Pathol. Oncol. Res. (2008) 14:213–217
DOI 10.1007/s12253-008-9055-7

BRIEF COMMUNICATION

Orbital Solitary Fibrous Tumor: A Case Report and Review of the Literature

Giuseppe Leoncini · Vincenza Maio · Marco Puccioni · Alessandro Franchi · Vincenzo De Giorgi · Francesca Ucci · Marco Santucci · Daniela Massi

Received: 12 April 2008 / Accepted: 17 April 2008 / Published online: 21 May 2008

Abstract Solitary fibrous tumor (SFT) is a rare spindle cell neoplasm typically arising in the pleura and involving the orbit as its most common extra-pleural location. We herein describe a well documented case of orbital SFT arising in a 62-year-old woman presenting with progressive swelling of the right upper eyelid and proptosis. The tumor had a benign clinical course, with radical surgical excision followed by regression of the clinical symptoms. We review the clinical, histopathological, and immunohistochemical features of the orbital SFT described so far, with particular emphasis on differential diagnosis with other spindle cell orbital neoplasms.

Keywords Solitary fibrous tumor · Orbit

Introduction

Although originally described in the pleura [1–2], solitary fibrous tumor (SFT) has been reported at several extra-pleural sites, including peritoneum [3], liver [4], mediastinum [5], upper respiratory tract [6], kidney [7], adrenal gland [8], urinary bladder and prostate [9], soft tissues [10], skin [11], nasal cavity, and paranasal sinuses [12]. The soft tissues of the orbit, yet another site unrelated to mesothelium, have also been reported as an additional extrapleural location. In the past years, orbital SFT has been underdiagnosed, due to unfamiliarity with its clinical presentation, variable histopathological profile overlapping with other spindle cell neoplasms, as well as inconsistencies regarding nomenclature and histogenesis [13–15].

We herein describe a case of orbital SFT arising in a 62-year-old woman presenting with progressive swelling of the right upper eyelid and proptosis and pursuing a benign clinical course. We review the clinical, histopathological and immunohistochemical features of orbital SFT described so far, with particular emphasis on differential diagnosis with other spindle cell orbital neoplasms.

Case Report

A 62-year-old woman presented with a slowly arising swelling of the right upper eyelid and proptosis. She complained no pain or vision changes, she had no history of glaucoma and intraocular pressure was normal in both eyes. Ocular movement was normal and pupils were symmetric, round and reactive to light. A computed tomography (CT) scan revealed a solid extra-conal mass that measured 1.4×3×4 cm adhering to the lateral wall of the right orbit and to the orbital roof, without any signs of osteolysis, calcifications or bone infiltration. The intravenous injection of gadolinium diethylenetriamine pentaacetic acid (Gd-DTPA) showed a homogeneous enhancement of the tumor mass (Fig. 1a). The tumor was surgically excised by antero-lateral approach. No adherences of optic nerve or involvement of other adjacent structures occurred. Postoper-

G. Leoncini · V. Maio · A. Franchi · M. Santucci · D. Massi (✉)
Department of Human Pathology and Oncology,
University of Florence,
Viale GB Morgagni 85,
50134 Florence, Italy
e-mail: Daniela.massi@unifi.it

