

Primary chest-wall leiomyosarcoma: a rare mimic of a malignant rib lesion

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PPrimary chest-wall leiomyosarcoma (LMS) is an uncommon, malignant, soft-tissue tumor that most often affects the extremities. Malignant LMS originates from mesenchymal cells with smooth muscle differentiation. It is rare in adults, forming only 7% of all soft-tissue sarcomas (STS), but it is the most common STS. In adults, this type of tumor is usually found in the retroperitoneum and extremities.¹ Chest-wall LMS is rare and most often occurs in men aged 50-70 years.² When LMS is associated with rib destruction, it may mimic a primary bone tumor or metastasis. We present here the case of histologically proven chest-wall sarcoma with associated rib destruction that was initially mistaken on imaging for either a metastasis or primary bone tumor.

Case presentation and summary

A 69-year-old man presented to the emergency department complaining of pain over the right side of the chest. The pain, which was pleuritic in nature, had worsened over the previous 6 months and was severe at presentation. The patient had no fever, shortness of breath, or loss of weight. He had no history of chest trauma or chest wall radiation, and nothing noteworthy was discovered in his medical history. Subsequent test results for hemoglobin, white blood cell count, lymphocyte count, and cardiac enzymes were normal.

A frontal chest radiograph showed an osteolytic destructive lesion involving the posterior right 6th rib (Figure 1). A contrast-enhanced computed-tomography (CE-CT) scan of the chest showed a heterogeneously enhancing, ovoid, soft-tissue mass of 5.6 x 3.6 cm (2.2 x 1.2 in) centred on the postero-lateral right 6th rib, with associated rib erosion. There was another 2.0-cm (0.8-in) subpleural nodule in the left upper lobe (Figure 2). On the

basis of those imaging results, we became concerned about possible primary bone tumor or metastasis. However, the results of a CE-CT scan of the brain, abdomen, and pelvis were normal.

We performed a CT-guided biopsy of the mass centred on the right 6th rib. A histological examination of the biopsy mass showed that cores of the tumor tissue were composed of spindle cells with pleomorphic hyperchromatic nuclei arranged in an interlacing fashion in a fibrous stroma. Many mitoses were identified, of which some were atypical. Immuno-histologically, the tumor stained positive for smooth-muscle h-Caldesmon, a selective stain for LMS, but negative for desmin, s-100, and AE 1/3 (Figure 3). Desmin is a mesenchymal marker, and s-100 is a marker for neural tissue. The final histopathological diagnosis on the basis of these results was high-grade primary LMS.

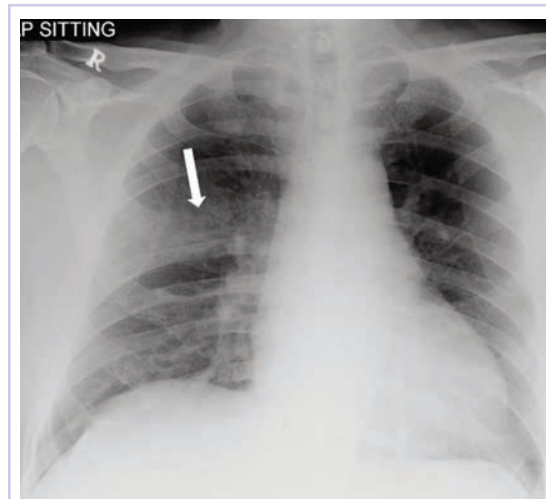


FIGURE 1 Frontal radiograph of the chest shows an osteolytic destructive lesion involving the right, posterior 6th rib (arrow).

