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Case Report

Mediastinal liposarcoma: Case report with radiology review ☆

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ABSTRACT

De-differentiated primary mediastinal liposarcomas account for less than 1% of all liposarcoma pathology. We report the case of an 82-year-old male who was suffering from progressive dysphagia, shortness of breath, and dysphonia for a period of 2 months. A CT scan of the chest with contrast revealed a large heterogeneously enhancing posterior mediastinal mass extending into the posterior soft tissues of the neck, abutting bilateral carotid arteries, and displacing the trachea and esophagus. Treatment chosen for our patient was surgical resection followed by adjuvant radiation therapy which resolved the patient's presenting symptoms. The insights gained through the diagnosis, management, and treatment of our patient can be utilized to approach this type of rare neoplasm.

Introduction

Liposarcomas are soft-tissue neoplasms that arise from primitive mesenchymal cells and are currently classified by the WHO into 5 different histological subtypes: well-differentiated, pleomorphic, myxoid, round cell, and dedifferentiated [1]. We present a rare case of a dedifferentiated mediastinal liposarcoma in an 82-year-old male patient presenting with shortness of breath, dysphagia, and dysphonia. Typically, liposarcomas are found in the extremities (75%) and retroperitoneum (20%). Mediastinal liposarcomas however account for less than 1% of all liposarcoma pathology [1]. These tumors can be detected by imaging such as X-ray, computed tomography (CT), or magnetic resonance imaging (MRI) but definitive diagnosis can only be made by histopathology. Mediastinal liposarcomas are generally insidious and typically manifest with symptoms of cough, chest pain, dyspnea, dysphagia,

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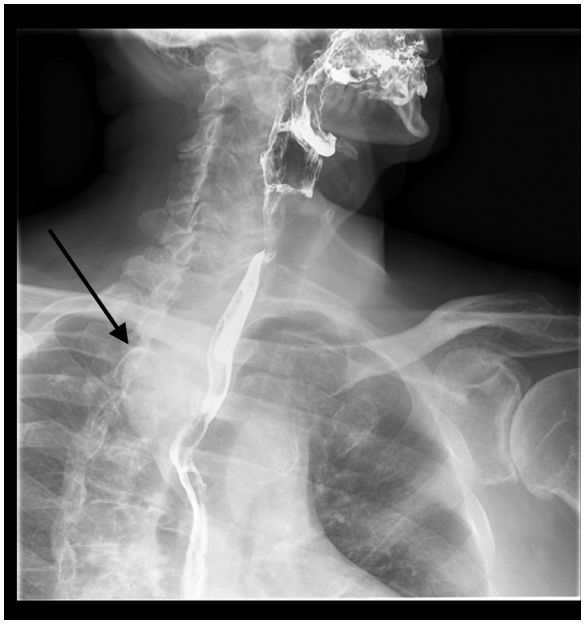


Fig. 1 – Esophagram in right anterior oblique demonstrates an opacity at the super mediastinal region (black arrow) with mild compression of the esophagus concerning for a mass.

and dysphonia [2]. In severe cases, the mass effect caused by these tumors can cause shortness of breath, tachypnea, tachycardia, hypotension, and orthopnea [3]. Currently, surgical resection is the mainstay of treatment and confers the best overall prognosis to patients [4].

Case

An 82-year-old male with a history of shortness of breath, dysphagia, and dysphonia presented to the emergency department. On initial presentation, an esophagram was performed which showed a superior mediastinal mass with mild mass effect on the esophagus without discreet luminal narrowing (Fig. 1). Afterwards a CT chest with contrast was ordered for further evaluation. This examination displayed a large posterior mediastinal heterogenous mass with soft tissue and fat components (Fig. 2). The mass abuts the bilateral carotid arteries as well as aortic arch without invasion. The mass extended posteriorly to the prevertebral soft tissue with mass effect anteriorly on the trachea and esophagus. MRI of the neck was performed for further characterization as well as for complete visualization of the mass superiorly (Fig. 3). The MRI showed a large retropharyngeal mass extending superiorly to C3 measuring 15.2 × 9.5 and 4.4 cm.

