CASE REPORT

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A case of extrapleural solitary fibrous tumor of the thigh with eight years follow-up

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Abstract

Solitary fibrous tumor (SFT) is a rare neoplasia. Now classified as SFT, it was first described in 1942 as "hemangiopericytoma" (HPC), and its origin was supposed to be the pericytes. The location outside the pleura is considered uncommon and the tumor situation on the thigh is much more unusual. In this article, we present a case of a SFT located in the hip. Even if the limb situation of the SFT is considered rare, some tumors in lower limb were reported in the past few years. Our initial clinical diagnosis, in this case, was a variant of lipoma (fibrolipoma), so no further investigations were performed before surgery at that time, and the surgery was planned as a simple procedure. The mimics of SFT with a large variety of benign tumors, especially in long-term evolution cases, make the preoperative diagnosis much more difficult. After the excision of the tumor and histological and immunohistochemical (IHC) tests, the diagnosis was finally stated. Even the findings in the exam conducted us to a most likely benign evolution, the eight years follow-up allowed us to strongly correlate the evolution with laboratory findings in this case. Further follow-up in this case can and will be performed.

Keywords: solitary fibrous tumor, thigh, surgery, CD34, Ki67, ACT.

Introduction

Solitary fibrous tumor (SFT) is a rare mesenchymal tumor, characterized by a proliferation of ovoid and fusiform cells immunoreactive for CD34 [1] and thin wall ramified blood vessels. This entity now called solitary fibrous tumor was first described in 1942 as "hemangiopericytoma" (HPC), and was supposed at that time to have the origin in the pericytes [2]. Today, even if the differentiation cell line is uncertain, there are elements that suggest the tumor cells fibroblastic/myofibroblastic origin, and the term of hemangiopericytoma is used more as a descriptive term [3].

The fraction of HPC, comparable to SFT, shows frequent positivity for CD34 and CD99, both of them usual expressed in fibroblastic tumors; endothelial markers are negative; actin and desmin are also negative in most cases [1].

The SFT is considered to be benign in more than 70% of cases [1], but recent studies suggest that the malignant rate is about 50% [4]. Even so, some other studies show only a 29% local recurrence (with 50 months median time to event) and 34% metastasis rate (with 44 months median time to event) [5]. Because of this risk of recurrence even after a great period of time, long term follow-up is mandatory [5, 6].

Even the SFT of the thigh are considered to be extremely rare, the percentage varies in different studies. In a 2002 study, thigh SPF was found in 2% of the cases [7], in 2014 was found in 4% of them (one in 28 cases) [6] and in 2015 was found as 16% (cases on both extremities) [4]. Another 2015 study found that thigh SFT was found in 50% of the only six extremities cases [8].

The diagnosis of the SFT in thigh is difficult [9], especially when the tumor is small, superficial and with slow or no grow during a long period of time and almost no symptoms. Confusion with a lipoma or a fibrolipoma can be done with ease if the tumor is suprafascial and small. Imaging can be useful. Soft tissue ultrasound will offer quite limited information, as well as X-ray examination [9, 10]. Magnetic resonance imaging (MRI), positron emission tomography (PET) and angiography are much more useful and other supplementary investigations can be done if necessary [10]. Differential diagnosis has to be done with posttraumatic conditions, other benign tumors of the soft tissue, and/or soft tissue malignancies. Total body scan is useful to detect distant metastasis, if the suspicion of malignancy exists.

We present a rare case of a 36-year-old woman with a thigh SFT, followed-up eight years after surgery, and discuss the results of the histological and immunohistochemical (IHC) examination and possible correlations with the outcome.

Case presentation

The patient, NC, Caucasian woman, aged 36, living in urban environment, presented on May 2007 to Division of Plastic and Reconstructive Surgery, Emergency Clinical Hospital Bucharest, "Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania for a tumor on her right thigh.

