Lipomatous hemangiopericytoma–solitary fibrous tumour of low malignant potential

G Liapis, IS Pateras, SH Abu Arqob, MJP Lopez, AC Lazaris*

Abstract

Introduction
Perivascular stem cells with the capacity for lipocytic and fibroblastic/myofibroblastic differentiation are involved in the pathogenesis of various mesenchymal tumours. This report discusses a case of a lipomatous hemangiopericytoma–solitary fibrous tumour of low malignant potential.

Case report
We report a case of an elderly male patient with a 6-month history of a tumour located deeply in his thigh. The tumour was surgically resected in its entirety and was well circumscribed; its maximum diameter was 6 cm. Microscopically, the tumour consisted of an adequate amount of mature fat tissue combined with cellular areas of pericytes with focal nuclear atypia. Based on the immunoreactivity of neoplastic cells to CD34 and CD99 and on the negative staining for desmin and cytokeratin, the tumour was diagnosed as a lipomatous hemangiopericytoma–solitary fibrous tumour (HPC-SFT) of low malignant potential.

Conclusion
Such lesions have been reported to behave in a benign fashion, although a few have recurred. Macroscopic features should be correlated to histological ones so that these tumours are not mistaken for well-differentiated liposarcomas.

Introduction
The border between extrapleural solitary fibrous tumour and hemangiopericytoma has recently become increasingly blurred. Perivascular stem cells with the capacity for lipocytic and fibroblastic/myofibroblastic differentiation are involved in the pathogenesis of various mesenchymal tumours. This report discusses a case of a lipomatous hemangiopericytoma–solitary fibrous tumour (HPC-SFT) of low malignant potential.

Case report
We report a case of a tumour in the deep soft tissues of the thigh of a 73-year-old man. The tumour had developed in the last 6 months before its surgical resection, and its maximum diameter was 6 cm. Morphologically, it was a well-demarcated neoplasm consisting of a varying combination of patternless cellular areas, prominent hemangiopericytoma-like vessels, variably collagenized extracellular matrix and lipomatous areas of mature lipocytes (Figures 1–4). There was a focal presence of variably ectatic or compressed, thin-walled branching vessels often having a staghorn configuration (Figure 3). Tumour cells are spindle shaped to round, generally of uniform size with small amounts of pale or eosinophilic cytoplasm with indistinct margins and often bland nuclei; however, some giant tumour cells with atypical nuclei were occasionally noticed (Figure 4). Up to three mitoses per 10 high-power fields were counted.

Figure 1: Abundant mature adipose cells intermingled with spindle cells (H-E ×100).

* Corresponding Author
E-mail: alazaris@med.uoa.gr

First Department of Pathology, School of Medicine, The National and Kapodistrian University of Athens

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Tumour cells were found to express CD99 and CD34 by immunohistochemistry (Figures 5 and 6). Immunohistochemical stains for desmin and cytokeratin were negative. No necrosis was noticeable, so due to the focal atypia and the relatively increased maximum diameter of the tumour, it was reported as a HPC-SFT of low malignant potential.

**Discussion**

HPC-SFT that originates from pericytes shows features of the classic SFT that consists mainly of spindle cells; it also contains a changeable amount of adipose tissue. It also exhibits a characteristic well-developed branching vascular pattern, and in most cases, vessels are surrounded by a thick layer of collagen. Macroscopically, HPC-SFT tumours are located in deep soft tissue. They are usually well demarcated and their size ranges from 5 to 10 cm. The vast majority of HPC-SFTs are reactive to CD34 and CD99.

First cases of HPC-SFT were reported in 1990 and 1993 by Theunissen et al. and Taccagni et al. as ‘HPC with lipomatous components’ and ‘SFT with lipomatous areas’, respectively. Nevertheless, the term lipomatous hemangiopericytoma was not coined till 1995 by Nielsen et al. In most studies, hemangiopericytoma and cellular solitary fibrous tumours are usually considered as synonyms because considerable immunohistochemical and morphological features overlap between SFT and HPC. The World Health Organization recognizes that SFT and HPC are two similar entities, if not identical.

Although HPC and SFT are not the same entity, features of both diseases can be found in the same patient. This is called HPC-SFT, the diagnosis of the case that we report. Cases of HPC-SFT are quite rare; in fact, a little amount of cases (around 60) have been reported.

SFTs are commonly associated with a benign clinical course; however, a few cases that show benign histologic features can behave aggressively. Malignant features for SFT are hypercellularity, nuclear atypia, mitoses >4/10 HPFs, necrosis, haemor-

**Figure 2:** Proliferation of spindle cells with evident collagen deposition. Presence of two dilated vessels (H-E ×200).

**Figure 3:** Anastomosing vessels surrounded by neoplastic spindle cells and a few mature adipose cells. Focal collagen deposition (arrows). (H-E X200).
Figure 4: Nuclear atypia in a tumour giant cell (H-E ×400).

Figure 5: Intense CD34 immunoreactivity in endothelial and neoplastic cells (immunoperoxidase stain ×200).

With regard to differential diagnosis, there are some closely interrelated entities that complicate the diagnosis of HPC-SFT. For example, HPC-SFT that contains fat may be misdiagnosed for well-differentiated or dedifferentiated liposarcoma. We can differentiate between HPC-SFT and myxoid liposarcomas by the presence of interstitial hyalinization, coarse-walled vessels and the absence of immature lipoblasts. Another histological finding that helps differentiation between these two entities is that in myxoid liposarcomas, vessels are not surrounded by pericytes as opposed to SFT.

Another myxoid tumour that can cause confusion with SFT is myxofibrosarcoma. The main difference is that SFT contains lipoblast-like cells that have fat vacuoles or mature adipocytes, in contrast to myxofibrosarcoma that does not form fat.

Conclusion
The diagnosis of HPC-SFT can be confused as well with angiofibroma, spindle-cell/pleomorphic lipoma and mammary-type myofibrolipoma because they are also collagenous spindle-cell tumours containing adipocytes and sometimes are reactive to CD34. However, the last three entities have a specific pattern, whereas HPC-SFTs demonstrate a distinctive hemangiopericytomaticus pattern.

Abbreviations list
HPC-SFT, hemangiopericytoma–solitary fibrous tumour

References
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Figure 6: CD99 membranous immunoreactivity of neoplastic cells (immunoperoxidase stain ×200).