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Malignant solitary fibrous tumour of the chest wall: a challenging case

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Abstract. – Solitary fibrous tumors are very rare neoplasms that seldomly appear in extra-serosal soft tissues. In such cases, an accurate preoperative diagnosis is often difficult and challenging, expecially in extrapleural ones. Traditionally, extrapleural solitary fibrous tumours have been regarded as indolent neoplasms similar to their intra-thoracic counterparts, althought there has been some evidence that this subgroup could be a subset of more aggressive malignant tumours. For these reasons, surgical excision is mandatory and represents, to date, the best therapeutic option. In this article we report a case of a malignant solitary fibrous tumor of the chest wall in a 58-year-old man. Problems related to differential diagnosis and the possible pitfalls that can be encountered in the diagnostic process of such rare tumors are discussed.

Key Words:
Chest wall tumors, Solitary fibrous tumor, CD 34.

Case Presentation

A 58-year-old man come to our attention for the surgical treatment of a left parascapular mass, initially interpreted as an elastofibroma dorsi or lipomatous tumor. Fisical examination showed a slightly painful swelling parascapular mass, well-marginated, elastic and firm at palpation. No axillary lymph nodes were detected. At ultrasound scan an oval-shaped solid hypoecogenic lesion (65 × 90 × 6 mm of diameter) was detected. Magnetic resonance imaging (MRI) examination revealed the accurate anatomical location of the mass (beneath the rhomboid major and latissimus dorsi muscles) and rule out local infiltration of surrounding structures. Moreover, a high-flow vascular pattern (supported by a vascular pedicle from adjacent intercostal vessels) was detected (Figure 1) at angiographic MRI. Considering the risk of potential seeding of the malignant cells, a fine needle aspiration was not performed and a surgical resection was planned. At surgery, the mass was explored via left dorsal parascapular incision. After amyotomical incision between latissimus dorsi and serratum, a thinly encapsulated whitish mass was visualized. No macroscopic signs of peripherical infiltration were evident. Thus, the tumor was radically removed (95 × 60 × 10 mm in size and weighting 460 g, Figure 2).

Postoperative course was uneventful and the patient was discharged home the following day.

Histopathological definitive examination showed a proliferation of round to spindle-shaped cells with small cytoplasm and indistinct cell borders, with vesicular nuclei, sometimes separated by thin bands of collagen; branching haemangipericytoma-like vessels were clearly evident (Figure 3). Tumor cells showed cytoplasmic immunoreactivity to CD34, Bcl-2 and CD99 (Figure 4).

No further adjuvant treatment such as radiotheraphy was administered. At present time (36 months after surgery), the patient is alive with no sign of tumor recurrence.

Discussion

Solitary Fibrous Tumor (SFT) is an uncommon mesenchymal neoplasm originally described by Klemperer and Rabin with approximately 800

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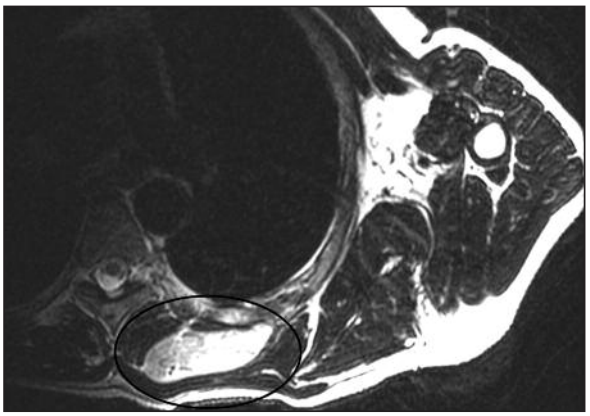

Figure 1. MRI scan visualized the lesion and accurately defined its anatomical location.

cases reported in the literature between 1931 and 2002[1]. Although SFTs mostly occur in a pleural location, up to one-third of cases has been reported in various extrapleural sites such as mediastinum[2], lung[3], meninges[4], prostate, urinary bladder, kidney[5], liver[6], nasal cavity[7], thyroid[8], salivatory glands, upper respiratory tract, peritoneum, spinal cord[9] and esophagus[10]. Extrapleural malignant SFT, especially those arising from chest wall, represent an exceedingly rare subset of soft tissue neoplasms. From literature review, only two cases have been described as parascapular lesions[11-12] and five are described as trunk lesions[13] not well specified as location (Table I).

These tumors are observed in middle-aged adults between 25 and 77 years (average age of 45 years) with no sex predilection[14]. They occur only occasionally in children and adolescents. Sometimes hypoglycemia has been observed in patients with SFTs and this is attributed to the production of insulin-like growth factor II[15], but this represents an ongoing controversial issue. In the present case hypoglycemia was not detected. Usually, ultrasound scan is not considered as the gold-standard for differential diagnoses because of the pattern findings similarity with other more common neoplasms such as liposarcomas. At computed tomography (CT)-scan SFTs appear as well delineated, smooth, soft–tissue masses that may occasionally contain scattered calcifications. MRI usually shows a lesion with a low intensity on T1-weighted images and high intensity on T2-weighted scans[16-17]. These findings are often extremely useful in the pre-operative differential diagnostic work-up.

