B-ENT, 2006, **2**, 201-204

Solitary fibrous tumour of the soft tissue of the face: a case report

S. Schmitz, B. Weynand**, B. Lengelé*** and M. Hamoir**

*Department of Oto-Rhino-Laryngology and Head and Neck Surgery, **Department of Pathology, ***Department of Plastic and Reconstructive Surgery, St-Luc University Hospital, Brussels, Belgium

Key-words. Fibrous tumour; face; soft tissue

Abstract. *Solitary fibrous tumour of the soft tissue of the face: a case report. Introduction*: Solitary fibrous tumour (SFT) is a rare mesenchymal neoplasm described first in the pleura. Recently, SFTs have been found in various extraserosal locations, including the head and neck region.
Case presentation: We report a case of SFT originating from the periorbital region of the face.
Preoperative cytological examination by fine needle aspiration biopsy diagnosed a mesenchymal tumour. The patient underwent surgical resection. The mass was completely resected. Definitive histostopathologic and immunohistologic examination confirmed the diagnosis of SFT.
Discussion: The rare localisation in extrapleural sites and the multiplicity of histological patterns can explain the difficulty in arriving at a definitive diagnosis in SFT. Usually, SFT is a benign tumour, although malignant variants exist. Clinical behaviour is unpredictable and recurrence or malignant transformation can also occur, especially in cases with macroscopically or microscopically invaded margins. Complete surgical excision and long follow-up is therefore always recommended. The recent increase in reports of extrapleural SFT indicates that this rare tumour should be included in the differential diagnosis of soft tissue head and neck tumours.

Introduction

Solitary fibrous tumour (SFT) is a spindle cell neoplasm, which was first described in 1931 by Klemperer and Rabin as a localised fibrous mesothelioma of the pleura.[1] Nevertheless, recent publications have also reported this tumour in extra-pleural sites and areas remote from serosal surfaces. We report the case of a SFT arising from the periorbital soft tissue in a female adult.

Case presentation

A 41-year-old female was referred to our department with a 4-month history of a slowly growing mass of the left internal periorbital region (Figure 1). The patient had no significant medical past history. The mass was bilobated, firm, and well-demarcated, and measured about 2×1 cm. Magnetic resonance imaging (MRI) described a well-defined, low-signal mass on T2-weighted images developed in the soft tissue without bone destruction. Fine needle aspiration biopsy (FNAB) was performed, resulting in the diagnosis of mesenchymal tumour. The tumour was then removed through a lateral rhinotomy approach without complications. The mass was well-circumscribed and relatively easily removed in totality, including the surrounding skin intimately adherent to the tumour (Figure 2).

Histopathological analysis revealed a pale and firm mass that measured $3.5 \times 1.3 \times 1.5$ cm, displaying interlacing fascicles of fusiform cells. Vascularisation was prominent, with branching vascular channels virtually indistinguishable from those usually reported in haemangiopericytoma (Figure 3). Some areas appeared to have a high cellularity and a high mitotic activity (> 4/10 HPF). The cells stained positively for CD-34 and vimentin but were negative for S-100, AE1/AE3, NSE, actine and desmin, confirming the diagnosis of SFT. The postoperative course was uneventful and close postoperative follow-up was proposed. There was no clinical or radiological evidence of tumour recurrence six months after surgery.

Discussion

SFTs are known to occur predominantly in the visceral pleura of the lung and are now considered to be of mesothelial origin.[2] After considerable controversies about the precise aetiology of this tumour, recent publications acknowledge that SFTs originate from mesenchymal cells of submesothelial tissue.[3-5] Recently, SFTs have been described in a variety of extrapleural localisations such as the head and neck region, including

