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Fat-forming solitary fibrous tumor of the orbit with typical imaging findings

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ABSTRACT

Purpose: We describe a case of fat-forming solitary fibrous tumor (SFT) of the orbit with typical findings on imaging that may improve the awareness of orbital fat-forming SFT.

Observations: An 88-year-old female presented with exophthalmos and pain in her right eye. Preoperative imaging showed an oval, well-defined mass with soft-tissue density, interspersed with a well-circumscribed lesion. The lesion showed low-density in computed tomography (CT) scans, hyperintense in T1/T2 weighted images of magnetic resonance imaging (MRI) scans and hypointense in fat-suppressed images of MRI scans. The tumor was removed en bloc and diagnosed as low-grade malignant fat-forming SFT by pathological examination. There was no evidence of recurrence 9-month postoperatively.

Conclusions: The imaging feature of orbital fat-forming SFT is a well-defined solid tumor interspersed with adipose tissue. Such findings are vital for the preoperative diagnosis and the choice of the treatment.

1. Introduction

Fat-forming solitary fibrous tumor (SFT) is commonly found in the pleura and thigh.[1] However, the occurrence of orbital fat-forming SFT is exceedingly rare, with only three documented cases in the literature.[2–4] The limited understanding of orbital fat-forming SFT is primarily due to the absence of comprehensive clinical and radiographic data. This study presents a case with typical imaging characteristics, aiming to enhance the accuracy of diagnosis and treatment for this particular disease.

2. Case report

An 88-year-old female patient suffered from exophthalmos in her right eye for approximately 20 years (Fig. 1A), with pain intensifying over the last year. The patient had not undergone any specific treatment previously and had experienced vision loss five years prior. The eye movements of the right eye were restricted in all directions. An immobile and slightly tender hard mass was detected in the supraorbital areas, resulting in the subocular displacement. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a substantial isodense or isointense mass, interspersed with a well-circumscribed lesion (Fig. 1B and C). The lesion showed low-density in CT scans (Fig. 1B), hyperintense in T1/T2 weighted images of MRI scans (Fig. 1C and D) and hypointense in fat-suppressed images of MRI scans (Fig. 1E). Additionally, a non-enhancing necrotic region with irregular and indistinct boundary was observed (Fig. 1F).

The patient underwent an anterior orbitotomy via a *trans*-eyebrow approach, which revealed an oval-shaped, enveloped and well-circumscribed mass. The tumor was removed en bloc and measured 5 × 4 × 4 cm (Fig. 2A). In that case, we did not perform extra frozen sections perioperatively. Upon cross-sectioning, the specimen showed signs of tissue hemorrhage and necrosis (Fig. 2B). Hematoxylin-eosin (HE) staining identified oval and spindle tumor cells. The tumor cells were in a hypercellular or hypocellular distribution in different areas (Fig. 2C). Besides, there are some staghorn vessels decentralized in the tumor which characterized by ordinary SFT. The tumor also contained clusters of mature adipocytes (Fig. 2D) and necrotic tissues (Fig. 2E). Immunohistochemical analysis showed that the tumor was negative for MDM2 and CDK4, positivity for CD34, BCL2, CD99, and STAT6 (Fig. 2F). The positive rate of Ki67 was 10 % and some of tumor cells grew actively with nuclear atypia. Based on these pathological results, the patient was diagnosed with low-grade malignant fat-forming SFT. At last follow-up 9 months later, the patient's right eye showed no obvious exophthalmos. And there was no well-defined tumor-like lesion on CT findings. All the data showed no recurrence of the patient.