The tumor had a very slow onset, and was first noticed about two years before. The patient could not link the tumor with any trauma, and was completely painless. The

situation was superficial, and the fist reason the patient asked for medical assistance was the cosmetic reason. It became more evident after the patient underwent a voluntary diet, and lost about 6 kg in the last three months. The patient had no other health problems and no family history of malignancies (Figure 1).

Clinical exam showed a relatively increased consistency tumor mass, in the anteromedial side of the right thigh, of approximately 5/6 cm in diameter. The situation was very superficial, in the suprafascial plane, and the tumor was mobile both on the fascia and skin, and deformed the contour of the thigh, aspect visible and disturbing for the patient after the diet. No pain and no pulses were detected at palpation. Usual lab examinations were normal (Figure 2).

We suspected a benign tumor, most likely a lipoma or a fibrolipoma. The X-ray exam was inconclusive, and the soft tissue ultrasound detected a single tumor mass, of approximately 6/4 cm, with uniform structure and surrounded by a capsule. No further investigation was considered necessary. A simple excision with intravenous anesthesia was planned.

Surgical procedure went with ease, considering the suprafascial situation of the tumor, and the capsule allowed a straightforward dissection from the surrounding fatty tissue.

The tumor was processed to paraffin blocks, cut in 3 μm thickness sections, and stained with Hematoxylin–Eosin (HE) initially. After this, two sets of supplementary IHC examinations were performed.

The follow-up continued for eight years, and after that, a MRI exam was performed. The follow-up is planned to continue until 12 years after surgery.

After the resection, the tumor measured approximately 55–40–35 mm and was covered with a thin well-defined capsular sheet (Figure 3).

However, a partial section of the tumor offered an unforeseen look of the inside, completely different from what we expected (Figure 4).

The first the HE staining histopathological examination concluded a border malignancy hemangiopericytoma, with relative frequent mitosis, cell pleomorphism, and no anaplastic cells and no tumor necrosis or bleeding.

Tumor was built up of monomorphic oval and spindle cell proliferation, with minimal cytological atypia, cellular indistinct edges, forming various cellular density areas (Figure 5). The tumor appears to be well defined, bordered by a thin fibrous capsule (Figure 6). Thin wall intratumor branched vessels (hemangiopericytoma-like vessels) were found. Mitotic rate was low, about 1–2 mitoses/10 HPF (high-power fields) (Figure 7).

The diagnosis was confirmed and refined by IHC tests. A first IHC test concluded: CD34 diffuse positive in tumor cells; Ki-67 unquantifiable and S-100 / α-SMA / Bcl-2 / CD99 were negative.

After this first examination, another IHC investigation was performed on 3 μm serial sections from one of the paraffin blocks. The sections were incubated with primary antibodies: CD34, ACT and Ki67. Then was applied the detection system Poly-HRP-GAM/R/R IgG (Immunologic) and DAB (3,3'-diaminobenzidine) solution for developing. The slides were counterstained with Mayer's Hematoxylin.

CD34 was diffusely positive in tumor cells (Figure 8). ACT was found negative in tumor cells, and positive in blood vessels (Figure 9). Ki-67 was found in 5% of tumor cells (Figure 10).

The IHC tests sustain the diagnosis of extrapleural SFT. Even there is no strict correlation between histological and IHC aspects and clinical evolution, the findings on this case suggest a further benign evolution (WHO code 8815/1).

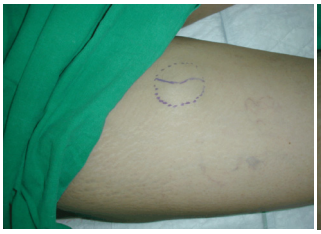

Figure 1 – ***Right thigh seen from front before surgery. Tumor limits are marked.***