The patient was referred for surgical evaluation and palpable mass was identified bilaterally, lateral to the trachea. The lesion was excised by the head and neck and thoracic surgical team using a trans-cervical approach. The strap muscles were splayed over the mass which was abutting but not invading the carotid and subclavian arteries bilaterally. The anterior portion of the mass was just posterior to each thyroid gland without local invasion. Laterally and inferiorly, there was no pleural invasion and with a superior mediastinal blunt dissection the entire tumor was removed intact. The patient recovered uneventfully.

Gross pathological findings demonstrated a 460.0 g, 18.0 × 16.0 × 4.0 cm pale-tan-yellow, multilobulated mass, which is covered by a thin, focally disrupted capsule (Fig. 4). The tumor consists of mildly to moderately cellular myxoid neoplasm with areas of necrosis (Fig. 5). The tumor does not penetrate overlying visceral mediastinal pleura. The tumor is positive for MDM-2, S100 (rare cells), while negative for desmin and SOX-10. The tissue submitted for MDM-2 and DDIT-3 Fluorescence in situ hybrydization (FISH) studies reveals MDM-2 amplification and negative DDIT-3, supporting the diagnosis of dedifferentiated liposarcoma.

Discussion

De-differentiated liposarcomas typically present as a primary tumor rather than as a recurrence of a well-differentiated liposarcoma. Currently, the dedifferentiated subtype carries the worst prognosis with a mean age of 63.3 +/- 13.2 years [4]. In regards to location, de-differentiated mediastinal liposarcomas are located in the posterior mediastinum in more than half of the cases, followed by the anterior, middle, and superior mediastinum in order of frequency [4]. Additionally, the most common symptoms in order of frequency were dyspnea, dysphagia, and hoarseness [4]. In congruence with these typical characteristics, our patient had a posterior mediastinal liposarcoma with primary symptoms of dysphagia, dyspnea, and dysphonia.

Initial imaging with a chest radiograph or computed tomography (CT) are important in the workup as they can reveal some basic characteristics of the mass such as size, shape, location, density, and vascularity with contrast uptake. There are many pathologies that can present as a mediastinal mass, some of which are thymic neoplasm, teratoma, lymphoma, neurofibroma, and esophageal cancer. Liposarcomas have nonspecific findings on a radiograph such as abnormal mediastinal contouring and will appear as an inhomogeneous lobulated mass with areas of hypoattenuation representing fat on a CT scan [5]. This requires the use of MRI to allow for better characterization of liposarcomas considering their heterogeneous architecture through differentiation of lipid/water components, pericardial/vascular invasion, and superior soft tissue visualization [5,6]. Imaging features suggesting liposarcoma over a simple lipoma lesion are lesion size greater than 10 centimeters (cm), presence of thick septa, nodular/globular area, and an associated nonadipoase mass [7]. Lipomas appear hyperintense on MRI T1 and T2 weighted images denoting its homogenous fatty architecture, while liposarcomas exhibit low intensity on T1 images with associated larger size and thicker septa [3,8]. An MRI finding in sarcomas on T2 weighted sequences is the “triple sign” due to the inhomogeneity of the lesion which can be represented by low, intermediate and high signal areas within the lesion [9]. In our particular case, CT scan and MRI imaging revealed that the liposarcoma was lo-