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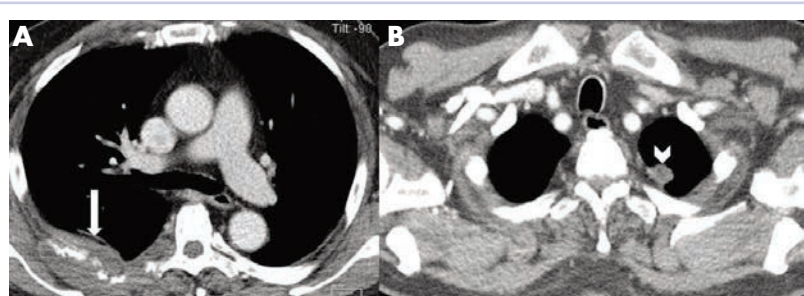


FIGURE 2 **A**, Contrast-enhanced axial computed-tomography chest images obtained in the soft-tissue window show a posterior chest wall mass (arrow) with rib destruction on right side, and **B**, a subpleural nodule in the apical segment of the left upper lobe.

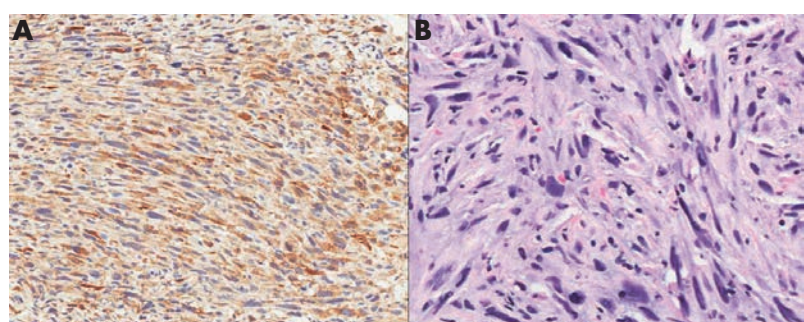


FIGURE 3 **A**, Spindle cell sarcoma with nuclear pleomorphism, hyperchromatism, and mitosis (high-power [hematoxylin and eosin] photomicrograph), and **B**, tumor cells show strong immune-reactivity for h-Caldesmon.

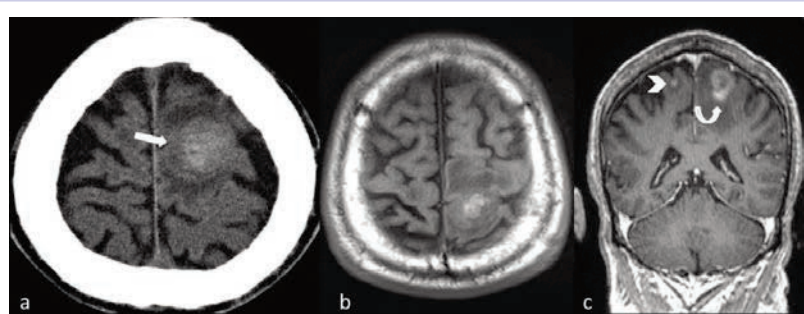


FIGURE 4 **A**, Non-enhanced computed-tomography image of the brain shows a hyper-lesion in the left frontal lobe (arrow). **B**, Non-enhanced axial T1-weighted magnetic resonance image of the brain shows a hyper-intense mass in the left frontal lobe. **C**, Contrast-enhanced coronal T1-weighted image shows enhancing mass lesions in the bilateral frontal lobe (arrows). These imaging features are in keeping with a hemorrhagic metastatic deposit.

The results of a whole-body, positron-emission CT scan showed a large hypermetabolic lesion in the right posterolateral chest wall and in the left lung apex, with standardized uptake values of 24.5 and 8.7 respectively, corresponding to the known tumor and the metastatic deposit. We discussed the case with colleagues at our institution's sarcoma tumor board meeting and in view of the lung metastasis, it was decided to start the patient on palliative che-

motherapy (gemcitabine) and radiotherapy.