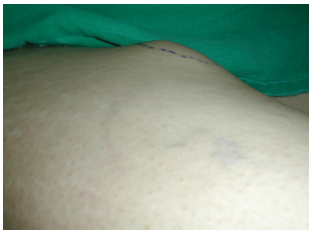

Figure 2 – ***Tumoral deformation of the right thigh.***

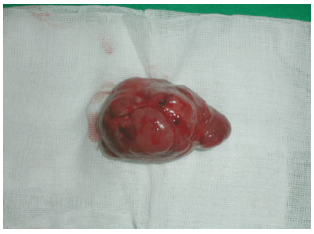

Figure 3 – ***Macroscopic aspect of the encapsulated mass.***

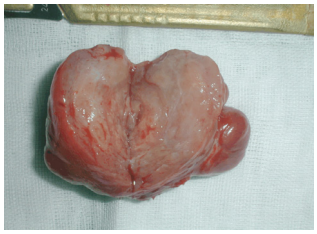

Figure 4 – ***Macroscopic aspect of the tumor half sectioned.***

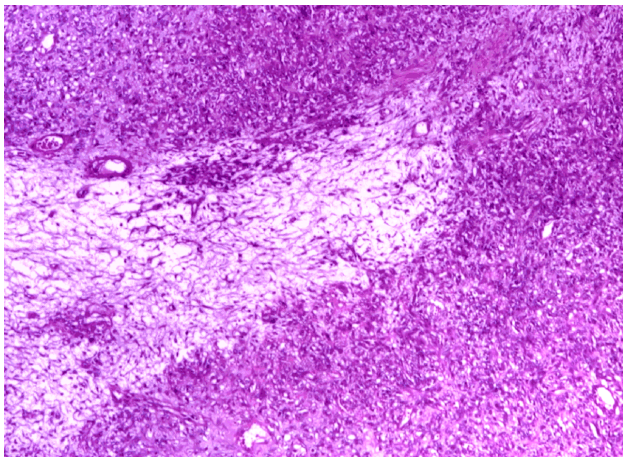

Figure 5 – ***Extrapleural SFT. Irregular cell density. HE staining, ×40.***

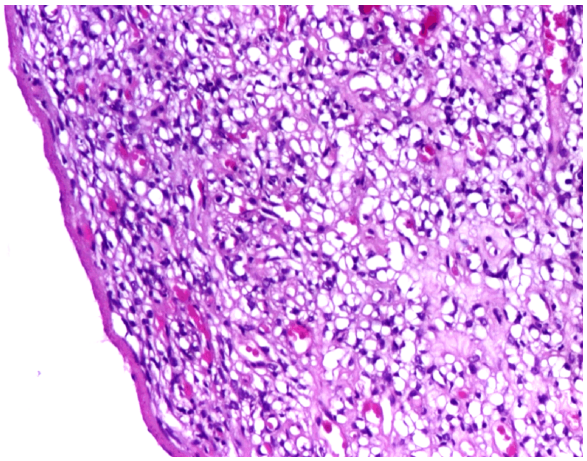

Figure 6 – ***Extrapleural SFT. Thin capsule bordering the tumor. HE staining, ×200.***

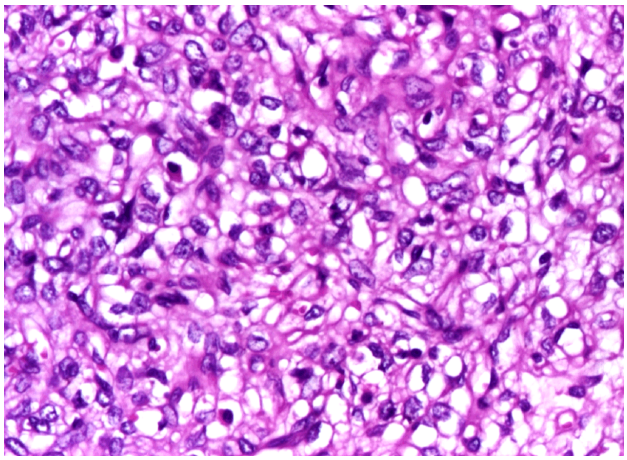

Figure 7 – ***Extrapleural SFT. Branched blood vessels with thin wall. Low mitotic rate. HE staining, ×400.***