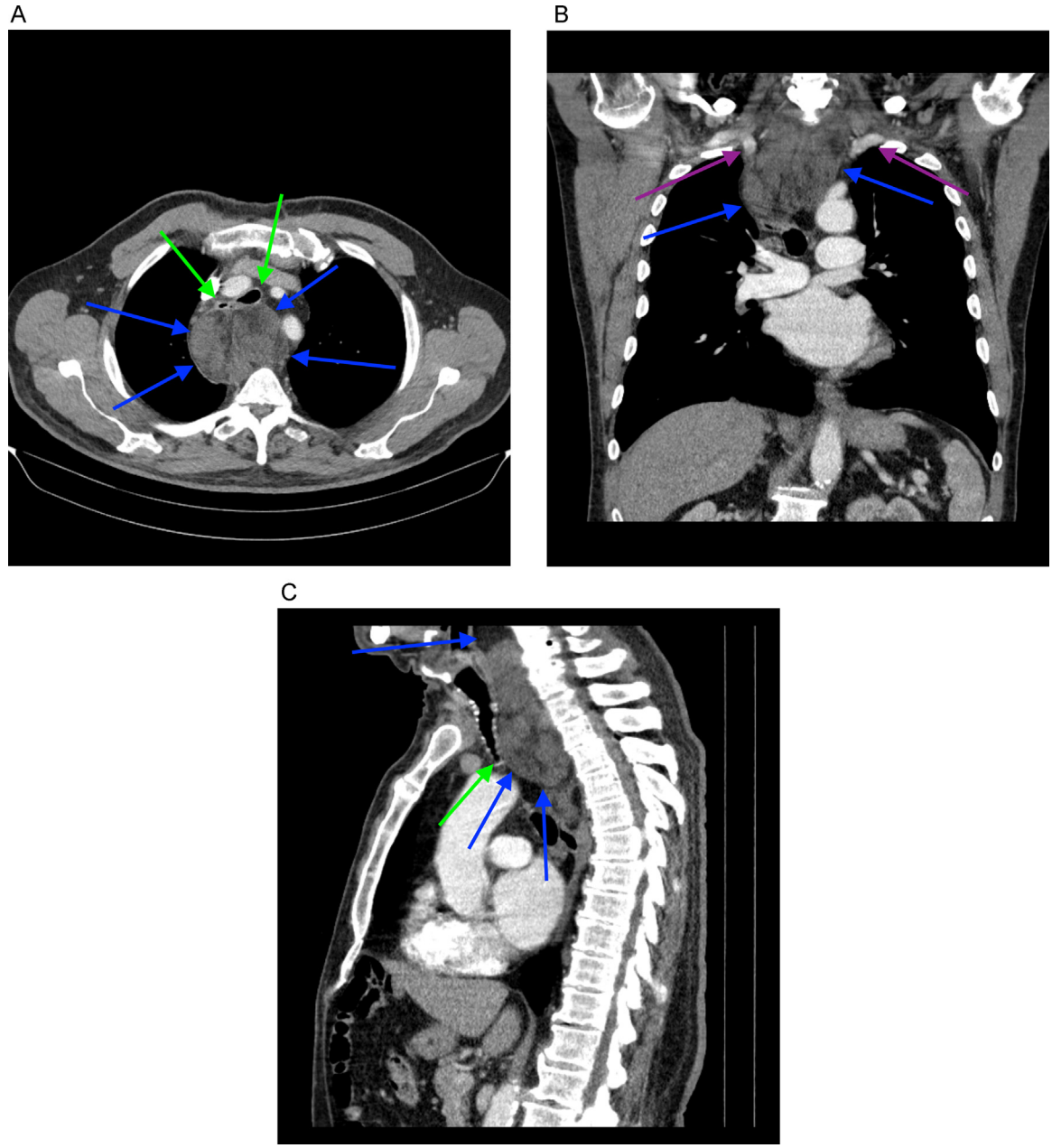


Fig. 2 – Demonstrates a large heterogeneous posterior mediastinal mass (dark blue arrows). Axial (A) and Sagittal (C) views show extension of the mass to the prevertebral soft tissue posteriorly as well as causing mass effect on the trachea and esophagus more anteriorly (light green arrows). The lesion measured 16.6 x 8.6 x 6.5 cm. The coronal view (B) displays the mass tracking adjacent the bilateral subclavian arteries without gross invasion (purple arrow). The mass has heterogeneous attenuation with that of fat and soft tissue density. Although there is significant mass effect, the mass seems to be encapsulated.

cated in the posterior mediastinum extending into the soft tissues of the posterior neck, abuts bilateral carotid arteries, and displaced the trachea and esophagus. These findings highlight the benefits and importance of advanced imaging such as MRI, endoscopic ultrasound, and PET scans all of which can provide valuable information that will ultimately guide management and treatment.

Since mediastinal liposarcomas account for 1% of all liposarcoma pathologies [1], there is no standard treatment algorithm. However, according to a limited number of case reports and series, surgical resection via en-bloc tumor debulking is currently the promoted treatment for de-differentiated or any type of mediastinal liposarcoma [4,10]. In cases of local recurrence, repeat surgical resection with or without adju-

