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CASE REPORT

Malignant solitary fibrous tumour of orbit

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ABSTRACT

Solitary fibrous tumor (SFT) is a rare neoplasm that is thought to be of mesenchymal origin. Occurrence of such a tumor in the orbit is rare, more so in its malignant form. Histopathologically, it can mimic several other tumors of the orbit and can be differentiated by CD34 positivity. We report a case of malignant transformation of an SFT of the orbit that recurred after 15 years. The differentiating histopathological features with special stress on the importance of CD34 positivity and principles of management are outlined. The need for long-term follow-up to detect recurrence and malignancy is stressed.

Key words: CD34, malignant, orbit, recurrent, solitary fibrous tumor

Introduction

Solitary fibrous tumor (SFT) of the orbit is a rare lesion, which may have been diagnosed as fibrous histiocytoma, hemangiopericytoma, neurofibroma, or other orbital lesions before the advent of immunohistochemical stains. Since the first report of orbital SFT in 1994, only about 70 cases of orbital SFTs have been reported till date, of which malignant transformation is even rarer.[1] We report a case of malignant orbital SFT in an elderly female who had been operated 15 years back.

Case Report

A 56-year-old female presented with painless progressive ptosis since one year. On examination, the right eyeball was pushed downward, vision was preserved, and both pupils were reacting to light normally. On fundoscopy, no abnormality was detected. The patient was operated 15 years back for progressive ptosis of eight years' duration (right frontal craniotomy and orbitotomy with excision of mass), and the histopathological finding was vascular tumor. No tumor marker studies were done.

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Magnetic resonance imaging (MRI) of the orbit showed intensely enhancing soft tissue lesion in the extraconal space of anterosuperior part of the right orbit involving adjacent orbital roof, resulting in inferior displacement of the right eye globe. The optic nerve was normal in course and optic foramina were symmetrical and normal. As shown in Figures 1-3

Re-exploration of the previous right frontal craniotomy and right lateral orbital osteotomy with complete excision of the mass was done. It was firm, lobulated, and extraconal in location. The postoperative period was uneventful. Vision and extraocular movements were normal. Fundus examination was normal. Partial ptosis was present.

Histopathologically, on gross examination, the tumor was partially encapsulated, gray-brown in color, and firm in consistency. On cross section, there were gray-white to yellow gelatinous areas. Microscopic examination disclosed hypo and hypercellular areas separated from each other by thick bands of hyalinised tissue [Figure 4]. Clusters of round cells with prominent nuclear rim and some prominent nucleoli were seen separated by collagenous stroma. There were foci of calcifications. At places, giant cells were also seen with mitotic activity more than 4 per 10 HPF (HPF: High-power fields), [Figure 5] cytological atypia, and areas of necrosis. Reticulin stain showed individual and small clusters of cells

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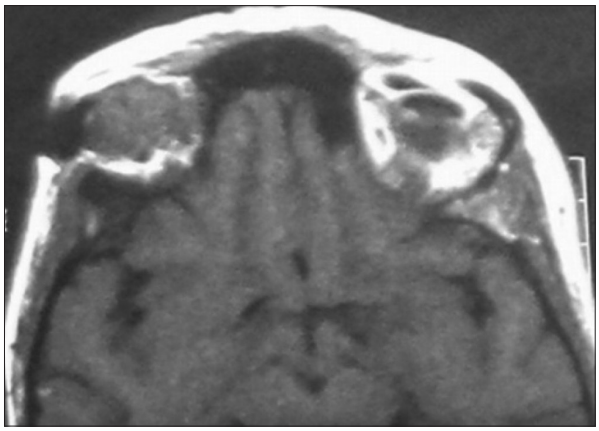