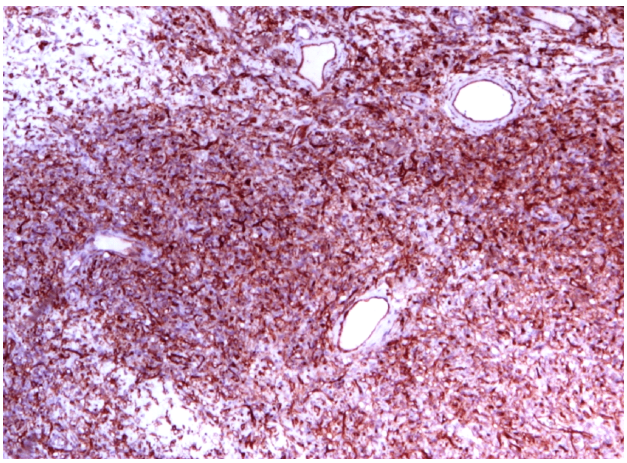

Figure 8 – ***CD34 immunostaining positive in tumor cells (×100).***

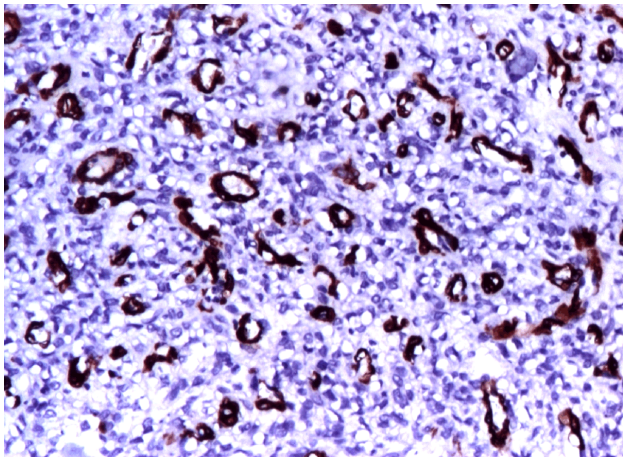

Figure 9 – ***ACT immunostaining negative in tumor cells, positive in blood vessels (×200).***

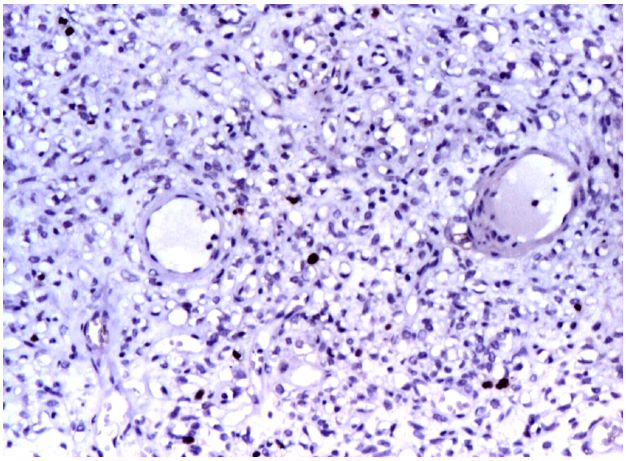

Figure 10 – ***Ki67 immunostaining – about 5% positive in tumor cells (×200).***

Considering all of those, we decided to make a long-term follow-up of the case. No clinical event was detected in the first eight years locally, and no sign of relapse could be pointed out on general exam. After eight years (94 months), in May 2015, we decided to make a MRI examination of the hip region, to detect with great accuracy any kind of local relapse. The result concluded that there was no sign of tumor recurrence on the antero-medial side of the right thigh (Figure 11).

