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Case Report

Low-grade fibromyxoid sarcoma arising in the mediastinum: Case report and review of the literature [☆]

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ABSTRACT

Low-grade fibromyxoid sarcoma (LGFMS) is a rare tumor characterized by bland histological features and aggressive clinical course, it is a distinctive variant of fibrosarcoma with high metastasizing potential and sometimes a long interval between tumor presentation and metastasis, the most common anatomic locations of occurrence are the lower extremities, thorax, inguinal area, and upper limbs, Here, we present the case of a 29-year-old patient with a deceptively benign-appearing mass arising in the mediastinum that had been discovered on computed tomography, a biopsy under cervicotomy was performed and histopathological examination revealed low-grade fibromyxoid sarcoma. In conclusion, LGFMS can arise in the mediastinum, and physicians should consider this entity as a differential diagnosis in the presence of such mass.

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Fig. 1 – In the thorax computed tomography image, a large circumscribed soft tissue density lesion of the upper and middle mediastinum with cervical extension (A, B), pushing back the trachea (star), esophagus (blue arrow), and azygous vein (green arrow) to the right, not enhanced after injection of contrast agent (C).

Introduction

In 1987, Evans was the first author to describe 2 cases of metastasizing soft tissue tumors with a deceptively benign histologic appearance that he named low-grade fibromyxoid sarcoma (LGFMS).

It may be located in any of the locations where other soft tissue sarcomas have been described. However, only a few cases with mediastinal locations have been reported to date [1].

On histological examination, this tumor is found to consist of alternating fibrous and myxoid areas, arranged in a spiral fashion with fibroblast spindle cells that appear benign [2].

Case report

A 29-year-old patient presented with a large non-painful left lateral-cervical mass, gradually increasing in size, initially neglected, worsening 15 days before admission by the onset of dyspnea, a computed tomography (CT) scan was then ordered and a mediastinal mass was found. The CT scan (Fig. 1) showed a large circumscribed soft tissue density lesion of the upper and middle mediastinum with cervical extension, not enhanced after injection of contrast agent, containing some peripheral microcalcifications, pushing back the trachea, esophagus, and azygous vein to the right, encompasses the left subclavian artery and the left vertebral artery without signs of invasion, and arriving in intimate contact with the vertebral bodies from C7 to D6 without bone lysis, The lesion measured 10.4 × 6.6 × 18.9 cm. Magnetic resonance imaging (MRI) evaluation of the area (Fig. 2) demonstrated a mass in

low T1 signal, in high T2 signal, without marked enhancement after injection of Gadolinium.

The patient subsequently underwent a biopsy under cervicotomy for additional information. Histological evaluation revealed a morphological appearance of a non-atypical myxoid spindle cell tumor proliferation, IHC staining was performed which showed an atypical myxoid spindle cell tumor proliferation suggesting low-grade fibro myxoid sarcoma or low-grade myxoid liposarcoma. A molecular analysis (t (12;16) (q13;p11.2) FUS-DDIT3 – t (12;22) (q13;q12): EWSR1-DDIT3) was also performed and showed an absence of mutation of the DDIT3 gene; therefore, the profile is in favor of a LGFMS.

Discussion

Benign soft tissue lesions may histologically mimic soft tissue sarcomas, and the reverse can also occur with sarcomas being mistaken for benign lesions [3].

In 1987, Evans first recognized LGFMS as a unique entity seen in young and middle-aged adults, which was initially identified as a slow-growing, asymptomatic tumor of the soft tissue with deceptively mild histology but with a higher risk of recurrence and metastasis [2].

Most frequently, these lesions are located in the deep soft tissues of the lower extremities and thighs. They are situated less frequently in the chest wall, the axilla region, the shoulder, and the inguinal area, the mediastinal occurrence is very rare [1].

In 1999, Takanami, and al reported the first case of mediastinal LGFMS in a 35-year-old male, which was primarily mis-

