Case Report

Malignant Solitary Fibrous Tumor of the Thigh: A Case Report

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Abstract

Malignant solitary fibrous tumor, previously known as hemangiopericytoma (HPC), is a rare spindle-cell mesenchymal tumor of probable fibroblastic derivation. It usually affects serosal surfaces and the pleura although extrapleural sites have been reported. We report here a case of a 56 year old male, admitted in our clinic for the management of an enlarged mass of the right distal femur. Histopathologic analysis of the surgical specimen retrieved a diagnosis consistent with a malignant solitary fibrous tumor, in R0 tumor margin resection. Malignant solitary fibrous tumor of the thigh is an extremely rare condition. A safe margin at the surgical excision and a close follow-up remain the standard of care.

INTRODUCTION

First described by Wagner in 1870, solitary fibrous tumor (SFT) was further defined by Rabin and Klemperer in 1931 reporting five cases of a primary pleural neoplasm [1,2]. Murray and Stout in 1942 introduced the term hemangiopericytoma describing a vascular tumor featuring Zimmermann's pericytes [3]. It is now known that the well-developed “staghorn” branching pattern of the vasculature is not a unique feature of hemangiopericytoma, since it can be found in all soft tissue tumors [4].

Although the etiology remains unknown, solitary fibrous tumors affect primarily adults with a median age of 45-50 years, representing approximately 2% of all soft tissue tumors [5]. According to the latest WHO classification of bone and soft tissue sarcoma, solitary fibrous tumor is classified into two forms; typical and malignant forms of solitary fibrous tumor are distinguished based on the number of mitosis (>4 per high power field), the presence of cellular atypia or necrosis and hypercellularity [6].

We report here a case of a 56 year old male, where the diagnosis of malignant solitary fibrous tumor was an incidental finding after surgical excision of an enlarged mass of the right distal femur.

CASE PRESENTATION

An otherwise healthy 56-yrs old patient was admitted to our department for the surgical management of an enlarged, painless mass of the right distal femur first noted by the patient about 11 months ago. Clinical examination revealed a non-tender, palpable mass of approximately 9cm maximum diameter of the distal thigh, proximal to the medial condyle of the femur. No reduction in the range of motion of the knee joint was noted and no inguinal lymphadenopathy was present. Ultrasonography showed the presence of a heterogeneous, solid, well circumscribed mass (10x6 cm) with intense vascularization, in the distal third of the thigh, as shown on (Figure 1a, 1b). The patient underwent a surgical excision in the Day Care Unit of our hospital and was discharged 6 hours postoperatively after an uneventful recovery.

Figure 1a Ultrasonography of the mass showing the well circumscribed borders and its length.
Histopathologic analysis of the surgical specimen confirmed the diagnosis of a malignant solitary fibrous tumor, in R0 tumor margin resection. Macroscopically the specimen weighted 416 gram with dimensions of 13x9.5x8 cm presenting a firm and whitish surface with an intact tumor capsule. Microscopically a mesenchymal neoplasm exhibiting a patternless architecture was noted along with spindle cells in a fibrous stroma. High cellular atypia was present as well as 7 mitoses per high power fields were noted. The immunohistochemical analysis was positive to antibodies against CD34 and vimentin and negative for desmin, SMA, S-100, h-caldesmon, DOG-1, Bcl-2 and CD117. The patient is under close follow-up and is well 15 months following surgery. There is neither any local recurrence nor any distant metastasis.

**DISCUSSION**

Malignant solitary fibrous tumor is a distinct neoplasm of mesenchymal origin often arising from the pleura. Various extraserosal sites have been reported such as in thyroid [7,8], kidney [9], liver [10], intra-meningeal [11] and in orbital space [12]. Until recently only nine cases of malignant solitary fibrous tumor affecting the thigh have been reported [13]. Symptoms are non-specific and usually the presentation consists of painless mass in deep soft tissue of the thigh. Hypoglycemia has been reported in up to 5% of the cases as a paraneoplastic phenomenon due to the avid production of insulin like growth factors by the tumor [14].

In our case, the diagnosis of malignant solitary fibrous tumor was set after the surgical operation although it has been reported that clinical examination, MRI and ultrasound may not be sufficient in setting the diagnosis preoperatively [15].

A definite diagnosis is set only after the histopathologic examination of the specimen. Immunohistochemistry using antibodies against the vimentin and CD34 antigen along with the presence of spindle cells, notable atypia, tumor necrosis, hypercellularity and more than 4 mitoses per HPF are the gold standard setting the diagnosis of malignant solitary fibrous tumor [16,17].

Tumor cells tend to metastasize via hematogenous dissemination primarily in the lungs. A wide surgical resection is of paramount importance offering the patient a five-year survival rate of approximately 90 - 100%. Although distant metastasis rate has been reported up to 19%, the local recurrence rate regarding a malignant solitary fibrous tumor of the extremity is less than 6% [18]. Radiation therapy may be considered as an option along with a close long-term follow-up in selected cases where primary surgical excision cannot be achieved (meningeal and retroperitoneal cases) or in case of patient refusal [19-23].

**CONFLICT OF INTEREST**

Financial & competing interest’s disclosure

The authors have no relevant affiliations or financial involvement with any organization or entity with a financial
interest in or financial conflict with the subject matter or materials discussed in the manuscript. This includes employment, consultancies, honoraria, stock ownership or options, expert testimony, grants or patents received or pending, or royalties. No writing assistance was utilized in the production of this manuscript.

**Ethical approval**

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

**Informed consent disclosure**

The authors state that they have obtained informed consent from the patients for the inclusion of their medical and treatment history within this case report.

**REFERENCES**