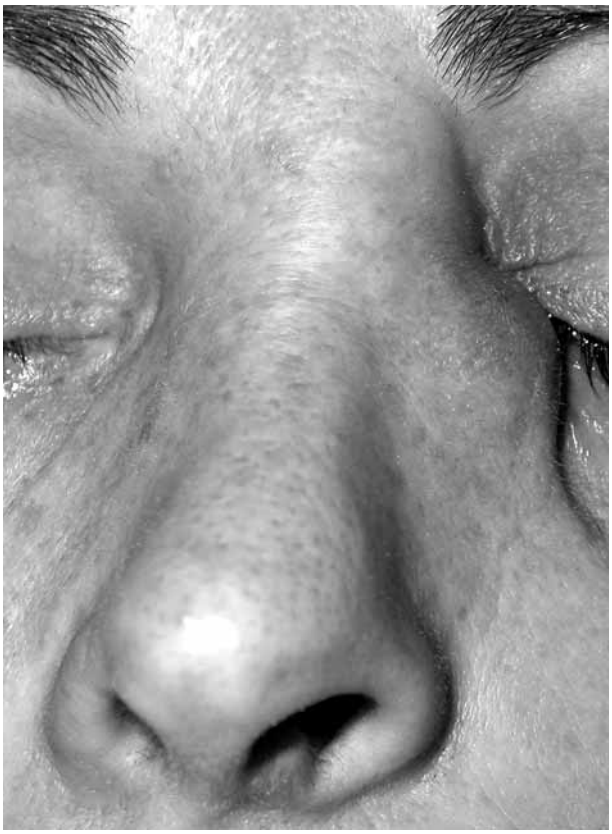

Figure 1
Clinical photograph of the tumour

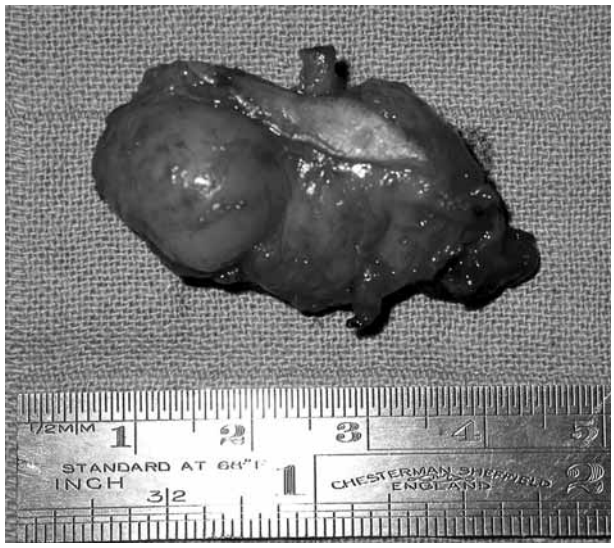

Figure 2
The lesion presents a caracteristic grey-white aspect

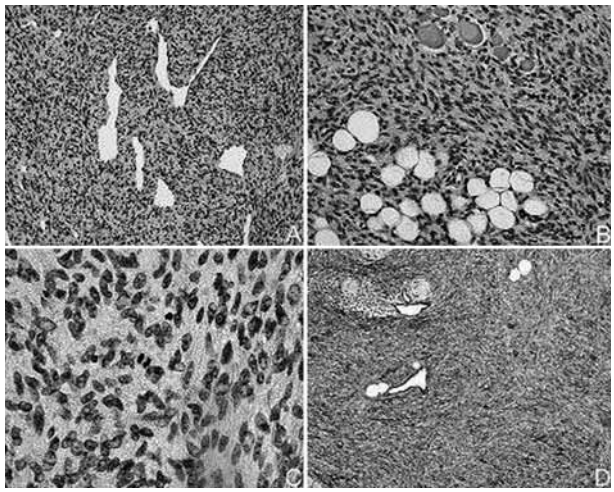

Figure 3
A. highly cellular tumour consisting of fusiform cells and characterised by haemangiopericytoma-like vessels
B. high magnification, atypical spindle cells and mitotic activity
C. the tumour infiltrates the surrounding adipose and muscular tissue
D. CD-34 expression of virtually all tumour cells

the orbit, oral cavity, thyroid gland, major salivary glands, sinonasal tract and soft tissue.[6-12] The pathologic and clinical features of extrathoracic and thoracic SFT are very similar. The symptoms are related to the anatomical site where the tumour is located or they may have a systemic expression such as hypoglycaemia, arthralgia, osteoarthropathy or clubbing.[13] Histologically, a multiplicity of pathological patterns characterise SFTs, particularly in extrathoracic locations. This heterogeneity explains the difficulty of differentiating extrathoracic SFTs from other tumours. Indeed, this tumour is composed of alternating areas of hypercellular and hypocellular zones made of spindle cells against a background of collagenised stroma with abundant "haemangiopericytoma-like" vascularisation. Areas of calcification or stromal degeneration, including myxoid change and focal pseudopalisading of tumour cells, are possible. In immunohistochemical analysis, SFT is consistently immunoreactive for vimentin, and CD34, BCL-2 and CD99 are expressed variably. The other markers, such as desmin, neural markers (S100 protein), epithelial markers (cytokeratin), vascular markers (factor VIII-related antigen) and smooth muscle actin are usually not expressed. The variability of the

