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INTERESTING MEDICAL IMAGE

Giant Mediastinal Myxoid Pleomorphic Liposarcoma

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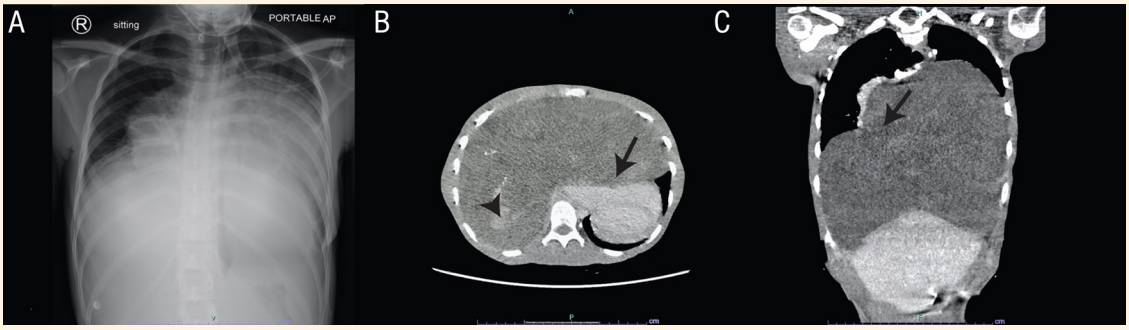

Figure 1: A: Anteroposterior chest x-ray of an 18-year-old female patient showing an abnormal contouring of the mediastinum with a large lesion in the anterior mediastinum and the left lower hemithorax. Hilar overlay sign is noted with both hilar shadows identified through the opacity of the abnormal lesion. **B & C:** Chest computed tomography images with IV contrast in the (B) axial and (C) coronal views showing a large heterogeneous mass occupying the anterior mediastinum, right and left hemithorax with posterior displacement of the heart and atelectasis of the lungs. The mass shows areas of low density (arrow) likely representing fat content and other areas of high density (arrowhead) representing calcification.

An 18-year-old female patient with no past medical history presented to a University hospital in Muscat, Oman, in 2020. She was tachypnoeic, tachycardiac, hypotensive, unable to lie flat and had had progressive shortness of breath for the previous three months.

Her chest x-ray revealed abnormal contouring of the mediastinum [Figure 1A]. Contrast-enhanced computed tomography (CT) of the chest showed a large heterogenous mass occupying the anterior mediastinum, right and left hemithorax with posterior displacement of the heart and atelectasis of the lungs [Figure 1B and C]. The patient underwent urgent surgical resection. The mass was exposed through a clam shell incision exposing the anterior mediastinum and bilateral pleura [Figure 2A]. The mass was yellow, encapsulated and lobulated. It was resected en-block with an intra-operative frozen section confirming negative margins. The mass weighed 3,208 g and was 22 × 18 × 13 cm in size [Figure 2B].

Post-operatively, the patient did well with complete resolution of her symptoms. The final histopathology proved the mass to be high grade malignant pleomorphic myxoid liposarcoma [Figure 3 and 4]. Murine double minute 2 (MDM2) and cyclin-dependent kinase 4 were both negative. She received adjuvant chemotherapy and is under close surveillance by the oncologist.

Consent was obtained from the patient for the publication of the case and images.

Comment

Liposarcoma is a relatively uncommon malignant tumour of adipose tissue that can occur at any site.[1] In rare instances, it arises primarily in the mediastinum.[2] The World Health Organization recently classified adipocytic tumours into: (1) benign; (2) intermediate (locally aggressive); and (3) malignant. Malignant tumors include: (1) well-differentiated liposarcoma (i.e. lipoma-like, sclerosing, inflammatory); (2) dedifferentiated liposarcoma; (3) myxoid liposarcoma; (4) pleomorphic liposarcoma; and (5) myxoid pleomorphic liposarcoma.[2–4]

Myxoid pleomorphic liposarcoma is a new entity in this classification. Unlike other liposarcomas, its most common anatomical site is in the mediastinum followed by the limbs and neck. Histologically, it shows a mixture of both myxoid and pleomorphic liposarcoma.[3] Mediastinal liposarcoma may grow to

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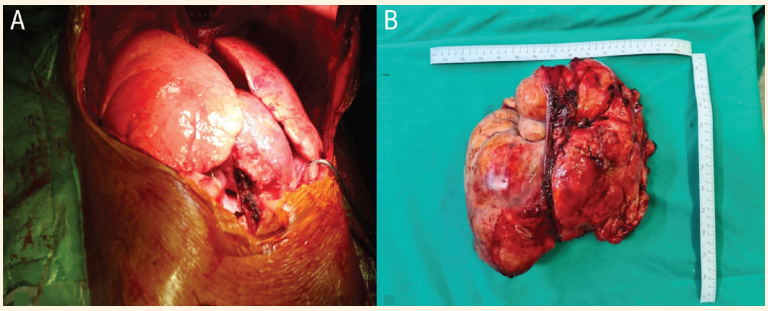

Figure 2: A: Intraoperative photographs showing clam-shell incision exposing the anterior mediastinum and bilateral pleura. **B:** Photograph of the excised mass which was yellow, encapsulated and lobulated measuring 22 × 18 × 13 cm in size.