Figure 1: Axial T1 image showing tumor isointense to grey matter

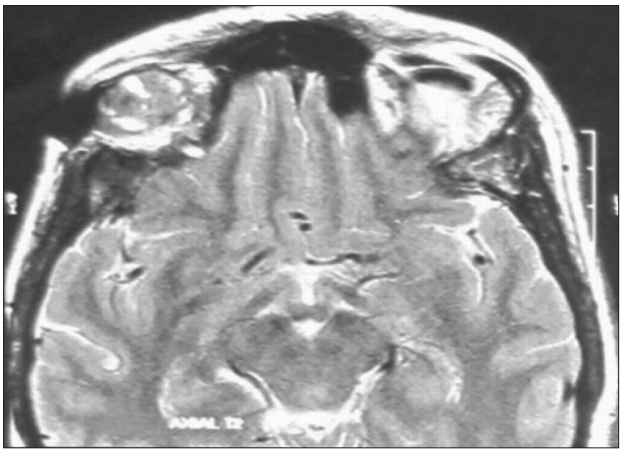

Figure 2: Axial T2 image showing mixed iso to hypointense tumor

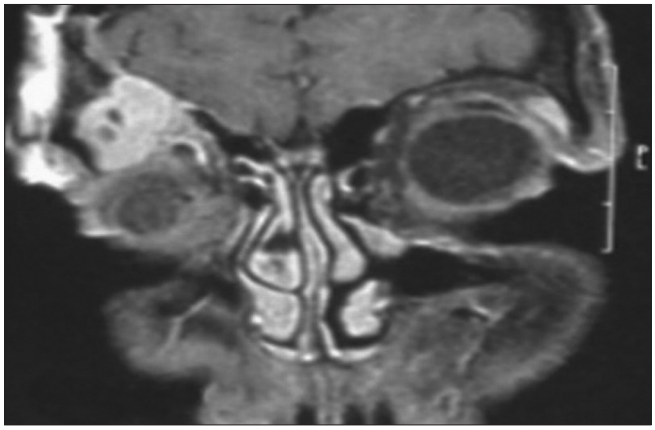

Figure 3: Post-Gd coronal section showing intense enhancement

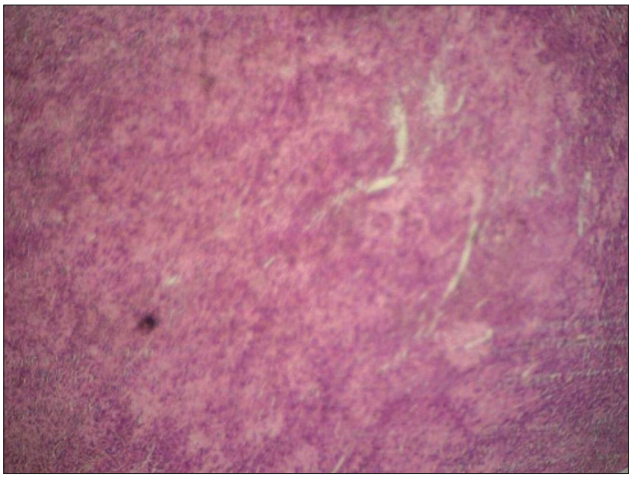

Figure 4: Hematoxylin and eosin stain low-power view hypo and hypercellular area

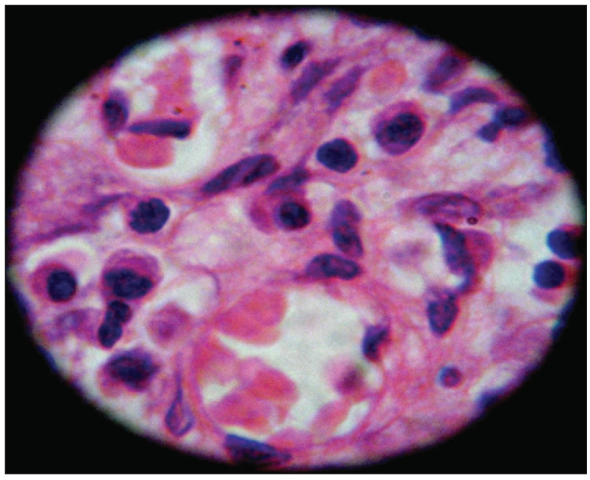

Figure 5: High-power ×40 view, nuclear pleomorphism and mitotic activity with two small blood vessels

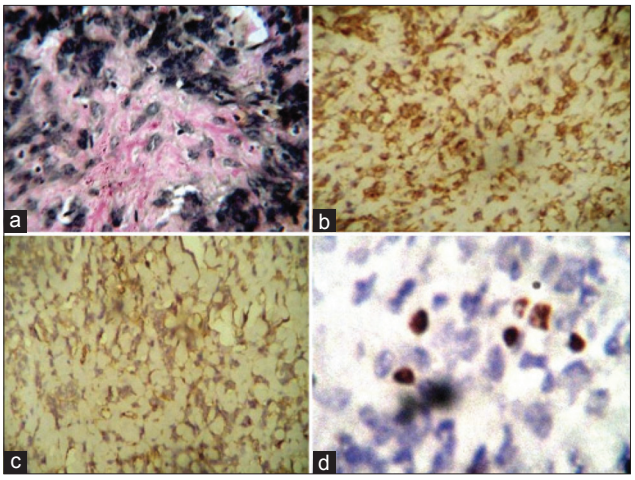

Figure 6: (a) Van Gieson stain collagens stain red (b) CD34 ×10 strong and diffused positivity (c) Bcl2 ×10 diffuse and strong positivity (d) Ki-67 ×40 high power, showing positivity