M. Puccioni · F. Ucci
Department of Ophthalmology, University of Florence,
Florence, Italy

V. De Giorgi
Department of Dermatological Sciences, University of Florence,
Florence, Italy

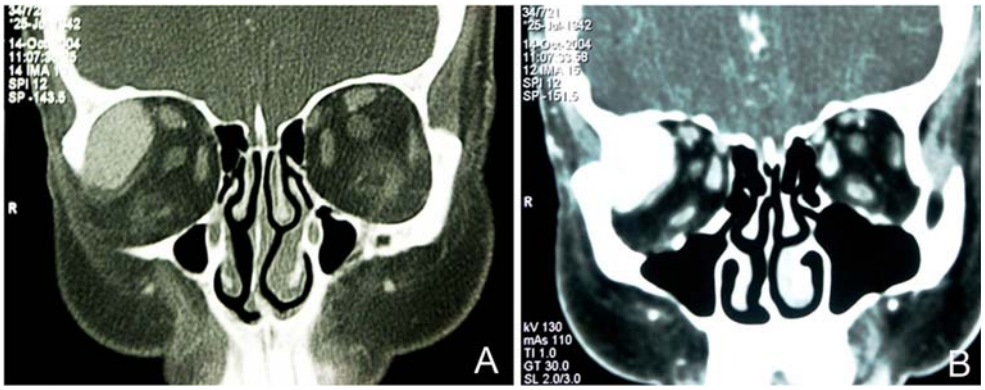

Fig. 1 Computer tomography scan showing a well-circumscribed mass, pre- (**a**) and post-enhancement (**b**) after intravenous injection of Gadolinium diethylenetriamine pentaacetic acid (Gd-DTPA)

ative course was uneventful, the patient is alive and well after 2-years follow-up.

Macroscopically, the tumor had hard-elastic consistence and reddish surface (Fig. 2a). Histopathological examination showed a dense proliferation of spindle cells with variable architectural pattern (Fig. 2b). There were areas with fascicled pattern and areas with random and "pattern-less" cell arrangement. Spindle cells showed a uniform, oval, vesicular nucleus and finely dispersed chromatin. Sclerotic areas with dense bundles of collagen and clefts between collagen and tumor cells were also demonstrated. By immunohistochemistry, tumor cells were CD34, CD99, and bcl-2 strongly and diffusely positive (Fig. 2b), whereas S-100 protein and CD45 were negative.

Discussion

Orbital SFT occurs over a wide age range (from 9 to 76 years; mean 42.5), male–female ratio amounts roughly to 1:1. It always presents as an extra-conal mass, although the intra-conal location was also been described [16], it equally involves both the right and the left orbit where it can be localized anywhere, including the lacrimal gland fossa [17, 18], with a mild predilection for the medial half of the orbit. During its slow development, the tumor may be associated with deficit of the oculomotion and optic nerve. The tumor often leads to eyelid swelling and progressive painless proptosis, till dislocation of the ocular globe. Vision and ophthalmic examination are generally unremarkable, with the exception of cases associated with a pronounced globe dislocation (approximately 20%). Space-occupying symptoms such as orbital proptosis may be present.

Like the intra-thoracic lesions, the clinical course of extra-thoracic SFTs is unpredictable and a long term follow-up is mandatory. Although the biological behaviour is commonly benign, the excision being curative, some cases have the potential for local relapse or distant spread. By review of the literature, it is apparent that eight cases were associated with an aggressive clinical course. In particular, Polito et al. [19] describe three cases of orbital SFT with aggressive behaviour (tendency to osteolysis) and recurrence 4 years after the first-look surgery. Hayashy et al. [20] report a case of orbital SFT extended to extra-dural middle cranial fossa and cavernous sinus. Carrera et al. [21] refer about a case of orbital SFT with two local recurrences after surgical excision.

As reported by Vallat-Decouvelaere et al. [22], features predictive of malignant clinical behaviour in intra-thoracic SFTs can be equally applied to extra-thoracic SFTs. Histopathological findings associated with local relapse or distant spread include hypercellularity and high mitotic rate (mitoses>4/10 HPF), cellular and nuclear pleomorphism, foci of necrosis and high MIB-1 labeling [5, 13]. Indeed, recurrent orbital SFTs showed increased cellularity and high mitotic rate; however areas of necrosis were usually absent and tumors had a low MIB-1 labeling (<10%) [22]. It is worth of mention that one metastatic tumor included in Hasegawa et al. [23] (case 20) did not show any atypical features in either the primary or metastatic lesion.

