

Exceptional Location of a Pleomorphic Leiomyosarcoma: Clinical Case

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Abstract

Case Report

Pleomorphic leiomyosarcoma is a rare entity that is part of the pleomorphic sarcomas with a complex genome with a high metastatic capacity and a usually poor prognosis. The most common location remains the lower extremities but other sites have been reported in the literature, particularly in the chest wall. Leiomyosarcoma (LMS) is an exceptionally rare tumor in the chest wall. Classical LMS may exhibit focal areas of pleomorphic cells; however, the coexistence of a large pleomorphic component allows the tumor to be designated as pleomorphic leiomyosarcoma (PLMS). The first cases of pleomorphic leiomyosarcoma were reported in the literature by Brooks in 1985. The pleomorphic component of these tumors is often heterologous with either cartilaginous differentiation; bony or rhabdomyosarcomatous. Since pleomorphic myogenic sarcomas, including high-grade leiomyosarcomas and rhabdomyosarcomas, have a poorer prognosis hence the importance of an accurate and valid histological diagnosis. The thoracic location of a leiomyosarcoma remains exceptional or even extremely rare and management requires multidisciplinary consultation involving the surgeon; the oncologist and the radiotherapist with additional molecular study to properly detect genetic anomalies in order to consider possible targeted therapy.

Keywords: Sarcoma, Pleomorphic, Thoracic, Surgery, Genomic.

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INTRODUCTION

Primary leiomyosarcomas originating from the chest wall for approximately 1 to 4% of chest wall soft tissue sarcomas [1, 2]. Pleomorphic leiomyosarcoma is a rare entity that is one of the pleomorphic sarcomas with a complex genome, a high metastatic capacity and a usually poor prognosis [1]. The most common location remains the lower extremities but other sites have been reported in the literature, particularly in the chest wall.

OBSERVATION

This is a 62-year-old patient, who quit smoking a year ago without any other notable history, admitted to consultation for a painless left chest mass measuring 7 cm in long axis with a notion of deterioration in general condition reported for 6 months. Chest computerized tomography (CT) reveals a well-defined mass with an encapsulated appearance at the expense of muscular

structures not invading the ribs, with an encapsulated appearance (Figure 1).

A surgical biopsy was done for which the pathological study showed a high-grade sarcomatous process with made of spindle and pleomorphic cells with abundant eosinophilic cytoplasm with polylobed nuclei, sometimes monstrous and dense chromatin with prominent nucleoli. The mitoses are atypical and frequent (Figure 2 and 3), The stroma is fibrous and inflammatory with absence of vascular emboli image.

An immunohistochemical complement was carried out confirming the smooth muscle nature of the tumor cells with diffuse positivity for markers such as desmin, SMA and focal for H caldesmone (Figure 4) which confirmed the diagnosis of a pleomorphic leiomyosarcoma. A surgical excision of the mass was carried out and the surgical specimen was received today at the pathological anatomy department of our structure (Figure 5).

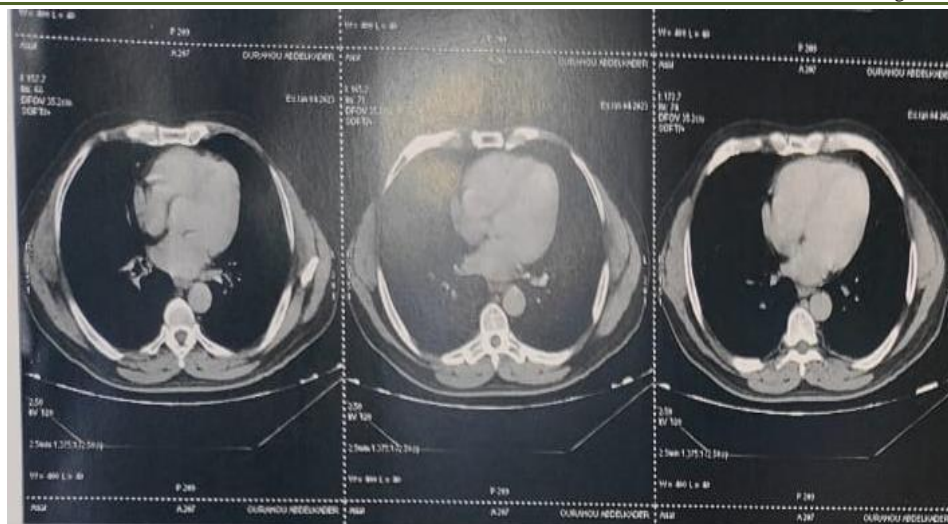


Figure 1: Chest computerized tomography (CT) reveals a well-defined mass with an encapsulated appearance at the expense of muscular structures not invading the ribs