Table 1
Differential diagnosis of SFT

	Haemangiopericytoma	**Solitary Fibrous Tumour**	**Dermatofibrosarcoma**
Location	Usually extremities	Pleura and extrapleural localisation	Trunk, proximal extremities
Vascular pattern	Abundant dividing sinusoidal vessels with a "staghorn" configuration	Focal "haemangiopericytoma-like" pattern	Inconspicuous vascularisation
Cell population	Homogeneous, highly cellular. Hyalinisation is uncommon	Varying cellularity, storiform arrangements of spindle cells and often thick and keloid-like hyalinisation	Uniform population of fibroblasts arranged in a monotonous storiform pattern
CD 34	Most are +	Virtually all +	Diffuse and extensive staining in most cases
Histologic malignant forms	Small number	Small number	Always

histological aspect of SFT can lead to a misdiagnosis of haemangiopericytoma or dermatofibrosarcoma. Indeed, focal haemangiopericytoma like stag horn and fibrous dermatofibrosarcoma-like storiform patterns have been described in SFTs (Table 1). When CD-34 and vimentin are expressed in most of these tumours, however, there are no specific tumour markers. Accurate microscopic evaluation by experienced pathologists is always recommended. Preoperatively, FNAB can be useful when there is a suspected SFT.[14]

When described, CT scan shows a well-circumscribed mass with a muscle-like density and intense and homogeneous enhancement.[15] The enhancement can be heterogeneous in the case of myxoid stroma, intratumoural necrosis or bleeding. Bone modifications may be reported in long-standing lesions. MRI reveals an isointense T1 signal, relative to muscle, and intense enhancement with gadolinium.[15,16] As demonstrated in our patient, T2 signal is hypointense to muscle and frequently shows intralesional heterogeneity.

Criteria for malignancy were suggested for pleural tumours by England *et al.*,[4] including high cellularity and mitotic activity (>4 mitosis per 10 high power fields), cellular polymorphism, haemorrhage and necrosis. In our case, the tumour met only a few criteria for malignancy, such as high cellularity and mitotic activity. The clinical behaviour of SFT is unpredictable and recurrence, metastasis or malignant transformation are possible even for histologically benign tumours.[17,18] Indeed, the resectability of the tumour is the best prognostic criterion. This explains why positive surgical margins, tumour size greater than 10 cm and recurrent disease are other relevant factors for local recurrence and metastasis. By contrast with previous reported series, Gold *et al.*[19] showed that extrathoracic SFTs had an increased risk of recurrence, suggesting that tumours involving the pleura were generally resected with larger margins than extrapleural SFTs. In any case, complete surgical excision is always recommended. Long follow-up is necessary because of the possibility of late recurrence and unpredictable behaviour. Radiotherapy or chemotherapy has been proposed as adjuvant treatment following incomplete resection, with variable results.[2,20,21]

In conclusion, we report a rare case of extrathoracic head and neck SFT showing typical characteristics (clinical, imaging, histopathological and immunohistochemical features) similar to those observed in other locations. Histopathological appearance can lead to confusion with haemangiopericytoma and dermatofibrosarcoma. Even when most extrathoracic solitary fibrous tumours pursue a benign course, there is a potential risk of recurrence or metastasis. Indeed, clinical behaviour is not always correlated to histological findings. Careful long-term follow-up is recommended for all patients with SFT. Complete resection is the therapy of choice. Other treatments like radiotherapy or chemotherapy have not been evaluated.

