

# A solitary fibrous tumor of the upper limb

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## ABSTRACT

يعد ظهور الورم الليفي الانفرادي في الأطراف العلوية من الحالات النادرة جدا. ونستعرض في هذا المقال ظهور هذا الورم في ذراع ذكر في الثلاثينات من عمره. لقد حضر المريض الى المستشفى وقد ظهرت في ذراعه كتلة حجمها 22 سم. أجريت للمريض عملية جراحية لاستئصال الورم بالكامل. واعتمد التشخيص النسيجي للورم على ظهور الأوعية الدموية التوسعية والتصبغ الإيجابي لكل من CD34 والفيمنتين، وبعدها تمتع المريض بالصحة خلال 3 سنوات من فترة المتابعة. يهدف هذا المقال إلى زيادة الوعي عن معايير التشخيص النسيجي للأورام الليفية الانفرادية، وكذلك مبادئ الاستئصال الجراحي لها والمتابعة.

Solitary fibrous tumors (SFT) of the upper limb are extremely rare, and we report this tumor in the arm of a 30-year-old male. He is presented with a 22 cm painless mass. Complete surgical excision was performed. The histological diagnosis of SFT was based on the presence of ectatic blood vessels and positive staining for CD34 and vimentin. He remains disease-free at the 3-year follow-up interval. The report aims to increase the awareness of the criteria for the histological diagnosis of SFT, as well as the principles of their surgical excision and follow-up.

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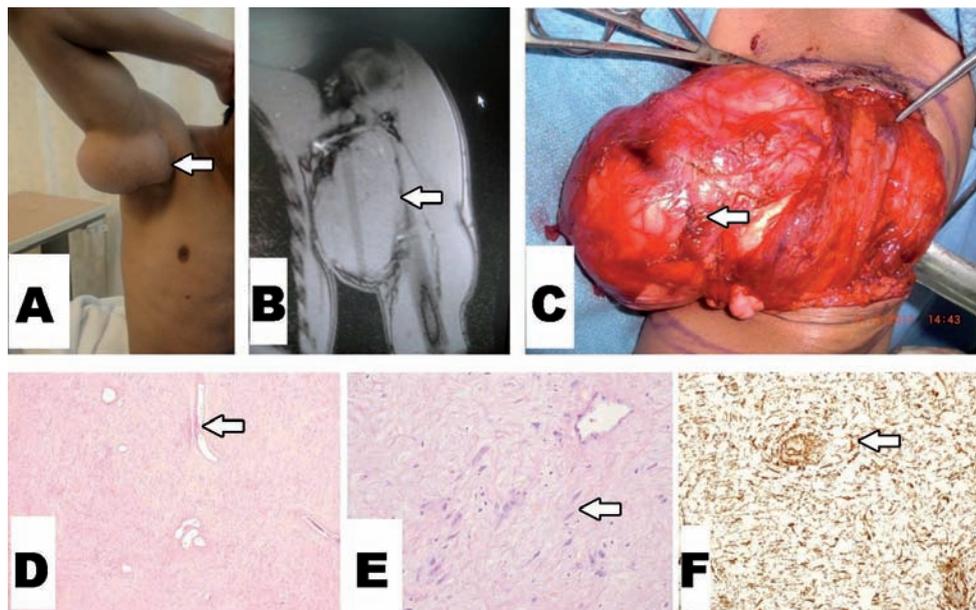
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Solitary fibrous tumors (SFT) are rare spindle-cell mesenchymal tumors, which are also known as fibrous mesotheliomas. As the name implies, the tumor is mainly seen in the pleura, the mesothelial lining of the lungs. In 5% of cases, it is accompanied by hypoglycemia secondary to tumor production of insulin-like growth factor II; and this is most often seen in tumors located in the pelvis and retroperitoneum.<sup>1</sup> The majority of the tumors are histologically benign; however, atypical features have been described in some, and malignant cases have been reported.<sup>2</sup> Extra thoracic SFT are very rare, and those involving the upper limb are extremely rare. In 2003, Akisue et al<sup>3</sup> reviewed the literature on SFT of the limbs and could only find 9 cases involving the upper limbs. Since 2003, few other upper limb tumors have been reported.<sup>2,4-7</sup> Our objective in presenting this particular case is to increase awareness, and focus on the criteria for histological diagnosis of SFT, as well as the principles of their surgical excision and follow-up.