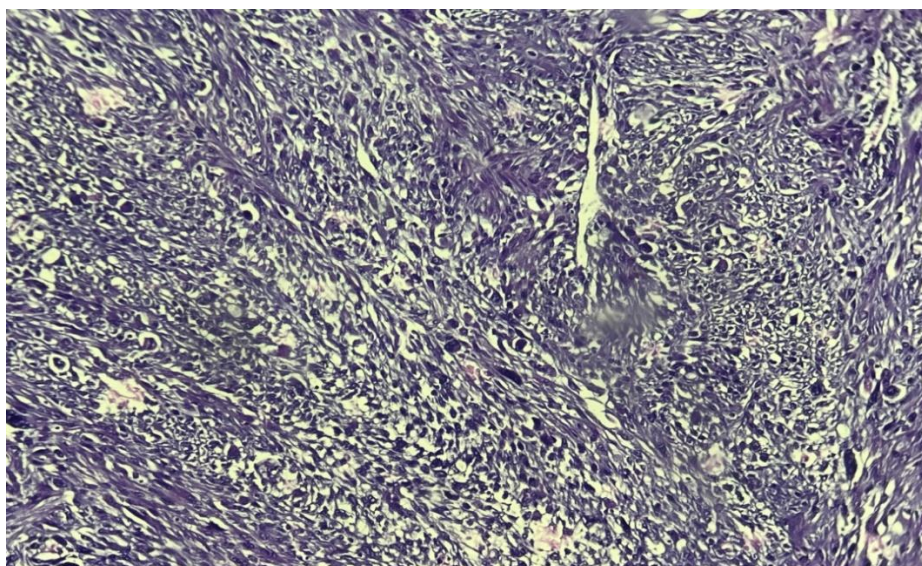


Figure 2: Image showing a high-grade sarcomatous process with made of spindle and pleomorphic cells with abundant eosinophilic cytoplasm with polylobed nuclei

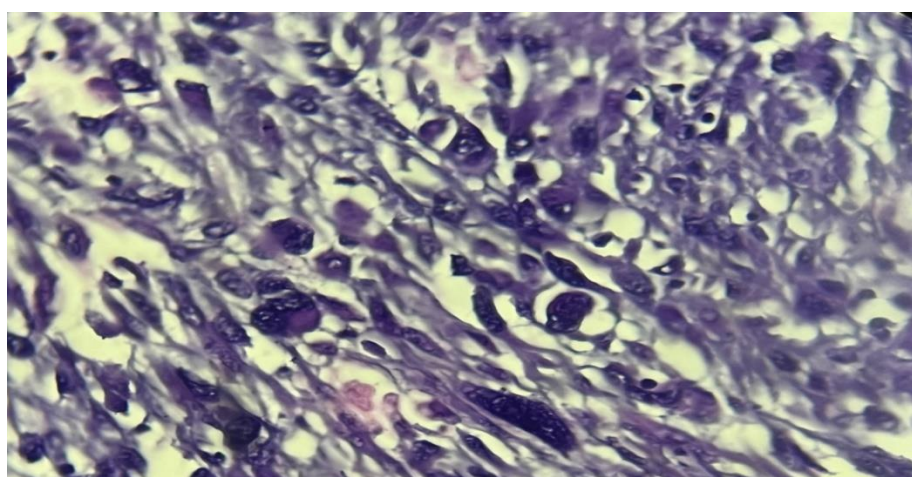


Figure 3: Histological image showing a high-grade sarcomatous process with polylobed nuclei, sometimes monstrous and dense chromatin with prominent nucleoli. The mitoses are atypical and frequent