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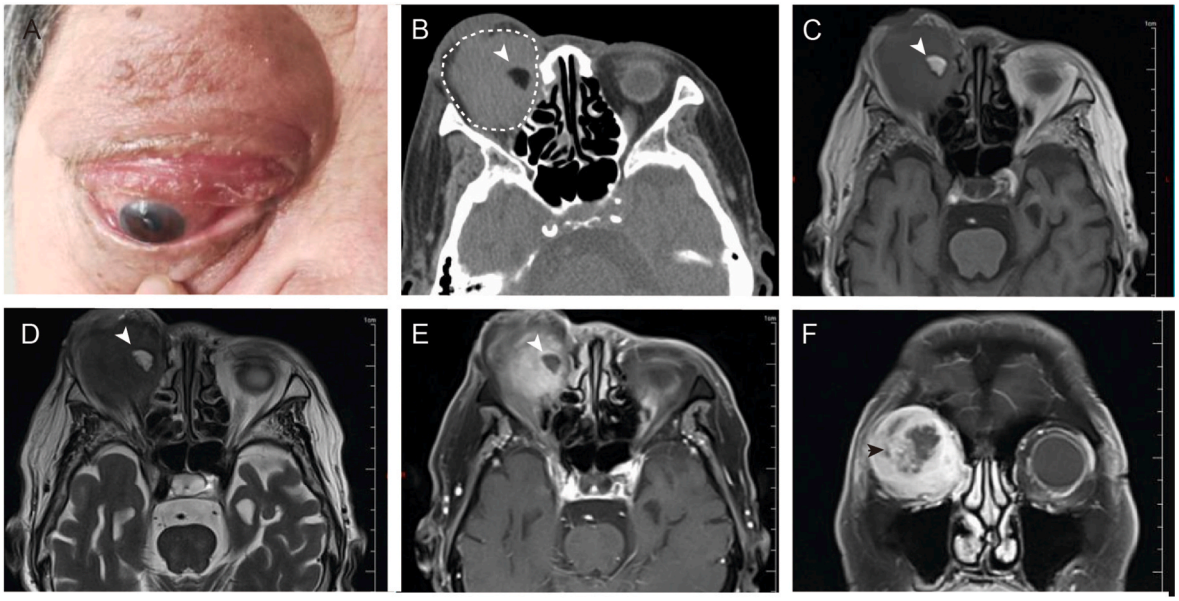

Fig. 1. A) Clinical photograph of the patient. B) CT scan depicted a substantial mass of soft-tissue density, intermingled with a well-circumscribed hypointense lesion. *C*-E) Magnetic resonance imaging (MRI) indicated the presence of adipose tissue within the tumor. F) MRI finding demonstrated a necrotic lesion within the tumor.

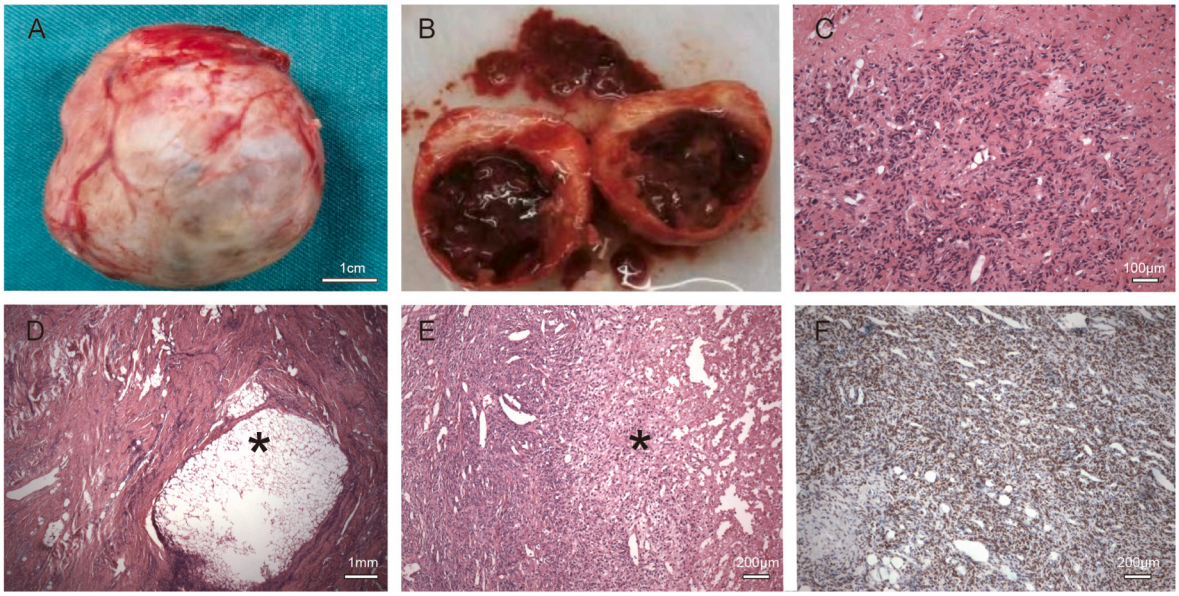

Fig. 2. Gross appearance and histopathological examination. A) In gross examination, the lesion is well circumscribed with a glistering surface. B) The sectioned tumor specimen showed extensive necrosis and cystic degeneration. C) Hematoxylin-eosin (HE) staining showed the fields of dense tumor cells and sparse cells arranged within a collagenous, vessel-rich stroma. D-E) In low magnification, mature adipose tissue and a necrotic tissue are indicated by the asterisks. F) Immunohistochemical test showed the tumor was positive for STAT6.