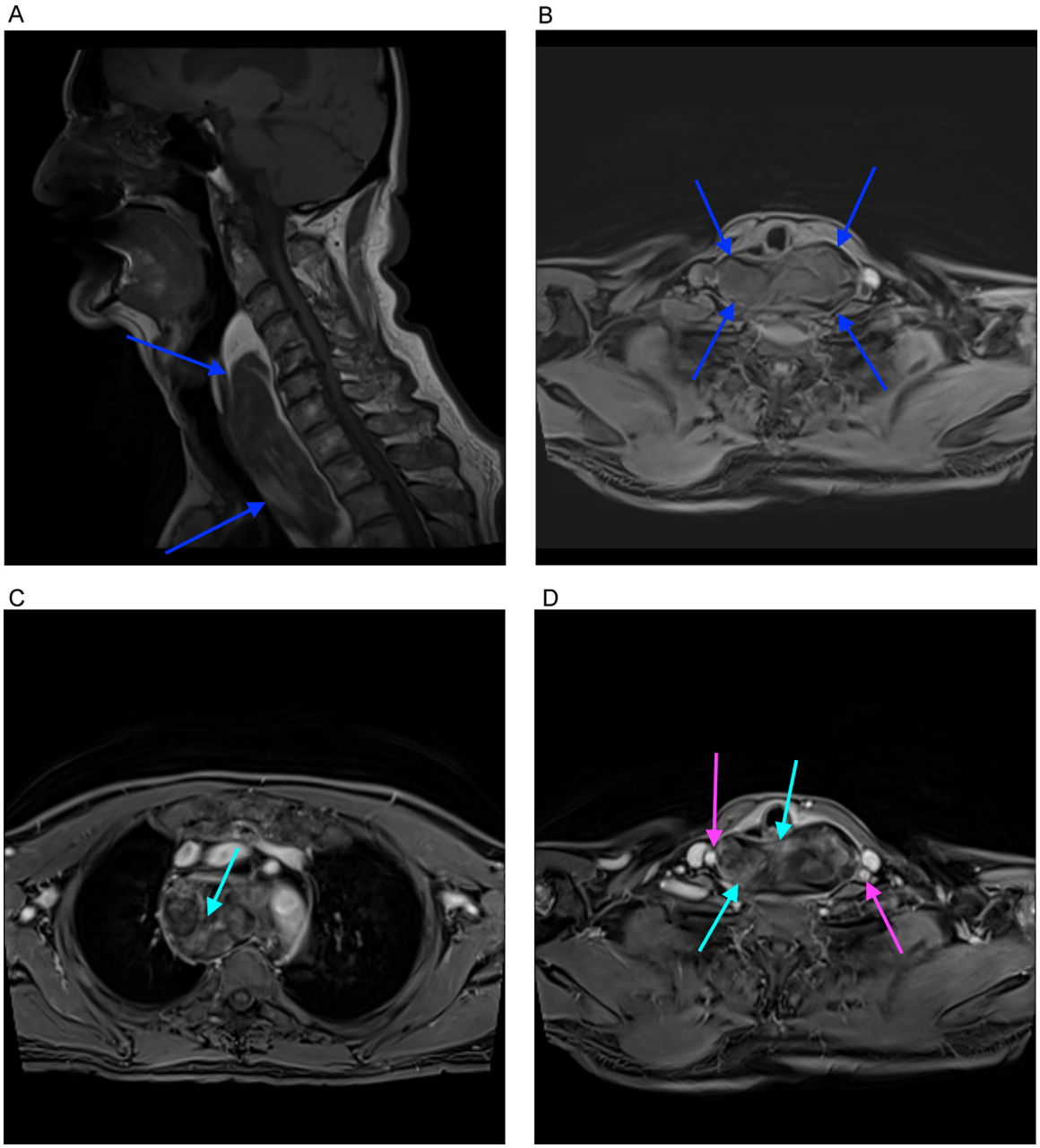


Fig. 3 – Sagittal T1(A) sequence shows a large heterogeneous retropharyngeal mass extending from C3 to the superior mediastinum measuring 17 x 9 x 5.7 cm with extension to the prevertebral soft tissue posteriorly. Anteriorly, the mass extends to the posterior thyroid, trachea, ascending thoracic aorta. Axial pre-contrast (B) and postcontrast (C-E) sequences show heterogeneous enhancement (blue arrows) of the mass with abutment of the bilateral carotid arteries without gross invasion (pink arrows). Further, postcontrast imaging demonstrates a T1 hypointense capsule surrounding the mass (dark green arrows). Out-of-phase (F) and axial STIR (G-J) sequences display a large portion of the mass being fat with india ink artifact (red arrows) and high STIR signal.