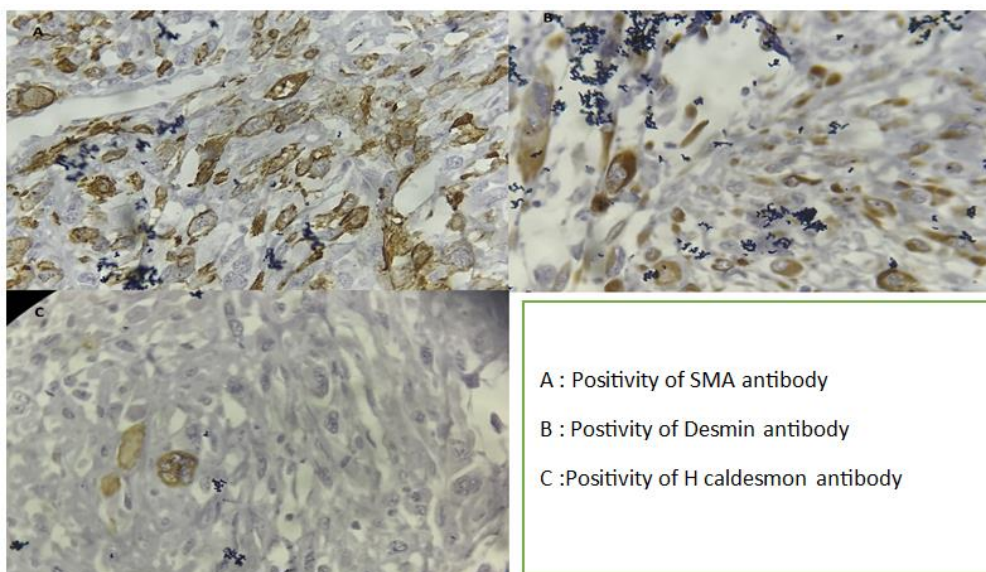


Figure 4: An immunohistochemical complement was carried out confirming the smooth muscle nature of the tumor cells with diffuse positivity for markers such as desmin, SMA and Hcaldesmone



Figure 5: A surgical excision of the mass was carried out and the surgical specimen was received today at the pathological anatomy department of our structure

DISCUSSION

Leiomyosarcoma is a malignant tumor presenting a pure, smooth form with muscular differentiation that accounts for 5-10% of all soft tissue sarcomas. This can occur at any anatomical location, but chest wall leiomyosarcoma is rather unusual [3, 4].

Cellular pleomorphism is defined by a marked variation in the size and shape of the nuclei or even the cytoplasm of tumor cells. Pleomorphism is often worrying but is not always synonymous with malignancy! It is appropriate to confirm the presence of high mitotic activity to confirm malignancy in this context. The diagnostic reasoning must be systematic [5].

Thoracic leiomyosarcomas may cause nonspecific symptoms, such as cough, pain, and shortness of breath, or may be asymptomatic [6]. On CT, they are usually characterized by huge soft tissue masses with heterogeneous enhancement due to bleeding, necrosis and other heterogeneous textures. Larger tumors can also grow against surrounding organs and can sometimes be invasive [7]. Leiomyosarcomas show enhancement on MRI, while they also show elevated FDG uptake on PET-CT [8].

Alternatively, the schwannoma presents a smooth tumor with homogeneous enhancement on CT. Schwannomas may be heterogeneous due to necrosis or hemorrhage within the mass. On MRI, the tumor is iso- to hypo-intense, while it is also enhanced with contrast

media on T1WI and is iso- to hyper-intense on T2WI [9]. Administration of gadolinium causes homogeneous enhancement [10] characteristic of schwannoma. Heterogeneity, FDG uptake by PET-CT, and rapid tumor growth were the only findings that raised suspicion of malignancy.

Since pleomorphic myogenic sarcomas, including high-grade myogenic sarcomas, leiomyosarcomas and rhabdomyosarcomas have poorer prognosis than non-myogenic tumors, the accuracy and classification of the myogenic and non-myogenic nature of these sarcomas is clinically significant [11-13]. Approximately one third of pleomorphic are histologically undifferentiated. sarcomas exhibit myogenic differentiation, and most of these will be found to be pleomorphic leiomyosarcomas when the histomorphology is reevaluated and ancillary studies including immunohistochemistry and/or electron microscopy are performed [14].

The optimal treatment of thoracic leiomyosarcoma has not yet been established. However, due to its resistance to chemotherapy and radiotherapy, surgery remains the first-line treatment [15]. The recommended excision range will be similar to other sarcomas. Kawaguchi *et al.*, Reported that a margin of 2 cm is acceptable for bone and soft tissue sarcomas [16], whereas King *et al.*, Advocated that a resection margin of 4 cm results in a higher resection duration at 5 years which is the survival rate of thoracic sarcoma [17]. The latest report by Mesko et al. recommended a margin of 2 cm for low-grade tumors and a margin of 4 cm with R0 resection for high-grade tumors [18].

However, the resection margin depends on the tumor type and anatomical location: high-grade histology is associated with a higher recurrence rate, and complete resection may be difficult when the tumor is in contact with major organs. In this case, despite its high-grade histology, no local recurrence was observed during 5 years of follow-up. It is therefore considered sufficient to remove the tumor with a margin of 2 cm. Adjusting the resection line when checking with a thoracoscope is helpful to obtain an appropriate margin.

CONCLUSION

The thoracic location of a leiomyosarcoma remains exceptional or even extremely rare and management requires multidisciplinary consultation involving the surgeon; the oncologist and the radiotherapist with additional molecular study to properly detect genetic anomalies in order to consider possible targeted therapy.

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