A Rare Case of Solitary Fibrous Tumour of the Pre-sacral Space: Morphological and Immunohistochemical Features

E. CRISTI, G. PERRONE, C. BATTISTA, P. BENEDETTI-PANICI and C. RABITTI

1Surgical Pathology and
2Department of Obstetrics and Gynaecology, Università Campus Bio-Medico, Roma, Italy

Abstract. A 28-year-old woman presented with abdominal pain. Ultrasonographic examination showed a pre-sacral mass, with complex structure and well delimited cysts with thick walls. The resected specimen was 7.5 x 6 x 4 cm in size, well circumscribed and yellow in colour, with cystic change containing mucoid-like material. Histologically, the lesion was composed of spindle cells with high cellularity and rich vascularization with a haemangiopericytoma-like pattern. The diagnosis of solitary fibrous tumour (SFT) was made. The differential diagnosis for SFT of the pre-sacral space involves haemangiopericytoma, GIST, malignant mesothelioma, synovial sarcoma, leiomyomatous tumours and granulosa cell tumour. Immunohistochemical studies revealed reactivity for CD34, CD99 and Bcl-2, but no staining for desmin, inhibin, c-kit, EMA, CK, SMA, S-100 and CD31, confirming a diagnosis of SFT. Although SFT is usually associated with a favourable prognosis, close follow-up is recommended because of the limited information on its long-term behaviour.

SFT of the pre-sacral space can be identified as a large mass and needs to be distinguished from gynaecological tumours on the basis of imaging studies (9).

We described an unusual case of a solitary fibrous tumour located in the pre-sacral space.

Materials and Methods

The surgical specimen was fixed in 10% neutral buffered formaldehyde and tissue samples were embedded in paraffin. Routine haematoxylin and eosin staining was performed on the sections for histopathological examination.

Based on the quality of the morphological preservation of all available haematoxylin and eosin-stained slides of the tissue samples sections, we selected a paraffin block for immunohistochemical study. Immunohistochemical evaluations were carried out using the avidin-biotin-peroxidase complex method. The antibodies employed are shown in Table I.

Results

Clinical findings. A 28-year-old woman presented with abdominal pain. MRI demonstrated the presence of a disomogeneous ovaric mass, with multiple cystic formations of 5-40 mm in diameter. Ultrasonographic examination showed a mass of 8.4 x 6.9 cm, with complex echographic structure, large solid hypoechogene areas and well delimited cysts with thick walls. ECO-doppler showed hypervascularization of the solid component. CT scan revealed a well-demarcated tumour, measuring 9.3 x 8.5 cm, located in the pre-sacral space; the walls of the lesion were inhomogeneous and thick, with large sedimentations. The cystic compound was principally on the left side of the mass. Intense enhancement of the tumour indicated highly vascular tissue, which compressed the pelvic structure, in particular the uterus and rectum. The presumptive radiological diagnosis was of a serous cystoadenoma. Laboratory findings, including CA 125 and AFP, were all within normal limits. The tumour was surgically removed.
Pathological findings. The resected tumour was 7.5 x 6 x 4 cm in size, well circumscribed, rounded and yellow in colour. On the cut sections, there was cystic change containing mucoid-like material. The cystic walls had a smooth surface.

Histologically, the tumour showed high cellularity and rich vascularization with a haemangiopericytoma-like pattern (Figure 1A). Most of the lesion was composed of spindle cells, with regular, oval or round nuclei, with dispersed chromatin and small nucleoli. These cells were arranged in interlacing thin collagen fascicles that, in some areas, became more abundant with amianthoid-body-like appearance (Figure 1B). The cystic walls were composed of fibrous tissue, lacking epithelial covering and with warehouses of hemosiderin. There was some extravasation of blood cells. There was no evidence of microscopic necrosis and mitosis was rare.

Immunohistochemical findings. Immunohistochemical studies were very helpful in confirming the SFT diagnosis. By immunohistochemistry, tumour cells revealed diffusely strong positivity for CD34, CD99 and Bel-2 (8, 10) but negativity for desmin, inhibin, c-Kit, EMA, CK, SMA, S-100 and CD31. The immunoreactivity patterns of SFT are summarized in Table II. Ki-67 (MIB-1) was positive in less than 2% of the tumour.