The classical histopathological features of SFT include a proliferation of spindle cells with a widely varying architectural pattern, alternating hypercellular (tumor cell-rich) and hypocellular (collagen-rich) areas with a haemangiopericytoma-like pattern. The presence of striking areas of hyalinization and the artifactual cracks are suggestive of SFT [24, 25] In addition synovial sarcoma-like areas, storiform and fascicular formation, and neural-type palisades have also been described [26].

It is of the greatest importance to differentiate orbital SFT from other spindle cell neoplasms that can occur in this location (Table 1). Malignant peripheral nerve sheath tumor shows focal positivity for S-100 protein and EMA, both negative in SFT, whereas monophasic synovial sarcoma stains for EMA and keratins. Fibrosarcoma rarely involves the orbit and a herringbone pattern of cells is uncommon in SFT. Regarding leiomyosarcomas, CD34 is negative in the large majority of cases and spindle cells stains for α-SMA, suggesting smooth muscle origin. In the past literature, the

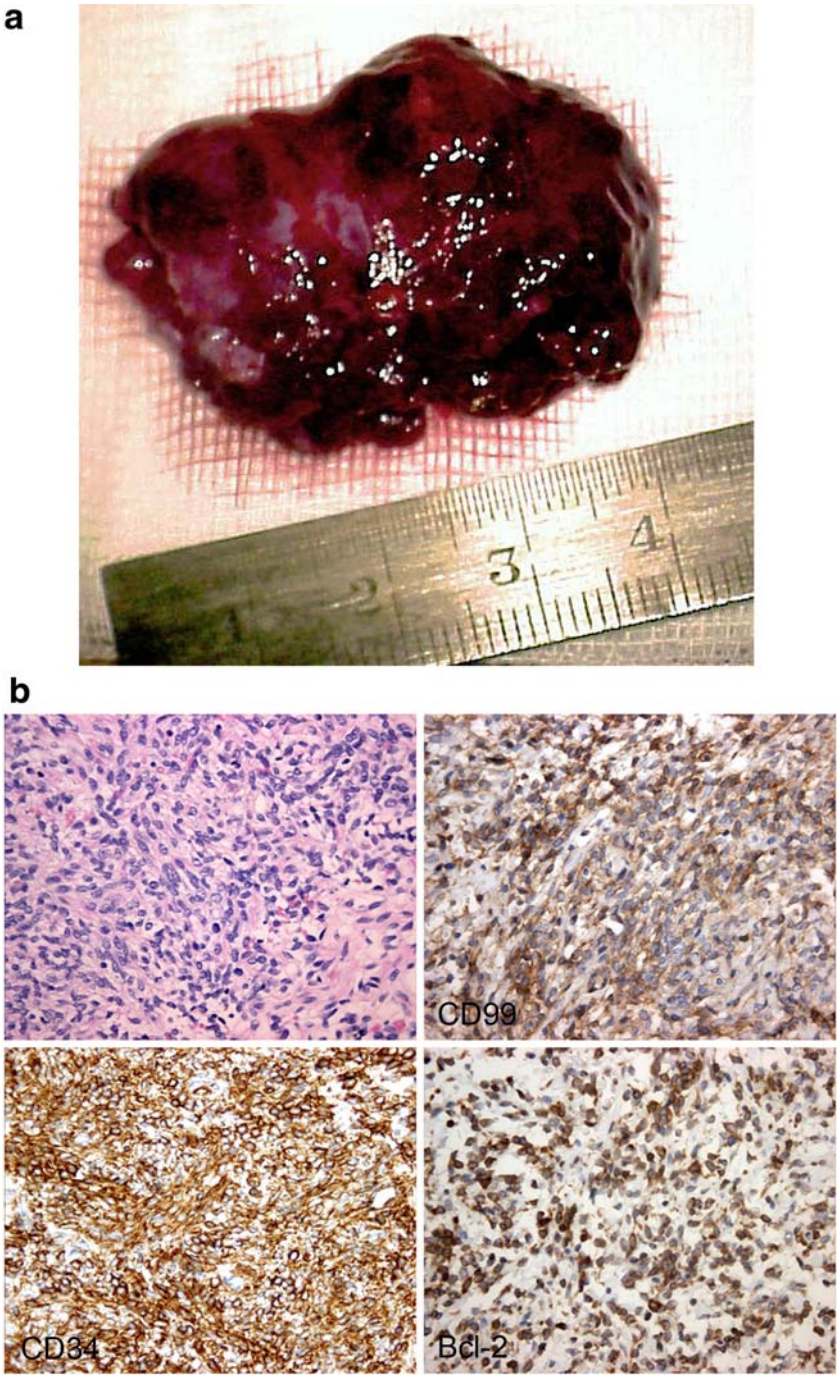

Fig. 2 **a** Macroscopically the tumor mass displays a reddish surface and a hard-elastic consistence. **b** Histopathological examination shows spindle-shaped cells in variable arrangement. The tumor shows a strong and diffuse positivity for Bcl-2, CD34 and CD99

differential diagnosis also included malignant fibrous histiocytoma, haemangiopericytoma, giant cell angiofibroma and giant cell fibroblastoma. At present, however, those cases classified in the past as malignant fibrous histiocytomas have been reclassified as other types of sarcomas while haemangiopericytoma, giant cell angiofibroma and giant cell fibroblastoma present overlapping features with SFT and are considered in the same spectrum of diseases.