**Case Report.** A 30-year-old man presented with a one-year history of a slowly growing mass of the left upper-medial arm (Figure 1A). There was no pain or neurovascular-related symptoms. On MRI (Figure 1B), a well circumscribed mass with intermediate signal intensity on T1 and T2 weighted images was seen. A metastatic work-up including CT scan of the chest and ultrasound of the abdomen was negative. Surgical excision was performed under general anesthesia. A well-circumscribed tumor was found with a firm whitish capsule (Figure 1C). The tumor mass along with the capsule was excised. The mass measured 22 x 10 cm and cut-sections showed a multi-nodular mass with a smooth surface feature. Histological examination showed a benign hypocellular fibrous tumor with ectatic blood vessels (Figure 1D); composed of fibroblasts separated by thin bands of collagen (Figure 1E). There were no areas of tumor cell necrosis, and mitotic figures were rare. There was no pleomorphism or atypia. All resection margins were free of tumor. Tumor cells stained positive for CD34 (Figure 1F), and vimentin; and negative for desmin, actin, cytokeratin, and S-100 protein. The final



**Figure 1** - Clinical photo showing: A) preoperative appearance, B) MRI showing an iso-intense mass, C) surgical excision of the mass along with its capsule, D) the tumor is hypocellular with ectatic blood vessels (hematoxylin and eosin stain x40), E) high power showing the fibroblasts surrounded by thin collagen (hematoxylin and eosin stain x200), and F) positive CD34 immunostain (x100).

diagnosis was a benign solitary fibrous tumor. He did well after surgery and after 3-years is disease free.

**Discussion.** Solitary fibrous tumors of the upper limb are extremely rare, and less than 20 cases were reported in the literature.<sup>2-9</sup> Historically, SFT were known as hemangiopericytomas because the presence of stag-horn vascular pattern was thought to be an indication of a pericytic origin. Later, this stag-horn vascular feature was found to be present in several other benign, and malignant tumors such as infantile myofibromatosis, and synovial sarcoma.<sup>10</sup>

The differential diagnosis of SFT is a long list of benign fibrous tumors that are known to affect the upper limb of children and adults. All benign fibrous tumors are mainly composed of fibroblasts (spindle cells) separated by collagen bands. The diagnosis of the type of tumor will be based on other histological features and on immunohistological analysis.<sup>11,12</sup> An accurate histological diagnosis is essential because it will determine the behavior, risk of recurrence, and the adequacy of surgical resection. For example, nodular fasciitis has a very low risk of recurrence and may even regress spontaneously; while desmoid tumors are locally aggressive with a high risk of recurrence and hence require wide local excision. The diagnosis of nodular fasciitis is based on the presence of myxoid areas and positivity for actin staining, while desmoid

tumors are positive for desmin staining. Therefore, an experienced histopathologist is an essential member of the team for managing fibrous tumors of the limb. The most characteristic histological feature of SFT is the presence of ectatic or stag-horn blood vessels; although other distinct histologic patterns have been described.<sup>6</sup> Furthermore, SFT cells will stain positive for CD34 and vimentin; and will stain negative for actin, desmin, and other epithelial (such as cytokeratin), and neural (such as S-100 protein) markers. Finally, all benign-looking fibrous tumors should be carefully examined for any areas of atypia or polymorphism. These features suggests the presence of a malignant component, and Baldi et al<sup>13</sup> have reported a distant metastases from tumors with such features. The tumor in our patient had no such features. However, it should be emphasized that histological features will not always predict biological behavior.

Surgically, the tumor mass should be removed along with its capsule since failure to do so is associated with increased risk of local recurrence.<sup>14</sup> Other factors that are known to be associated with local recurrence include positive surgical margins, accidentally going through the capsule during surgery, high cellularity, and the presence of frequent mitotic figures.<sup>13</sup> None of these features were present in our patient and there was no local recurrence through the 3-year follow-up visit. Long-term follow-up is recommended for benign

SFT since they may have a very slow growth, and late recurrences have been reported.<sup>13</sup> The histological features of the tumor of our patient are associated with a low risk of recurrence, and surgical excision was curative. Long-term follow-up is planned for our patient.

In conclusion, we report a rare case of SFT of the upper limb and emphasize the principles of excision, the diagnostic criteria, and follow-up of similar cases. Excision should include the capsule surrounding the tumor mass. The diagnosis should be confirmed by the presence of stag-horn vessels and positivity for CD 34 and vimentin. Long term follow up is required even for tumors with rare mitotic figures.

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## Case Reports

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