vant radiotherapy has proven to be most effective [4,10]. The long-term prognosis after surgery is promising but still needs more supporting data. This highlights the need to report more cases of mediastinal liposarcomas and the implementation of a longer follow up period with these patients. Our patient in particular did undergo adjuvant radiation therapy following surgical resection with good response. Chemotherapy has not shown much promise in efficacy against this type of neoplasm and is only considered in cases of metastatic disease or unresectable tumors [4]. Novel chemotherapies targeted against chromosome 12q14-15, including MDM2 and CDK4 cell cycle oncogenes, are being considered as dedifferentiated liposar-

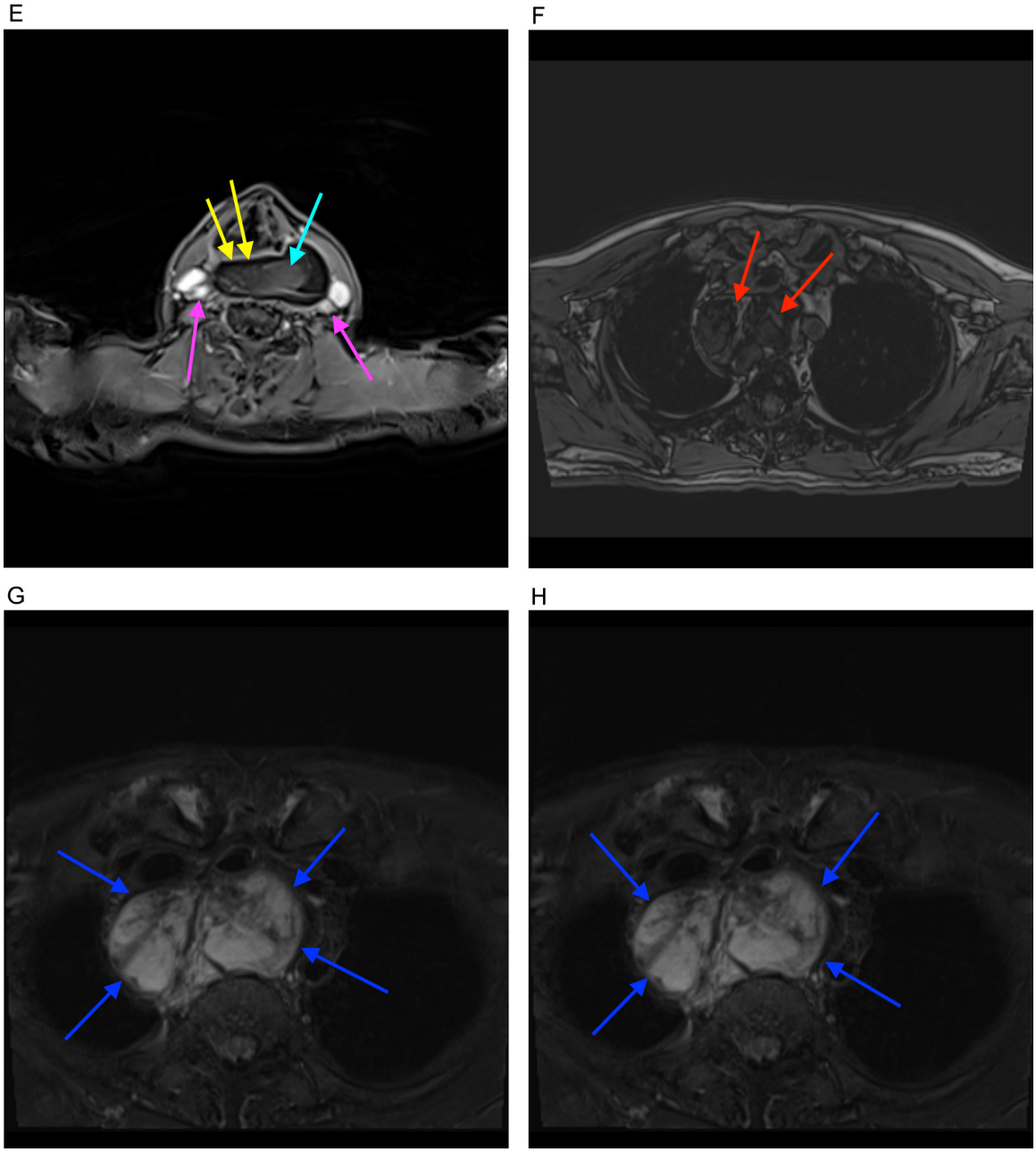
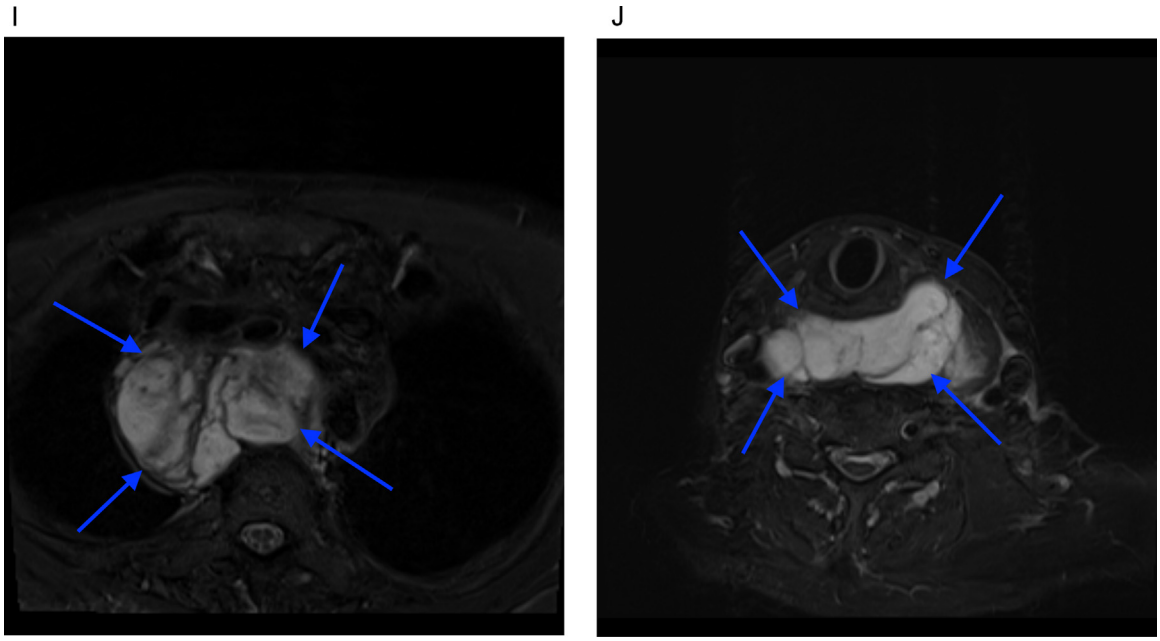