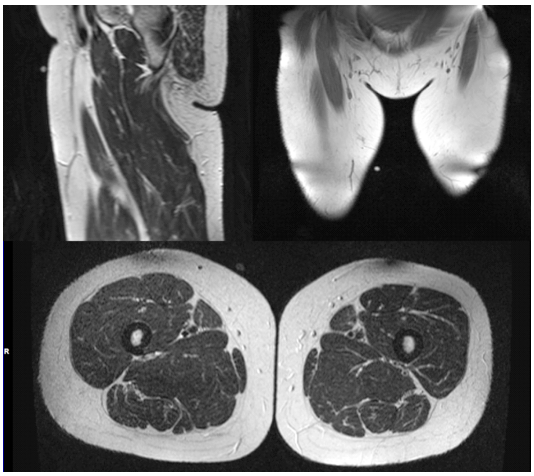

Figure 11 – ***Right thigh MRI examination at eight years checkup – sagital, frontal and horizontal sections without signs of local recurrence.***

Discussion

Extrapleural SFT is still considered a rare tumor [5] that means a prevalence of less than five cases/100 000 [11]. Even extrapleural location is considered unusual, the described locations of the SFT until now were head and neck, lung, mediastinum, diaphragm, liver, omentum, pancreas, retroperitoneal space, spermatic cord, trunk soft tissue [7], prostate [7, 12], penis [13], central nervous system [14], inferior vena cava [15], cheek [16].

At the lower limb were reported: one case of the ankle in 2015 [17]; one case of the leg (calf) in 2015 [9] and one in 2014 [18]; in the thigh was one case described in 2006 [10] and one in 2015 [19]. Between 1997 and 2003, other nine more thigh extrapleural SFT tumors were reported [10]. Another subcutaneous case of extrapleural SFT was described in the cheek in 2014 [16].

Even the tumor is rare, considering the prevalence, the number of studies and case reports cited by PubMed regarding this topic increased in the last decades from 9/1995 to 34/2005 and 125/2015. The interest on this kind of tumor is constantly growing, and this effort will allow a better understanding of the disease. Consequently, this condition will be better diagnosed and treated, and the evolution possibilities will become more predictable, as data accumulates.

The case described in our work seems to be the third of thigh situated SFT reported in the last 10 years, and

the second in the subcutaneous layer in the same period.

The distinctive feature of our case is the relatively small size of the tumor; the very slow grow rate, and its situation in the suprafascial layer of the thigh. Those particularities, together with the patient voluntary diet and the consecutive weight loss (6 kg in three months), which revealed the tumor, directed us to the fist clinical diagnosis a soft tissue superficial benign tumor (most probably a lipoma). Our simple initial evaluation was wrong. Even so, no major changes of the surgical treatment were made if the preoperatory diagnosis would have been more accurate.

After the excision, the histological and IHC examination was performed with care. Even the histological findings were somehow reassuring for a benign evolution, we continued with the IHC. Those findings were also reassuring [16]: S-100, α-SMA, Bcl-2, CD99 negative. On the other side, CD34 was diffusely positive, Ki67 was found in 5% of tumor cells, ACT was found negative in tumor cells, but positive in blood vessels, conducting to the same conclusion [4, 16, 5]. Also, the size of tumor, less than 10 cm and the low mitotic rate leaded us to the same conclusion, namely a benign behavior of the tumor [5].

We did not performed any molecular study to detect NAB2–STAT6 gene fusion, considered to be the hallmark for SFT [6]. Further tests will be considered.

The clinical survey of the patient lasted eight years after the surgery. Because no recurrences were detected a MRI of the region was performed. This one also showed no relapse of the disease. No other recurrence was detected at a detailed clinical exam. Even we can consider the patient cured, the follow-up will continue up to 12 years, because the latest recurrences in SFT occurred, as we know, after 88 months in an extrathoracic SFT and after 118 months intrathoracic case [7].

Conclusions

Any subcutaneous tumor can be an extrapleural SFT, even the dimension is small, the clinical aspect is benign and the growth rate is low. Proper histological and IHC examination after surgery is mandatory. The correlation of the histological and IHC examination with the outcome seems to be significant. Long-term follow-up is recommended to detect and treat any relapse of the disease. Further molecular studies are recommended, if possible.

Conflict of interests

The authors declare that they have no conflict of interests.

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