Discussion

SFT is a rare tumour in adults, principally found in the pleural cavity. Occasional reports of this tumour occurring in other sites, such as the mediastinum, pericardium, nasal cavity, peritoneum and liver, have been increasing (2-6). This tumour has many synonyms based on the histological features, including localized benign mesothelioma, submesothelioma, localized fibrous tumour, fibroma and

<table>
<thead>
<tr>
<th>Antibody clone</th>
<th>Dilution</th>
<th>Pre-treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>CD34</td>
<td>Monoclonal Mouse; Clone QBEnd10</td>
<td>1:25</td>
</tr>
<tr>
<td>CD99</td>
<td>Monoclonal Mouse; Clone 12E7</td>
<td>1:75</td>
</tr>
<tr>
<td>Bel-2</td>
<td>Monoclonal Mouse; Clone 12A</td>
<td>1:40</td>
</tr>
<tr>
<td>Desmin</td>
<td>Monoclonal Mouse; Clone D33</td>
<td>1:100</td>
</tr>
<tr>
<td>SMA</td>
<td>Monoclonal Mouse; Clone 1A4</td>
<td>1:50</td>
</tr>
<tr>
<td>c-kit</td>
<td>Polyclonal Rabbit Clone E29</td>
<td>1:50</td>
</tr>
<tr>
<td>S-100</td>
<td>Polyclonal Rabbit Clone MNF116</td>
<td>1:800</td>
</tr>
<tr>
<td>EMA</td>
<td>Monoclonal Mouse; Clone JC70A</td>
<td>1:100</td>
</tr>
<tr>
<td>Pancytocheratin</td>
<td>Monoclonal Mouse; Clone MNF116</td>
<td>1:50</td>
</tr>
<tr>
<td>CD31</td>
<td>Monoclonal Mouse; Clone R1</td>
<td>1:40</td>
</tr>
<tr>
<td>Inhibin</td>
<td>Monoclonal Mouse; Clone MIB-1</td>
<td>1:50</td>
</tr>
</tbody>
</table>

Source of all antibodies: Dakocytomation
fibromyxoma. Its histogenesis has been the subject of marked controversy, which is reflected by the varying nomenclature used for its description.

It is uncertain if this tumour arises from the mesothelial or mesenchymal cells, the latter being preferred (7, 8) on the basis of immunohistochemical studies, which identified the tumour cells as of mesenchymal origin with fibroblastic or myofibroblastic features (5, 6).

Solitary fibrous tumours may be difficult to diagnose outside their usual location because of their histological variability. Furthermore, other pre-sacral tumours, with similar histopathological findings, include haemangiopericytoma, malignant mesothelioma, synovial cell sarcoma, leiomyomatous tumour, granulosa cell tumour and gastrointestinal stromal tumour (GIST). If difficulties persist, immunohistochemistry is usually very helpful in identifying SFT (11, 12).

CD34, CD99 and Bcl-2 were proved to be markers which were consistently expressed in SFTs (8, 9); in contrast, SFT is characterized by negativity for desmin, inhibin, c-kit, EMA, CK, SMA, S-100 and CD31. Malignant mesothelioma, synovial sarcoma, leiomyomatous tumours and granulosa cell tumour are negative for CD34. Among the CD34-positive mesenchymal tumours, GIST can easily be distinguished from SFT by its positivity for c-kit, while a haemangiopericytoma differs from SFT by its positivity for CD31. Furthermore, GIST is negative for CD99 and haemangiopericytoma is negative for Bcl-2. Malignant mesothelioma and synovial sarcoma are strongly immunoreactive for CK, on the contrary to SFT. Synovial sarcoma and malignant mesothelioma are the only neoplasms considered in differential diagnosis which are positive for EMA. Fifty-90% of leiomyomatous neoplasms are reactive for Bcl-2 but, contrary to SFT, are also positive for desmin, SMA and S-100. Granulosa cell tumour consistently expresses CD99, but it also stains with inhibin, contrary to all the other neoplasms considered in differential diagnosis. The immunohistochemical evaluation is shown in Table II.

SFTs are though to be benign, but isolated cases of recurrence and distant metastasis have variously been reported at between 13% and 23% of cases in most large series of pleural tumors (13).

Some authors have attempted to divide them into benign or malignant neoplasms, based on criteria applied to their pleural counterpart, such as high cellularity, cellular pleomorphism, mitotic counts (cut-off point at 4/10 HPF), tumour necrosis and haemorrhage (13). Instead, the most reliable prognostic indicator appeared to be the gross appearance of the tumours and resectability (14). Polypoid, pedunculated tumours behave in a benign fashion regardless of their histological appearance. The malignant cases, however, presented as poorly-circumscribed, are often infiltrative masses. Although most extrapleural SFTs are associated with a good prognosis, many authors believe that these criteria do not correlate with the clinical outcome. For this reason, complete surgical excision with careful long-term follow-up was recommended, because recurrences have occurred up to several decades after surgery (15, 16). Therefore, careful long-term follow-up is recommended, even in cases which histologically appeared benign.

In conclusion, we reported the present case of SFT of the pre-sacral space, to emphasize the importance of bearing this entity in mind, and to show that the characteristic microscopic appearance of this unusual tumour and immunohistochemistry are useful for differentiating it from other spindle cell neoplasms.

Acknowledgements

We thank Fabiola D’Ingiullo for technical support and Dr. Maria Crapulli for useful collaboration.
References


Received February 2, 2005
Accepted March 30, 2005