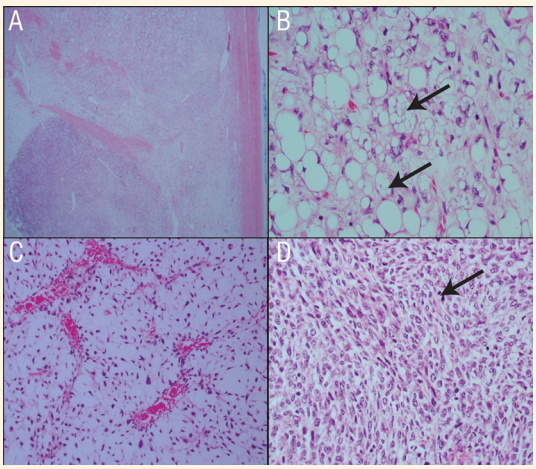

Figure 3: A: Low power examination showed an encapsulated multinodular malignant neoplasm of variable cellularity (hematoxylin and eosin stain, magnification ×40). **B:** lipomatous areas showing atypical multivacuolated lipoblasts (arrows) (hematoxylin and eosin stain, magnification ×100). **C:** other areas showed myxoid stroma containing plump spindle to stellate cells (hematoxylin and eosin stain, magnification ×100). **D:** highly cellular nodules formed of fascicles of pleomorphic spindle cells with frequent mitotic figures (arrow) (hematoxylin and eosin stain, magnification ×200).

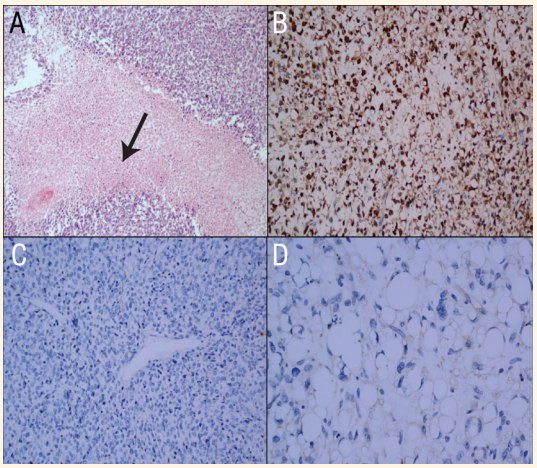

Figure 4: A: Areas of geographic coagulative type necrosis seen (arrow) (hematoxylin and eosin stain, magnification ×100). **B:** S-100 immunostain showed positive staining of tumor cells in the lipomatous areas (magnification ×100). **C:** s-100 immunostain was negative in the cellular pleomorphic areas (magnification ×100). **D:** Tumor cells were negative for cyclin dependent kinase 4 (CDK4) (magnification ×400).

a substantial size before causing any symptoms. It is most commonly found incidentally on chest x-ray. Symptomatic patients may have cough, dyspnoea, dysphagia and chest pain.[5]

Radiologically, the tumour appears heterogenous on a CT scan with a difficulty to differentiate it from other forms of sarcoma. Magnetic resonance imaging (MRI) can differentiate lipoma from well differentiated liposarcoma (WDL).[6] For lipomas, MRI shows high intensity in T1 and T2 weighted images as this represents the uniform structure with fatty tissue. In contrast, high grade liposarcoma show low intensity in T1 images. In addition, liposarcomas tends to be larger and have more thick septa.[7,8] As the entity is rare, there are no published randomised trials that assess different treatment modalities. Current practice is based on case reports and series.[4] Complete surgical resection remains the mainstay for the treatment of myxoid pleomorphic liposarcoma.[8] Wide resection with negative margins is the goal. However, anatomical location plays an important role in whether this is possible. Patients with huge mediastinal mass that present late may be high risk surgical candidates as the compression of vital structures, such as the heart and the lung, may cause critical haemodynamic issues during surgery.[9]

Myxoid pleomorphic liposarcoma has an aggressive clinical presentation with high recurrence rate and distant metastasis following surgical resection; approximately 40% recur after surgery.[8] Thus, a multidisciplinary tumour board is recommended to assess the need for adjuvant radiotherapy with or without chemotherapy after surgical resection.[9] Although 90% of mediastinal WDL show 12q12-15 amplicon that represents amplified oncogenes MDM2 and cyclin-dependent kinase 4, no genetic aberration

has been associated with myxoid pleomorphic liposarcoma.[3,9]

Primary mediastinal myxoid pleomorphic liposarcoma is a rare entity and tends to present at an advanced stage. Recognition of the need for complete surgical resection followed by adjuvant therapy is very important.

AUTHORS' CONTRIBUTION

AHK performed the surgery and EK assisted in the surgery. FAK prepared and reported the radiological images. MR prepared the pathology of the patient and reported the pathological images. AHK and EK drafted and edited the manuscript. All authors approved the final version of the manuscript.

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