Macroscopically, SFTs are described as well-circumscribed or encapsulated lesions with firm, grey-white cut surface, often peduncolated, and have a remarkable blood supply.

Microscopically, this kind of tumors generally are composed by alternating cells with variable amount of thick, often hyalinized or keloid-like intercellular collagen bundle, areas of necrosis and/or hemorrhage, calcifications, increased vascularity and atipia may also been seen in these lesions, as in our report Histologically, differential diagnosis of solitary fibrous tumor includes a wide variety of other benign and malignant lesions, such as sarcoma, liposarcoma, leiomyosarcoma, angioma, histiocitoma, schwannoma, neurofibroma and elastofibroma. A certain diagnosis is essentially based on this characteristic microscopic appearance in combination with immunohistochemical studies. Typically, strong and diffuse positivity for CD34 and, less significantly

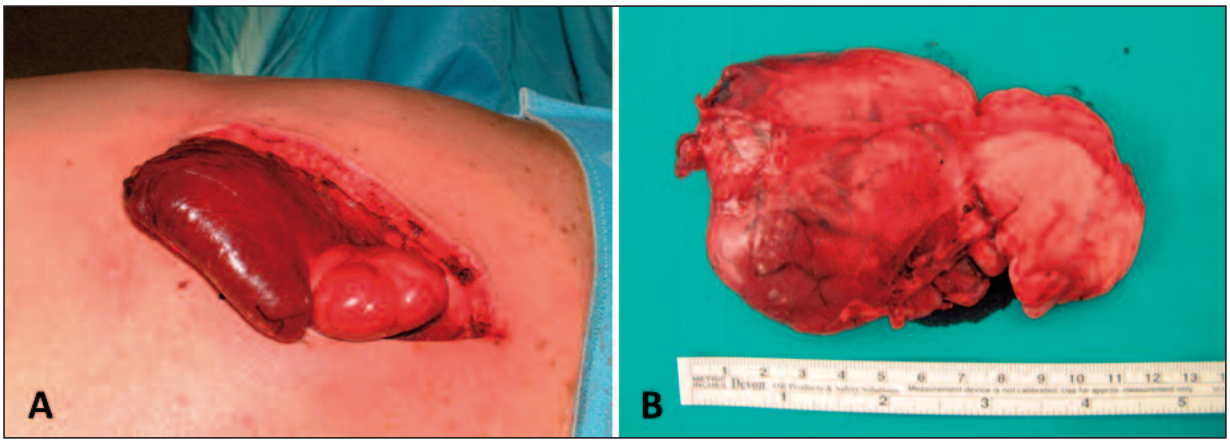

Figure 2. Macroscopic features (*A* and *B*).

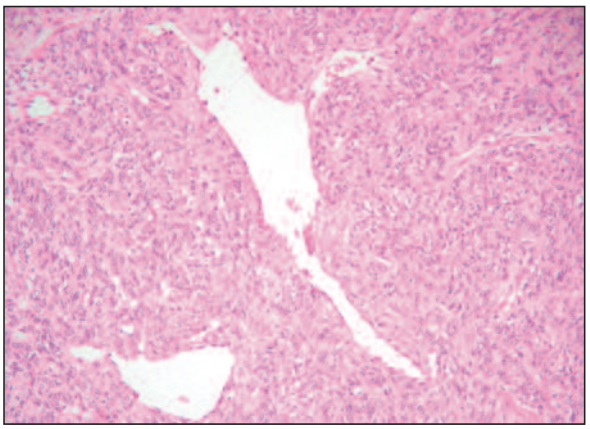

Figure 3. Microscopic features (HE × 200): Round to spindle-shaped cells with little cytoplasm and indistinct cell borders, with vesicular nuclei, sometimes separated by thin bands of collagen.

for bcl-2 and CD99, is currently regarded as the key finding of SFT. Additionally, diagnosis of malignancy was established on the basis of criteria suggested by England et al[9]. [The presence of high cellularity, cellular pleomorphism, necrosis, and high mitotic count (>4/10 HPF)], though biological behaviour of these neoplasms remains still unclear.

The majority of these tumors are histologically benign but up to 20% of all SFTs may be malignant. Histological features of extrapleural SFT are essentially similar to those of its pleural counterpart[18].

From literature review, radical surgical resection is mandatory and represents the gold standard. Up to now there is no evidence of effectiveness of chemotherapic and/or radiotherapic adjuvant treatments[19] in terms of long-term survival improvement: local recurrence or onset of metastases mainly depends on the radicality of the surgical resection and on histological findings[20]. Indeed, SFTs have an unpredictable course and, although most of them are characterized by a no aggressive clinical course, locally recurrence or disseminated metastases has been reported[21]. Thus, according with Rovera et al[22] a strict and long term follow-up is recommended.