separated by reticulin. Van Gieson staining showed foci of collagen [Figure 6a]. Small to large blood vessels were also seen [Figure 6b]. Marker study showed diffuse and strong positivity for CD34 and Bcl2 and negativity for smooth muscle actin (SMA) [Figure 6c], pan-cytokeratin, S100, and epithelial membrane antigen (EMA) [Figure 6d]. KI-67 MIB index positivity in hypocellular areas was approximately 5-10% and 10-15% in

hypercellular areas. A final diagnosis of malignant extrapleural SFT of the orbit was made.

Discussion

SFT is a rare, but distinct spindle cell neoplasm that was initially described in the pleura by Klemperer and Rabin in 1931; thereafter, this tumor has been documented in almost every anatomic location[2] with both benign and malignant variants identified.[3]

Histogenesis of this neoplasm, originally thought to be of mesothelial origin, is now considered to be of mesenchymal, and possibly of fibroblastic origin.[4] The clinical behavior is variable. Most SFTs are benign but local invasion or recurrence of the lesion has been demonstrated.

The MRI findings of SFT describe the mass lesion as isointense or hypointense to gray matter in a T1-weighted image.[5,6] In a T2-weighted image, the lesion shows hypointensity, hyperintensity, or variegated intensity.[6] When gadolinium is used as contrast, the lesion shows homogeneous enhancement[5,6] or variable enhancement with central areas of less enhancement. The variable findings of MRI are considered to be due to the diversified pathological components of the tumor including cellularity, rich collagenous fibers, and vascularity in the hemangiopericytoma-like pattern.

These radiological features are common to other orbital neoplasms such as hemangiopericytoma, schwannoma, or meningioma,[7] further emphasizing the need for definitive histopathological diagnosis.

It has been reported that a small percentage of SFTs of the pleura and mediastinum have aggressive features such as tumor necrosis and large tumor size, as well as hypercellularity, high mitotic count (4 per 10 HPF), and cellular pleomorphism. SFTs with these aggressive features may manifest with invasion of adjacent tissue, recurrence, or distant metastases.[8]

SFTs can mimic other spindle cell tumors of the orbit, which include fibrous histiocytoma, meningioma, schwanoma, and hemangiopericytoma. In view of the varied and potentially confusing histologic appearance of the tumor, it is possible that previous, unrecognized cases of orbital SFTs may have been classified under one of these diagnostic headings. CD34 immunoreactivity has been found to be a highly sensitive marker for SFTs.[4] All of the SFTs demonstrated strong cytoplasmic staining with a monoclonal antibody to CD34. CD34 is an antigen expressed on the surface of the vascular endothelium and hematopoietic progenitor cells.[9] Intense CD34 immunohistochemical staining helps to differentiate SFTs from hemangiopericytoma, which shows weak and patchy CD34 staining, and also from fibrous histiocytoma, which shows CD34 negativity. Schwanoma is also CD34 positive, but it is very focal, and it will be strongly positive for neural markers such as the S100 protein.

Treatment of an SFT of the orbit consists of complete surgical excision with long-term follow-up. Various surgical approaches are available for resecting orbital tumors: The fronto-orbital approach, the pterional approach, lateral orbitotomy, and medial orbitotomy. Lesions with intracranial extension, involving the optic canal or medial to the optic nerve in the apex, are addressed via the transcranial fronto-orbital approach.[10] A long-term follow-up is requisite because recurrences may occur several years after the excision of the primary tumor and the efficacy of either radiotherapy or chemotherapy in the management of residual SFT remains unclear.

Conclusion

SFT of the orbit is a rare tumor that can present in a malignant variant that can recur after many years. Awareness is needed among surgeons about this entity and about the possibility of malignancy in it. Differentiation from other similar orbital tumors such as schwannoma, hemangiopericytoma, meningioma, and fibrous histiocytoma can be challenging. Histopathological analysis with immunohistochemical staining for CD 34 is the key to differentiate it from other orbital tumors. More than four mitoses per HPF is a criterion for diagnosis of malignancy. Long-term follow-up is mandatory.

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Conflicts of interest
There are no conflicts of interest.

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