The use of immunohistochemical markers is extremely helpful in the diagnosis of orbital SFT [14]. SFT shows a strong and diffuse positivity for vimentin, CD34 and Bcl-2, whereas it is negative for cytokeratins, EMA, S-100 protein, α-SMA and desmin. Recently, CD99 (O-13) has been found to be a reliable marker for primary and recurrent SFT. Interestingly, in contrast with dedifferentiated mesenchymal tumors, SFT retains in the recurrences the same

Table 1 Differential diagnosis of orbital SFT

Neoplasm	Age	Involvement	Histopathological features	Immunophenotype	
Malignant peripheral nerve sheath tumor	Childhood	Limbs	Sweeping fascicles of spindle cells, occasionally arranged in whorls around the vessels	Vimentin	+
				CD34	+
				Bcl-2	+
	Adulthood			CK	±
				EMA	±
				S-100	±
Monophasic synovial sarcoma	Adulthood	Limbs Neck	Monotonous growth of spindle cells, organized in dense fascicles	Vimentin	±
				CD34	-
				Bcl-2	+
				CK	+
				EMA	+
				S-100	±
Fibrosarcoma	Childhood	Trunk Limbs	Spindle cells in sweeping fascicles. Herringbone pattern; scanty cytoplasm, prominent nucleoli	Vimentin	+
				CD34	+
				Bcl-2	+
	Adulthood	Head & Neck		SMA	±
				CK	-
Leiomyosarcoma	Childhood	Retroperitoneum	Intersecting groups of spindle cells with cigar-shaped nuclei; storiform/palisaded/haemangiopericytoma-like pattern; fibrosis/myxoid change	SMA	+
				Desmin	+
		Large blood vessels		Vimentin	+
	Adulthood			CK	+
				CD34	-
		Limbs		Bcl-2	-

± Focal positivity

immunohistochemical profile as the primary lesion, with strong and diffuse CD34 and CD99 positivity [22].