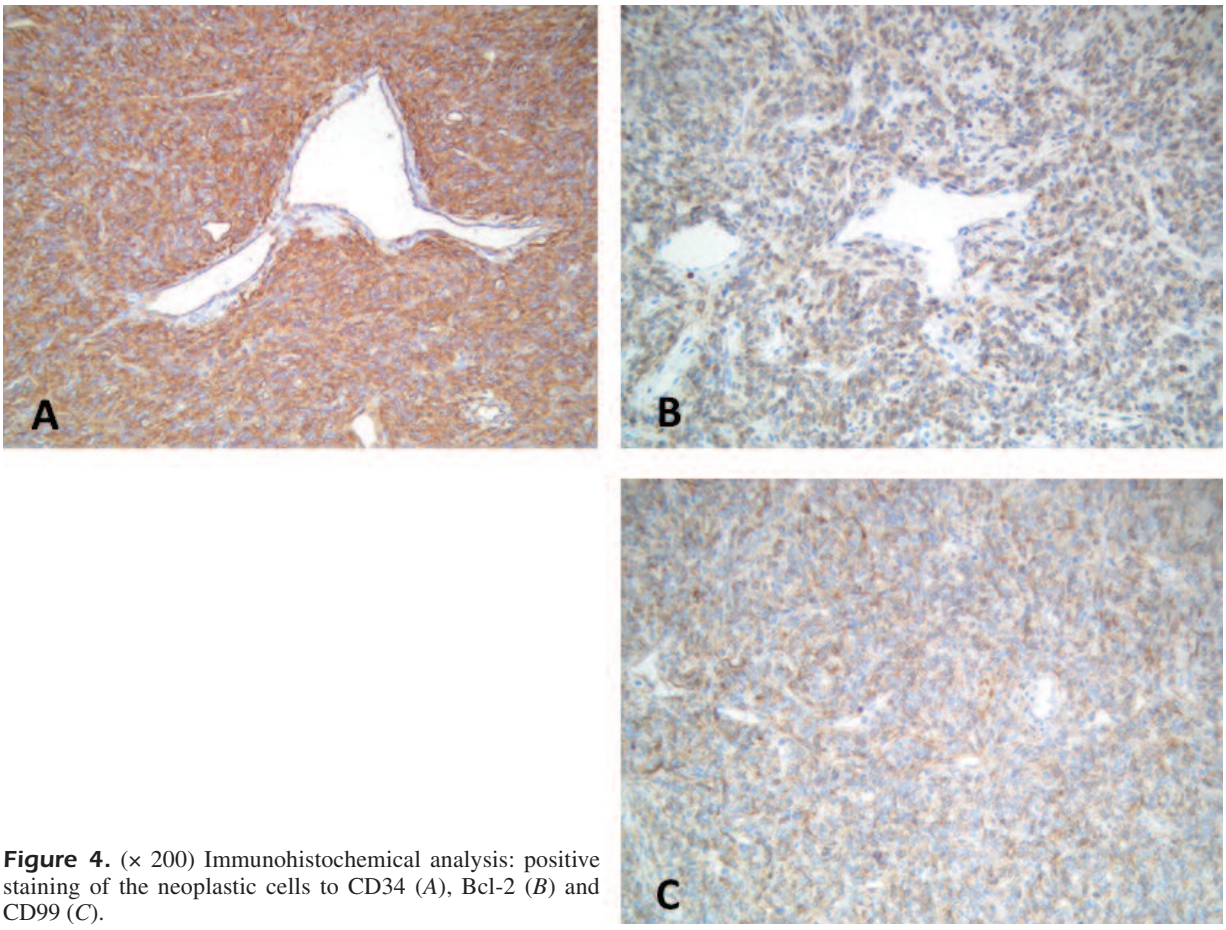

Figure 4. (× 200) Immunohistochemical analysis: positive staining of the neoplastic cells to CD34 (*A*), Bcl-2 (*B*) and CD99 (*C*).

Table I. Chest wall SFTs: literature review.

Author	N. of patients	Site	Size of (cm) primary tumor	Symptoms in first presentation	Surgical margins in final procedure	Locale recurrence	Metastases	IHC findings	Malignant components	Treatment	Chemo-therapy	Radiation therapy
Cranshaw I.M. et al[5]	5	Trunk	NA	NA	NA	NA	NA	NA	NA	Excision	No	No
Mentzel T. et al[16]	1	Left scapula	3.0	NA	Negative	No	No	CD34, V9, CD99	Absent	Excision	No	No
Daigeler A. et al[6]	1	Shoulder epifascial	4.5 × 4 × 2.5	Painless mass	Negative	No	No	CD34, V9	Absent	Excision	No	No
Our case	1	Left scapula	9.5 × 6.0 × 10	Painfull and	Negative	No	No	CD34, Bcl-2, CD99	Absent	Excision	No	No

Conclusions

Chest wall SFTs are a very rare entity with unpredictable behaviour. Precise diagnosis is usually based on the correct interpretation of specific pathologic and immunohistochemical features. Surgical excision as well as a subsequent oncological follow-up are mandatory

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