Six months after the patient's initial presentation to the emergency department, he presented to the emergency department a second time after he had a fall, and he complained of weakness in his right side, specifically in his right upper and lower limbs. A non-enhanced CT scan of the brain showed an ill-defined hyper-dense mass in the left frontal lobe (Figure 4A). Subsequent contrast-enhanced magnetic-resonance imaging (MRI) of brain confirmed a hyper-intense mass in the left frontal lobe on T1-weighted images (Figure 4B). It showed heterogeneous enhancement on post-contrast T1-weighted images with another small enhancing lesion in right frontal lobe (Figure 4C).

The imaging features were in keeping with hemorrhagic metastasis. In this second admission, a repeat non-contrast CT of the thorax, abdomen, and pelvis revealed an increase in the size of the right-side chest-wall mass and left lung metastatic deposit (Figure 5A, B). There was a new peritoneal nodule in the left hypochondriac region, probably a metastatic deposit (Figure 5C). In view of disease progression, the patient was continued on palliative chemotherapy only (gemcitabine and vinorelbine). The patient died a few weeks later of progressive disease.

Discussion

Primary LMS of the chest wall is quite rare, accounting for 1%-4% of soft-tissue chest-wall sarcomas.³ These tumors are typically painful and manifest as large, palpable, rapidly growing masses.²

Plain radiographs may be useful to detect and localize the lesion.² Both CT or MRI are useful to characterize and define tumor extent. On CT, LMS may show a range of findings, from a poorly marginated soft-tissue mass to a mass with areas of necrosis or cystic changes. It sometimes may be associated with involvement of adjacent structures like the underlying rib, pleura, or lung. These tumors exhibit heterogeneous enhancement. CT is better than plain radiographs and MRI for detecting tumoral calcification and cortical destruction.²

MRI usually reveals a large lobulated mass with relatively low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. After intravenous administration of a contrast agent, the tumor exhibits an enhanced rim and a central area of low signal intensity.²

It is difficult to distinguish the various histological subtypes of chest wall sarcomas on the basis of imaging findings, because most of them present as large, infiltrative masses with non-specific imaging findings.² The eventual

diagnosis invariably lies in histological analysis. The role of the radiologist is to diagnose, evaluate disease extent, and help determine whether the tumor is resectable. However, a radiologist must always look for subtle clues that may help narrow the differential diagnosis, such as the presence of intratumoral fat, osteoid, or chondroid matrix, which suggest liposarcoma, osteosarcoma, or chondrosarcoma, respectively.²

Treatment options are local excision, radiation, and chemotherapy.⁴ A high-grade LMS may require chemotherapy and radiation followed by surgical resection. Adjuvant chemotherapy may be offered in selected cases. Low-grade LMS is often treated with surgical resection alone.¹ In the present case, the patient had high-grade LMS with a metastatic lung deposit, so on his second presentation, he was started on palliative chemotherapy and radiotherapy.

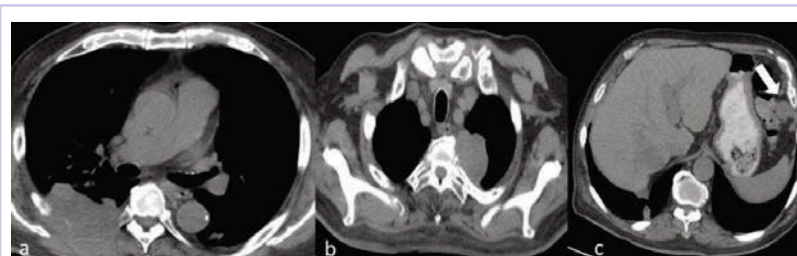


FIGURE 5 **A** and **B**, Follow-up non-enhanced axial computed-tomography images of the thorax and abdomen show increased size of the right-side chest wall mass and left lung metastatic deposit. **C**, There is a new peritoneal metastatic deposit in the left hypochondriac region (arrow).

In conclusion, primary chest-wall LMS with associated bone destruction may mimic metastasis or a primary bone tumor on imaging studies and pose diagnostic challenges. Radiologists need to be aware of this entity to avoid this potential diagnostic pitfall and help guide management.

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