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Leiomyosarcoma: a rare presentation as multifocal lesion

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Leiomyosarcoma is a rare type of connective tissue cancer, accounting for 5–10% of all soft tissue sarcomas. We present a case of leiomyosarcoma as unusual multifocal presentation. Retroperitoneal, mediastinal, pulmonary, uterine and bony regions were all involved at the time of presentation. The liver was normal without detected lesions. A 50-year-old lady presented to the emergency department with a history of right shoulder pain for 4 days. Right shoulder X-ray was done and showed a mediastinal mass at the edge of the film (Figure 1A). The patient noticed a growing abdominal mass over the past few months but didn't seek any medical advice.

Multiple imaging studies were performed including chest radiograph, chest, abdomen and pelvis CT scan with intravenous contrast, cervical and lumbar spine MRI with contrast. The investigations showed a multilobulated heterogeneously enhancing mediastinal mass with multiple hypoenhancing/necrotic areas (Figure 2A&B). It caused destruction and infiltration of the adjacent D1 vertebral body and directly extended through the right nerve root exit foramen to the spinal canal at the same level. Multilobulated lung parenchymal soft tissue mass involving the right lower lobe (Figure 2C). Multiple lung nodules likely metastatic in nature (Figure 2D). A third mass noted at the retroperitoneal region (Figure 3A&B) with similar characteristics to the aforementioned right lung mass. Its epicenter at the left upper aspect of the peritoneum with no local invasion. A fourth mass was found in the uterus with similar characteristics of other primary tumors (Figure 3C). Histopathology of ultrasound-guided biopsy of the retroperitoneal mass confirmed the diagnosis of high grade leiomyosarcoma (Figure 6).

Keywords: Leiomyosarcoma; soft tissue sarcomas; multifocal; Retroperitoneal; mediastinal; pulmonary

Biography:

Dr. Aftan work in Rashid Hospital- Dubai- UAE. Former clinical MSK fellow in Royal National Orthopaedic Hospital- London. Former radiology resident in Jordan University Hospital- Amman-Jordan.

Figures

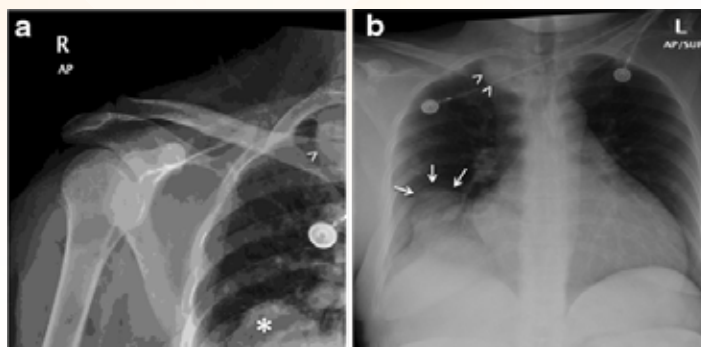


Figure 1. (A) AP view of the right shows a soft tissue opacity projecting over the bottom right corner of the film (white asterisk), another mediastinally based mass (arrow head).

(B) AP view portable chest X-ray confirms the presence of masses in the lower zone of right lung (white arrows) and the above mentioned mediastinal mass (arrow heads). AP, anteroposterior.

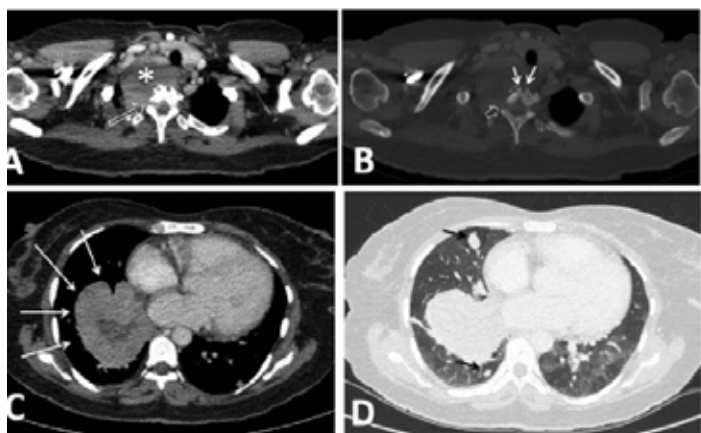


Figure 2. CT chest with i.v. contrast. (A) Mediastinal window at the level of lung apex shows multilobulated heterogeneously enhancing soft tissue mass with multiple hypo-enhancing/necrotic areas (white asterisk). It directly extends through the right nerve root exit foramen to the spinal canal at the same level (long white open arrow). (B) Bone window at the same level shows posterior extension to the adjacent D1 vertebral body causing infiltrative lytic bony changes (white arrows) and directly extending through the right nerve root exit foramen to the spinal canal at the same level (white open arrow). (C) soft tissue window at a lower level shows another multilobulated soft tissue mass with central necrotic areas involving the medial, anterior and basal aspects of the right lower lobe. It abuts but does not invade the adjacent pericardium (long white arrows). (D) Lung window shows multiple nodules (short black arrows) likely metastatic in nature.