References

1. De Saint Aubain Somerhausen N, Rubin BP, Fletcher CDM (1999) Mixoid solitary fibrous tumor: a study of seven cases with emphasis on differential diagnosis. Mod Pathol 12:463–471
2. Klemper C, Rabin CB (1931) Primary neoplasm of the pleura; a report of five cases. Arch Pathol 11:385–412
3. El-Naggar AK, Ro JY, Ayala AG et al (1989) Localized fibrous tumor of the serosal cavities. Am J Clin Pathol 13:199–209
4. Kottke-Marchant K, Hart WR, Broughan T (1989) Localized fibrous tumor (localized fibrous mesothelioma) of the liver. Cancer 64:1096–1102
5. Witkin GB, Rosai J (1989) Solitary fibrous tumor of the mediastinum. Am J Surg Pathol 13:547–557
6. Witkin GB, Rosai J (1991) Solitary fibrous tumor of the upper respiratory tract. Am J Surg Pathol 15:842–848
7. Fukunaga M, Nikaido T (1997) Solitary fibrous tumor of the renal peri-pelvis. Histopathology 30:451–456
8. Prevot S, Penna C, Imbert JC et al (1996) Solitary fibrous tumor of the adrenal gland. Mod Pathol 9:1170–1174
9. Fisher C, Bisceglia M (1994) Solitary fibrous tumor of the spermatic cord. Br J Urol 74:798–789
10. Hasegawa T, Hirose T, Seki K et al (1996) Solitary fibrous tumor of the soft tissue. Am J Clin Pathol 106:325–331
11. Cowper SE, Kilpatrick T, Proper S et al (1999) Solitary fibrous tumor of the skin. Am J Dermatopathol 21(3):213–219
12. Zuckerberg ML, Rosemberg AE, Randolph G et al (1991) Solitary fibrous tumor of the nasal cavity and paranasal sinuses. Am J Surg Pathol 15:126–130
13. England DM, Hochholzer L, McCarthy MJ (1989) Localized benign and malignant fibrous tumors of the pleura: a clinicopathologic review of 223 cases. Am J Surg Pathol 13:640–658
14. Chan JKC (1997) Solitary fibrous tumor—everywhere and a diagnosis in vogue. Histopathology 31:568–576
15. Frederick A Font J, Font RL (1986) Orbit. In: Spencer WH (ed) Ophthalmic pathology: an atlas and textbook, 3rd edn. Saunders, Philadelphia, pp 2459–2860
16. Festa S, Lee HJ, Langer P et al (1999) Solitary fibrous tumor of the orbit: CT and pathologic correlation. Neuroradiology 41:52–54
17. Scott IU, Tanenbaum M, Rubin D et al (1996) Solitary fibrous tumor of the lacrimal gland fossa. Ophthalmology 103:1613–1618
18. Woo KI, Suh YL, Kim YD (1999) Solitary fibrous tumor of the lacrimal sac. Ophthal Plast Reconstr Surg 15(6):450–453
19. Polito E, Tosi M, Toti P et al (2002) Orbital solitary fibrous tumor with aggressive behaviourThree cases and review of the literature. Graefes Arch Clin Exp Ophthalmol 240(7):570–574
20. Hayashi S, Kurihara H, Hirato J et al (2001) Solitary fibrous tumor of the orbit with an extraorbital extension: case report. Neurosurgery 49(5):1241–1245
21. Carrera M, Prat J, Quintana M (2001) Malignant solitary fibrous tumor of the orbit: report of a case with 8 years follow-up. Eye 15:102–104
22. Vallat-Decouvelaere AV, Dry SM, Fletcher CDM (1998) Atypical and malignant solitary fibrous tumors in extra-thoracic locations.

Evidence of their comparability to intra-thoracic tumors. Am J Surg Pathol 22:1501–1511
23. Hasegawa T, Matsuno Y, Shimoda T et al (1999) Extrathoracic solitary fibrous tumors: their histological variability and potentially aggressive behaviour. Hum Pathol 30:1464–1473
24. Enzinger FM, Weiss SW (1997) Soft tissue tumors, 4th edn. Mosby, USA
25. Moran CA, Suster S, Koss MN (1992) The spectrum of histologic growth patterns in benign and malignant fibrous tumors of the pleura. Sem Diagn Pathol 9:169–180
26. Nielsen GP, O'Connell JX, Dickersin GR et al (1997) Solitary fibrous tumor of soft tissue. A report of 15 cases, including 5 malignant examples with light microscopic, immunohistochemical, and ultrastructural data. Mod Pathol 10:1028–1037