3. Discussion

Similar to previous reports,[2–4] the principal manifestations of orbital fat-forming SFT predominantly resulted from the mass effect. The patient presented with ptosis, pain, and visual impairment attributed to tumor compression. However, these symptoms were nonspecific and not enough to make a diagnosis.

To our knowledge, our study for the first time showed the detailed preoperative imaging characteristics of orbital fat-forming SFT. Firstly, the lesion demonstrated characteristic imaging features of orbital SFT. Specifically, it presented as an oval, well-defined mass with soft-tissue density which exhibited significant enhancement on contrast-enhanced MRI images. It also did not display evident bone destruction on CT scans.[5] Meanwhile, the tumor also exhibited imaging features of adipose tissue which showed hyperintense on T1/T2 weighted images and hypointense on fat-suppressed images of MRI scans. These imaging features played a crucial role in preoperative diagnosis. However, in previous cases, preoperative imaging failed to identify characteristic adipose tissue, despite subsequent histopathological analysis confirming the presence of adipose tissue.[2–4] Therefore, the absence of adipose tissue imaging does not exclude a fat-forming SFT diagnosis. This discrepancy might relate to patient-specific disease progression and size of adipose tissue. For our patient, the disease duration lasted for up to 20 years, while the duration of other cases is less than 3 years.[2–4] Only in our case was there a large adipose tissue sized 5 mm, while the other cases consisted of scattered adipose cells.

Furthermore, we identified necrotic tissue in the orbital fat-forming SFT through imaging and histopathology examinations. However, there is no necrosis noted in the three previously reported cases of orbital fat-forming SFTs. Only one case of the thigh fat-forming SFT exhibited necrosis and hemorrhage and showed no evidence of recurrence or metastasis 14 months postoperatively.[6] Notably, fat-forming SFT can exhibit a favorable post-operative prognosis compared to that of SFT.

4. Conclusions

Orbital fat-forming SFT exhibits a protracted duration and gradual progression. The presence of a solid tumor with concurrent fatty signal on CT or MRI scans holds substantial diagnostic significance. Complete surgical resection yields a promising prognosis.

Patient consent

The subject has given her written informed consent to publish her case (including publication of images).

CRediT authorship contribution statement

Lan Yao: Resources, Writing – original draft. **Xinji Yang:** Writing – review & editing. **Wei Wu:** Funding acquisition, Writing – review & editing.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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References

1. Tariq MU, Din NU, Abdul-Ghafar J, Park YK. The many faces of solitary fibrous tumor; diversity of histological features, differential diagnosis and role of molecular studies and surrogate markers in avoiding misdiagnosis and predicting the behavior. *Diagn Pathol*. 2021;16(1):32.
2. Guillou L, Gebhard S, Coindre JM. Lipomatous hemangiopericytoma: a fat-containing variant of solitary fibrous tumor? Clinicopathologic, immunohistochemical, and ultrastructural analysis of a series in favor of a unifying concept. *Hum Pathol*. 2000;31(9):1108–1115.
3. Davies PE, Davis GJ, Dodd T, Selva D. Orbital lipomatous haemangiopericytoma: an unusual variant. *Clin Exp Ophthalmol*. 2002;30(4):281–283.
4. Pitchamuthu H, Gonzalez P, Kyle P, Roberts F. Fat-forming variant of solitary fibrous tumour of the orbit: the entity previously known as lipomatous haemangiopericytoma. *Eye (Lond)*. 2009;23(6):1479–1481.
5. Yang BT, Wang YZ, Dong JY, Wang XY, Wang ZC. MRI study of solitary fibrous tumor in the orbit. *AJR Am J Roentgenol*. 2012;199(4):W506–W511.
6. Noh SJ, Jang KY. Malignant fat-forming solitary fibrous tumor of the thigh. *Korean J. Pathol.* 2014;48(1):69–72.