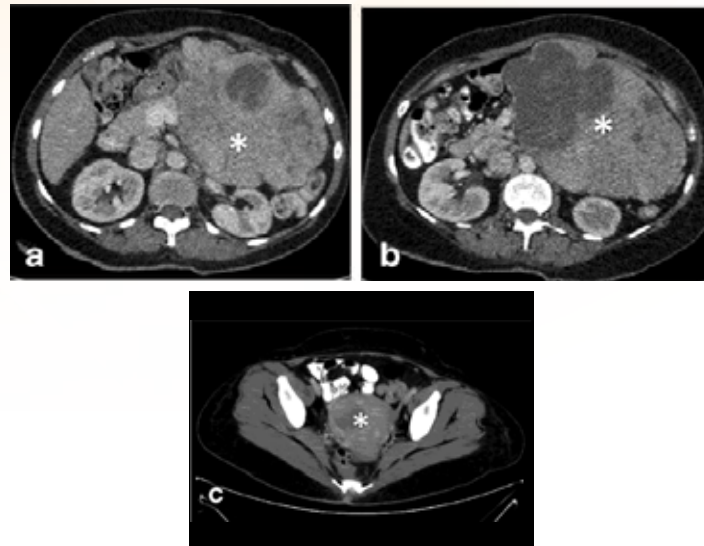


Figure 3. Abdomen and pelvis CT scan with IV contrast axial cuts. (A, B) Multilobulated soft tissue mass with necrotic center (White asterisk). Its epicenter at the left upper aspect of the peritoneum. It displaces but not invades the adjacent bowel loops, vascular structures and pancreas. (C) At a lower level shows another lesion with similar radiological characteristics in the uterus (white asterisk).

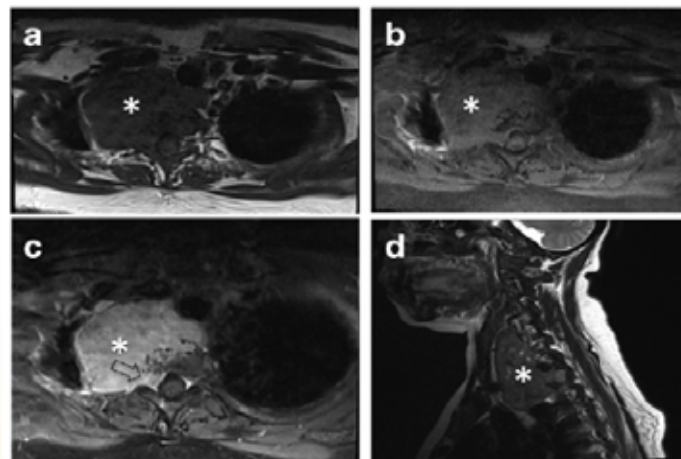


Figure 4. MRI. The mass is labeled with white asterisk. (A) Axial T1 sequence shows heterogeneously isointense signal relative to muscle signal. (B) Axial STIR sequence shows heterogeneously mildly hyperintense mass relative to muscle signal. STIR, short tau inversion recovery. (C) Axial T1 sequence post contrast shows heterogeneous hyperenhancement with involvement of D1 vertebral body and extension into the spinal canal through the right neural foramen (open black arrow). (D) Sagittal T2 sequence shows its cranial extension to cricoid cartilage level.



Figure 5. MR lumbar spine T2 sequence sagittal cut shows similar soft tissues lesion within the uterus (long white arrows) with central area of necrosis (open white arrow).

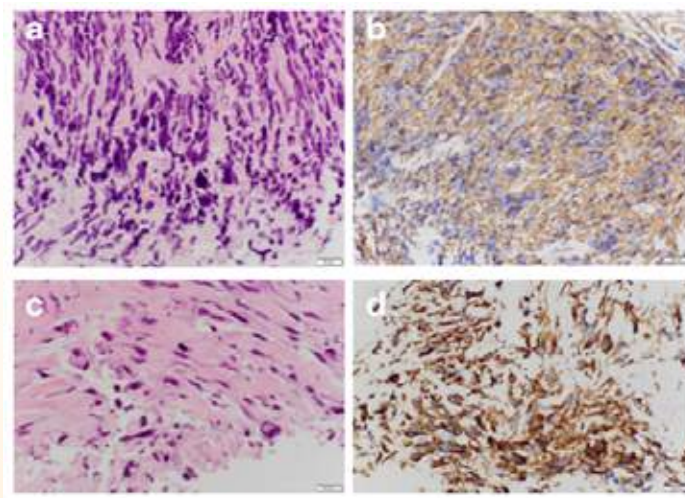


Figure 6. Histopathology of ultrasound guided biopsy of the retroperitoneal mass. (A) High power photomicrograph shows spindle cells, hyperchromatic nuclei, aneuploidy, pleomorphism and atypia. (B) SMA stain shows positive smooth muscle tumor. (C) Mitosis. (D) smooth muscle marker caldesmon. Other markers for other spindle cell tumors were negative. MDM2 test for de-differentiated liposarcoma is negative. Tests for lymphoma, nerve sheath tumor, solitary fibrous tumor is negative. SMA, smooth muscle actin.