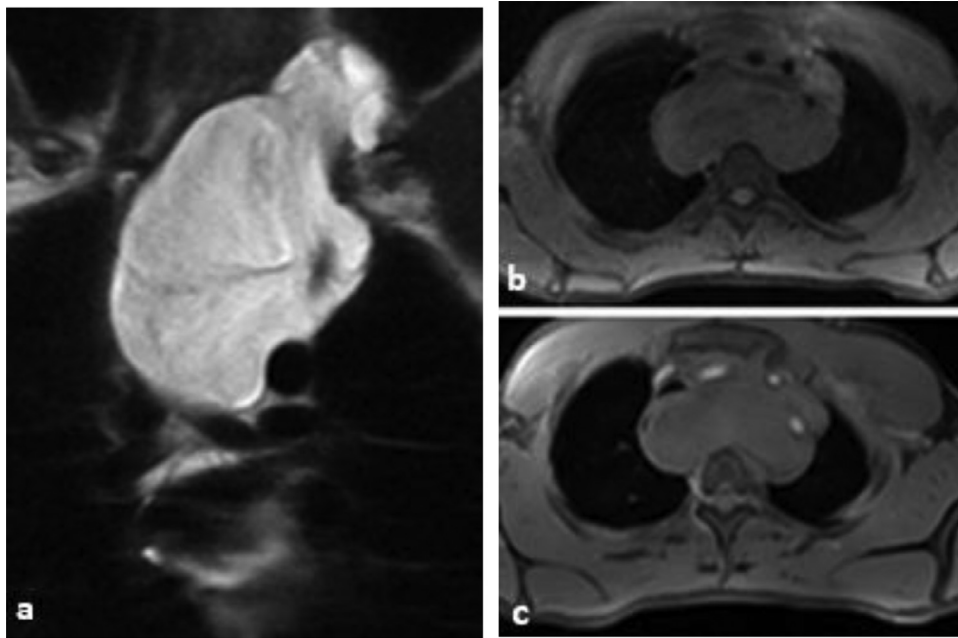


Fig. 2 – Magnetic resonance imaging: mediastinal mass in low T1 signal (B), in high T2 signal (A), without marked enhancement after injection of gadolinium (C).

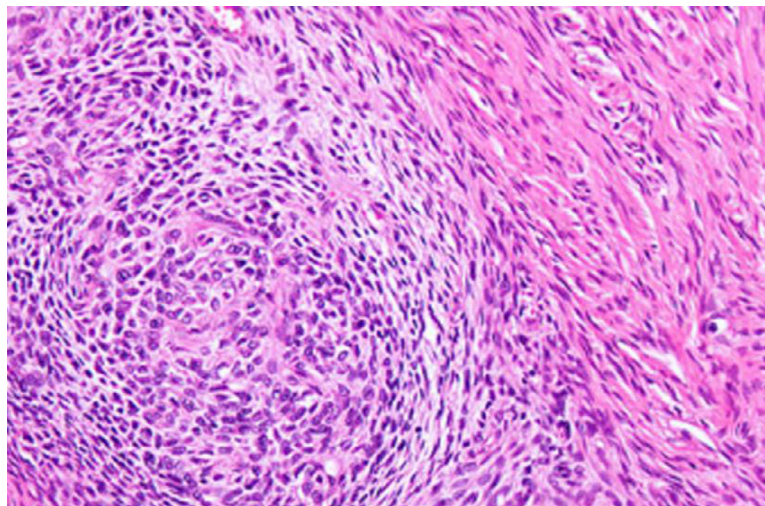


Fig. 3 – Low and intermediate power shows a tumor with alternating cellular myxoid and fewer cellular collagenized areas.

diagnosed as neurofibroma. After 9 years, it recurred and was correctly diagnosed as LGFMS [2].

Histologically, a spindle cell lesion is seen with alternating myxoid and hyalinized areas, it is characterized by an admixture of collagenized hypocellular zones and more cellular myxoid nodules [4] (Fig. 3).

The histological differential diagnoses of LGFMS include solitary fibrous tumor, low-grade peripheral sheath tumor, neurofibroma, and desmoid fibromatosis. Solitary fibrous tumor with dilated hemangiopericytoma-like vessels and variable stromal collagen exhibits a patternless growth pattern [2].

Therapeutically, these tumors require an aggressive therapeutic approach. Surgical excision must be as complete as

possible, which is not often the case, these tumors being often of large size and close to vital elements. The removal of mediastinal sarcomas is only possible in about two-thirds of cases. The quality of the excision is assessed by the absence of tumor invasion of the banks. This resection will be, on the one hand, for histopathological diagnostic purposes and, on the other hand, for therapeutic purposes.

The contribution of chemotherapy is poorly understood. She seems effective in metastatic forms and would allow reduction of tumor size for surgery secondary [5].

Although LGFMS is deceptively benign in appearance, it has a high local recurrence rate, and, in a majority of patients, it recurs frequently and metastasizes into the lungs. Local re-

currence has been reported in nearly 65% of LGFMS patients. In the literature, the recurrence has been reported to occur as early as 6 months after the excision of the first lesion and as late as 50 years after the excision [1].

Conclusion

In conclusion, we report a rare case of primary mediastinal LGFMS that was incidentally discovered in a young male patient. This aggressive sarcoma with a benign appearance should always be present in the differential diagnosis of all benign-looking neoplasms of the spindle cells even in the mediastinum.

Patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients un-

derstand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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