Fig. 3 – Continued

coma typically have high expression of these genetic abnormalities [4]. Our patient's pathology results revealed that his tumor was MDM-2 positive. Perhaps there could be a future role for chemotherapeutics for patients such as ours once their efficacy is proven but further clinical trials and studies are needed to establish any potential role. In conclusion, although mediastinal liposarcoma is rare, the presence of a large heterogeneously enhancing encapsulated lipomatous mass with thick septations in an older patient should put this entity at the top of the differential.



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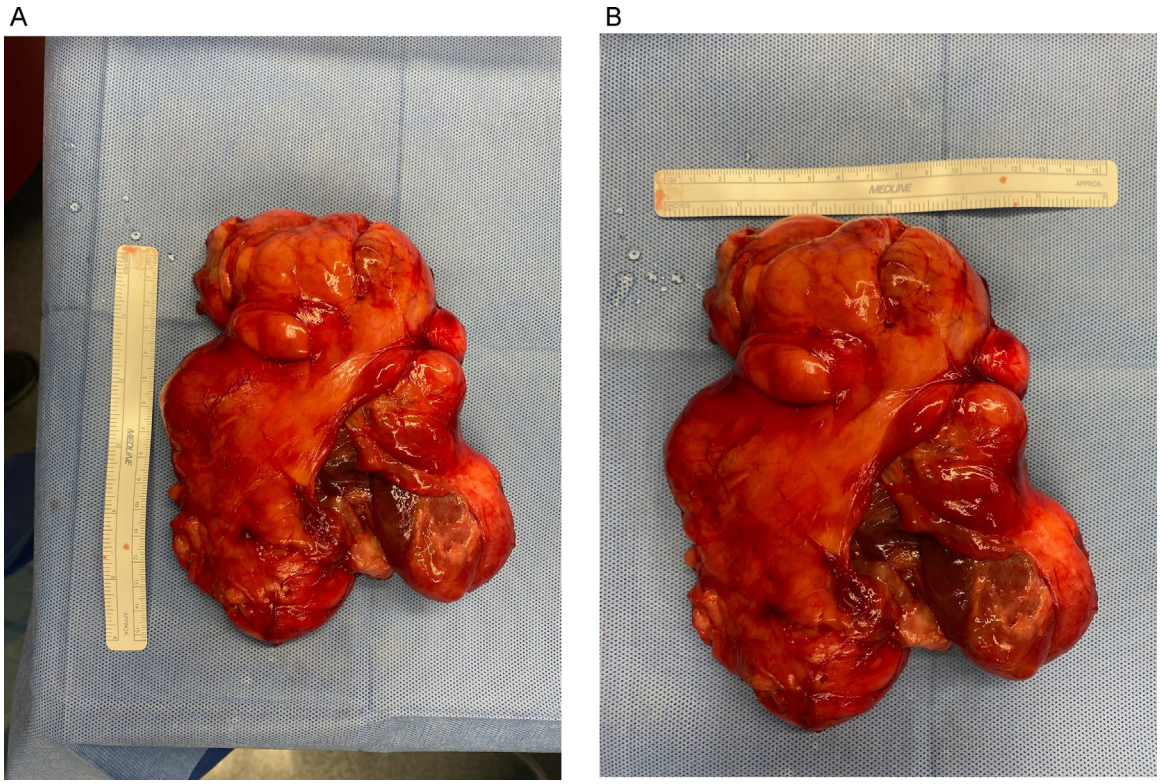

Fig. 4 – Shows an 18.0 x 16.0 x 4.0 cm pale-tan-yellow, multilobulated mass, which is covered by a thin, focally disrupted capsule. No areas of calcification or hemorrhage were identified.

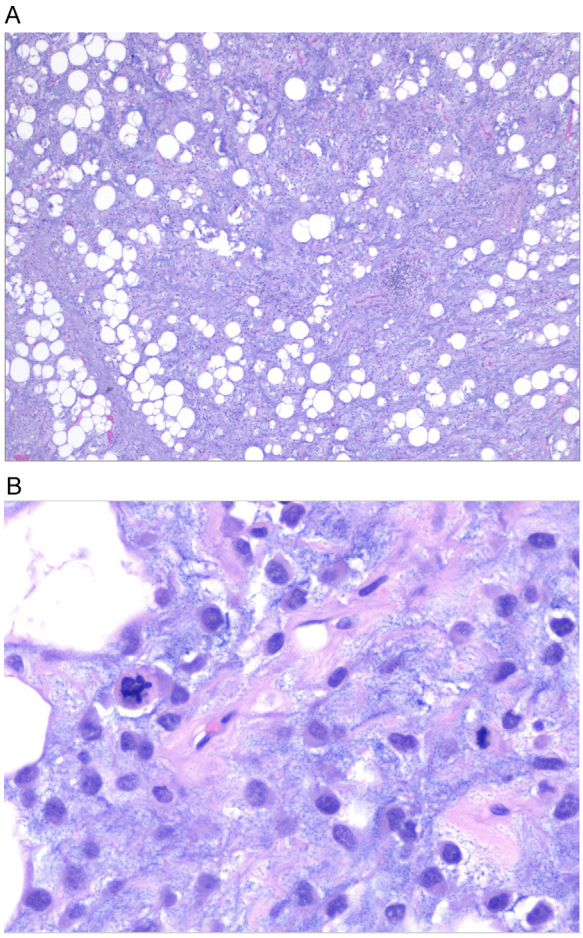

Fig. 5 – Images A (low power photomicrograph at 20 x magnification) and B (high power photomircograph at 40 x magnification). Photomicrographs demonstrate mildly to moderately cellular myxoid neoplasm with areas of necrosis, rare lipoblasts and rare mitotic activity consistent with dedifferentiated liposarcoma consisting predominantly of myxoid fibrosarcoma, low to intermediate grade.

Patient consent

The patient gives consent to be included as the subject of a case report.

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