presentation and CT findings were observed. Gupta *et al*.,[6] reported the same case as ours in 39-year-old female representing soft tissue swelling in the intraconal compartment of the right eye.

The typical histopathological feature of SFT is an arrangement of spindle cells in a haphazard manner in

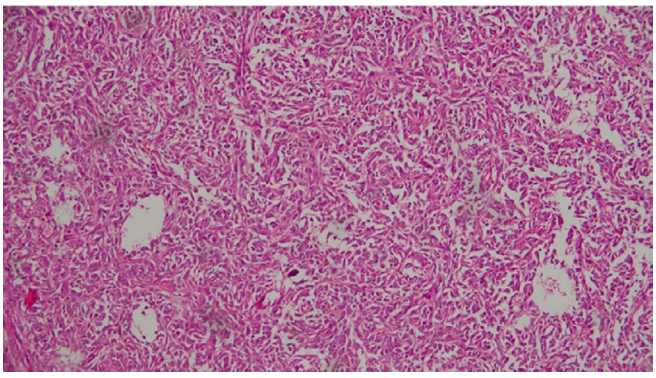

Fig. 1: SFT showing hemangiopericytoma and spindle cells arranged in patternless pattern (H&E, 400X)

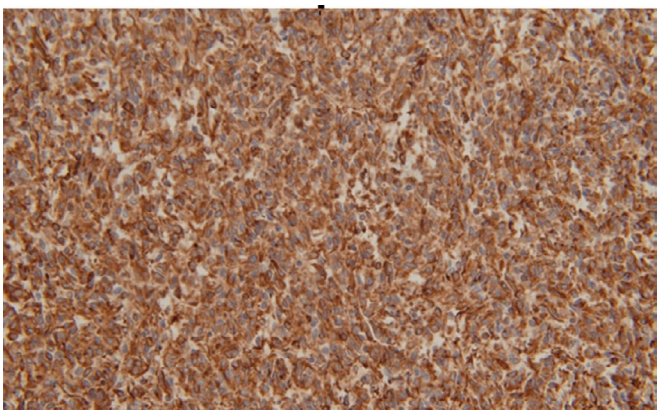

Fig. 2: Strong and diffuse positivity of Vimentin in tumor cells (400X)

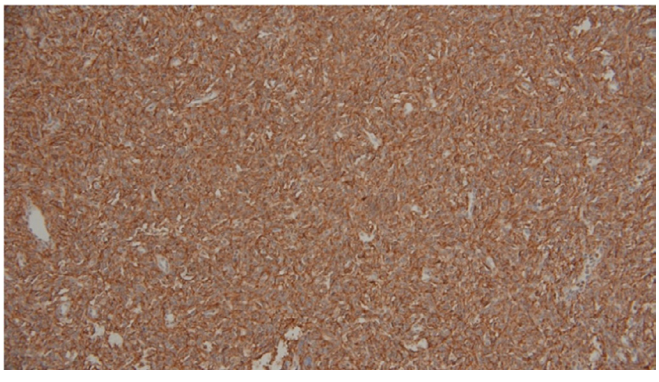

Fig. 3: Strong and diffuse positivity of CD34 in tumor cells (400X)

a cellular stroma consistent with a "patternless pattern." The cells can also arranged in a fascicular and storiform pattern, with thick bands of collagen interspersed between tumor cells and numerous thin-walled vessels forming occasionally staghorn-like channels. Some of the tumor cells are also arranged around the thin capillaries, forming haemangiopericytomatous pattern. [6]

Histopathology is a known gold standard when a definitive diagnosis is required; however, recent advances in the field of immunohistochemistry have further added accuracy in the diagnosis especially when it comes to rare tumors or neoplasms of unusual location. The differential diagnosis of orbital SFT includes a variety of tumors having spindle cell morphology, like fibroblastic tumors, angiofibroma, nerve sheath tumors, meningioma, and synovial sarcoma. The use of ancillary studies like immunohistochemistry and molecular genetics helps to distinguish and categorize such tumors.

Immunohistochemical studies show SFTs have strong and diffuse positivity to CD34, vimentin, bcl2, and focal or nonspecific reactivity to CD99. Recently, the use of immunohistochemistry for STAT6 has been introduced which acts as a surrogate marker for detecting the fusion gene.[4] We found diffuse positivity for Vimentin, CD 34, and focal positivity for CD99. In our case, the Ki67 proliferative index was 3%, while Lim *et al.*,[2] found the Ki67 proliferative index of 10 to 20% in SFT of the parotid gland in 51-year-old man and Ascione *et al.*[1] reported a Ki67 proliferative index of 10% in SFT of the pelvis in an 18-year-old young man.

Although most of the SFTs are benign in nature, some pleural cases have been reported for local invasion and recurrence.[9] Aggressive behavior of the tumor can be appreciated on histologic examination which includes hypercellularity, nuclear pleomorphism, high mitotic rate, necrosis, and high Ki67 proliferative index. Recurrent tumors have been reported to invade surrounding tissue. The most common reason for recurrence is subtotal or incomplete resection or positive margin, so the mainstay of treatment is complete resection of the tumor with long-term follow-up.[10]

CONCLUSION

A solitary fibrous tumor of an orbit is a rare benign neoplasm, generally presenting with painless proptosis of an eye. It should be included in the differential diagnosis of a well-circumscribed enhancing mass. Histopathology is the gold standard, while immunohistochemistry adds further accuracy in the diagnosis of rare tumors or tumors of unusual locations. Thorough histological evaluation with ancillary studies like immunohistochemistry to CD 34 and STAT6 plays a key role in the diagnosis of solitary fibrous tumors and prevent misdiagnosis, which may have significant clinical and therapeutic implications. Although SFT is benign in nature, recurrence is commonly seen if it is not resected properly. So, treatment should include complete resection of the tumor with long-term follow-up.

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