References

1. Klemperer P, Rabin CB. Primary neoplasms of the pleura: a report of five cases. *Arch Pathol*. 1931;11:385-412.

2. Dervan PA, Tobin B, O'Connor M. Solitary (localized) fibrous mesothelioma: evidence against mesothelial cell origin. *Histopathology.* 1986;10: 867-875.
3. Westra WH, Gerald WL, Rosai J. Solitary fibrous tumor. Consistent CD34 immunoreactivity and occurrence in the orbit. *Am J Surg Pathol.* 1994;18:992-998.
4. England DM, Hochholzer L, McCarthy MJ. Localized benign and malignant fibrous tumors of the pleura. A clinicopathologic review of 223 cases. *Am J Surg Pathol*. 1989;13: 640-658.
5. Hanau CA, Miettinen M. Solitary fibrous tumor: histological and immunohistochemical spectrum of benign and malignant variants presenting at different sites. *Hum Pathol.* 1995;26:440-449.
6. Krishnakumar S, Subramanian N, Mohan ER, Mahesh L, Biswas J, Rao NA. Solitary fibrous tumor of the orbit: a clinicopathologic study of six cases with review of the literature. *Surv Ophtalmol.* 2003;48:544-554.
7. Vargas PA, Alves FA, Lopes MA, *et al*. Solitary fibrous tumour of the mouth: report of two cases involving the tongue and cheek. *Oral Dis*. 2002;8:111-115.
8. Kie JH, Kim JY, Park YN, Lee MK, Yang WI, Park JS. Solitary fibrous tumor of the thyroid. *Histopathology.* 1997;30:365-368.
9. Ogawa I, Sato S, Kudo Y, *et al*. Solitary fibrous tumor with malignant potential arising in sublingual gland. *Pathol Int.* 2003;53:40-45.
10. Alobid I, Alos L, Blanch JL, Benitez P, Bernal-Sprekelsen M, Mullol J. Solitary fibrous tumour of the nasal cavity and paranasal sinuses. *Acta Otolaryngol*. 2003;123:71-74.
11. Hong JP, Chung YK, Kim SW, Kim TH, Lee KG. Solitary fibrous tumour of the face: a rare case report. *Br J Plast Surg*. 2002;55:75-77.
12. Ganly I, Patel SG, Stambuk HE, *et al*. Solitary fibrous tumors of the head and neck: a clinicopathologic and radiologic review. *Arch Otolaryngol Head Neck Surg*. 2006;132:517-525.
13. Goodlad JR, Fletcher CD. Solitary fibrous tumor arising at unusual sites: analysis of a series. *Histopathology*. 1991;19:515-522.
14. Clayton AC, Salomao DR, Keeney GL, Nascimento AG. Solitary fibrous tumor: a study of cytologic features of six cases diagnosed by fine-needle aspiration. *Diagn Cytopathol.* 2001;25:172-176.
15. Kim TA, Brunberg JA, Pearson JP, Ross DA. Solitary fibrous tumor of the paranasal sinuses: CT and MR appearance. *AJNR Am J Neuroradiol.* 1996;17:1767-1772.
16. Hyun JK, Ho KL, Jeong JS. MR imaging of solitary fibrous tumors in the head and neck. *Korean J Radiol.* 2005;6:136-142.
17. Vallat-Decouvelaere AV, Dry SM, Fletcher CD. Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors. *Am J Surg Pathol.* 1998;22:1501-1511.
18. Romer M, Bode B, Schuknecht B, Schmid S, Holzmann D. Solitary fibrous tumor of the orbit-two cases and a review of the literature. *Eur Arch Otorhinolaryngol*. 2005;262:81-88.
19. Gold JS, Antonescu CR, Hajdu C, *et al*. Clinicopatologic correlates of solitary fibrous tumors. *Cancer.* 2002;94: 1057-1068.
20. Macfarlane RG, Galloway M, Plowman PN, Thomas DG. A highly vascular intracranial solitary fibrous tumor treated with radiotherapy and toremifene: case report. *Neurosurgery*. 2005;56:E1378.
21. Dietrich CG, Roeb E, Breuer E, Matern S. Solitary fibrous thoracic wall tumor. Progression with percutaneous radiotherapy [in German]. *Dtsch Med Wochenschr.* 2001;126:12-15.

Schmitz S.
Department of Oto-rhino-laryngology and Head and Neck Surgery
St-Luc University Hospital
avenue Hippocrate 10
B-1200 Brussels, Belgium
E-mail: schmitz_sany@hotmail.com // hamoir@orlo.ucl.ac.be