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Case Report

Primary liposarcoma of the diaphragm: a rare intra-abdominal mass

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ABSTRACT

Primary malignant tumors of the diaphragm are rare, and primary liposarcoma of the diaphragm is extremely rare. The role of imaging is description of the anatomic relationships of the tumor as well as a suggestion of histologic diagnosis based on the presence of fatty and/or nonfatty components.

Case report

Clinical presentation

A 67-year-old woman with obesity, diabetes mellitus, hypertension, chronic obstructive pulmonary disease, and prior cholecystectomy presented with progressive shortness of breathing and pain in the right lower chest for the past 3 years. Physical examination and laboratory evaluation were unrevealing.

Radiology

A chest radiograph showed right hemidiaphragmatic elevation and/or eventration (Fig. 1A). An ultrasound showed a heterogeneous mass (Fig. 2). Computed tomography (CT) of the abdomen and pelvis with intravenous contrast revealed a large (20 × 18 × 10 cm), encapsulated, subdiaphragmatic, mostly fatty mass with scattered areas of soft tissue nodules and septations, as well as a few coarse calcifications, overall compatible with a liposarcoma (Fig. 3). There was mass effect on the liver and the diaphragm with apparent eventration of the right hemidiaphragm. CT-guided 18G core biopsy was performed (Fig. 4), which showed adipose tissue with pleomorphic lipoblasts suggestive of liposarcoma.

Surgery

Surgical resection was indicated. The operation was performed via a right subcostal and flank incision with the

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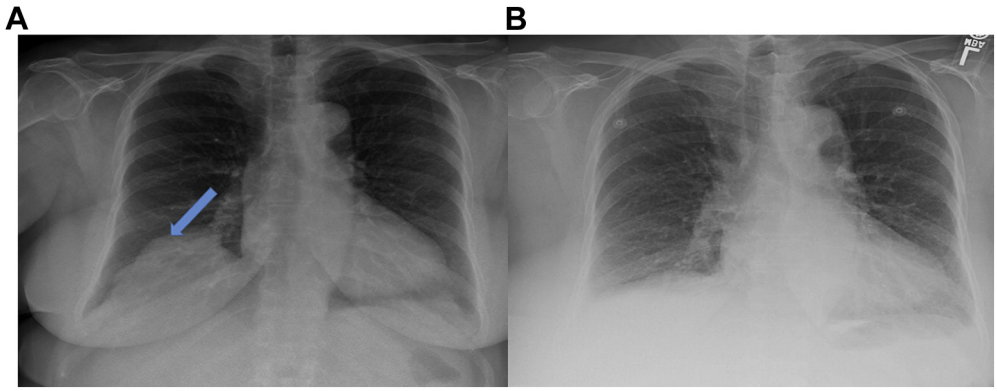

Fig. 1 – Frontal chest radiograph. (A) Right hemidiaphragmatic elevation and/or eventration (blue arrow), otherwise no acute process. (B) Postoperative normal appearance of the right hemidiaphragm.

patient in left lateral decubitus position (Fig. 5). A very large (20 × 18 × 15 cm, 1600 gram) lipomatous mass was found, covered with an intact glistening capsule and inseparable from the right hemidiaphragm. The mass was multilobulated and attached to the diaphragm anteriorly and posterolaterally, which required en bloc full-thickness removal of the diaphragm with the mass in those areas (Fig. 6). The diaphragm was reconstructed by primary repair. The patient recovered well and was discharged without complications.

Pathology

Well-differentiated liposarcoma arising from the diaphragm (Figs. 7-9) with otherwise intact capsule and without angiolymphatic invasion (grade 1; American Joint Committee on Cancer Stage pT2b). There was an associated intramuscular lipoma (3.5 × 2.5 × 1 cm) at the periphery of the main tumor.

Discussion

Epidemiology

Although liposarcoma is the most common soft tissue sarcoma [1,2], primary liposarcoma arising from the diaphragm is extremely rare. A PubMed search up to October 2016 identified a single case report [5]. Typical locations include the retroperitoneum and extremities (up to 75% of cases) [1,3]. There are case reports of other rare locations: orbit [6], oral cavity [7], esophagus [8], small bowel mesentery [9], colon [10], sigmoid mesentery [11], pancreas [12], scrotum [13], and other areas.

Pathophysiology

Liposarcomas are malignant mesenchymal tumors with diverse pathologic appearances, genetics, and natural history [1–4,14]. The 2013 WHO Classification of Soft Tissue Tumors divides all adipocytic tumors into 3 general categories: benign (includes lipomas), intermediate and/or locally aggressive (includes well-differentiated liposarcoma and/or atypical lipomatous tumor), and malignant (dedifferentiated, myxoid, pleomorphic, and not otherwise specified) [1,2]. Well-differentiated liposarcoma (the current case) is an intermediate-type of adipocytic tumor. It is locally aggressive but lacks metastatic potential [1–3]. When located in the superficial tissues, well-differentiated liposarcoma is frequently referred to as atypical lipomatous tumor [3,15]. Its 3 subtypes include adipocytic (lipoma-like), sclerosing, and inflammatory tumors. Although spindle cell liposarcoma is still described under atypical lipomatous tumors, its lack of MDM2 immunopositivity or 12q15

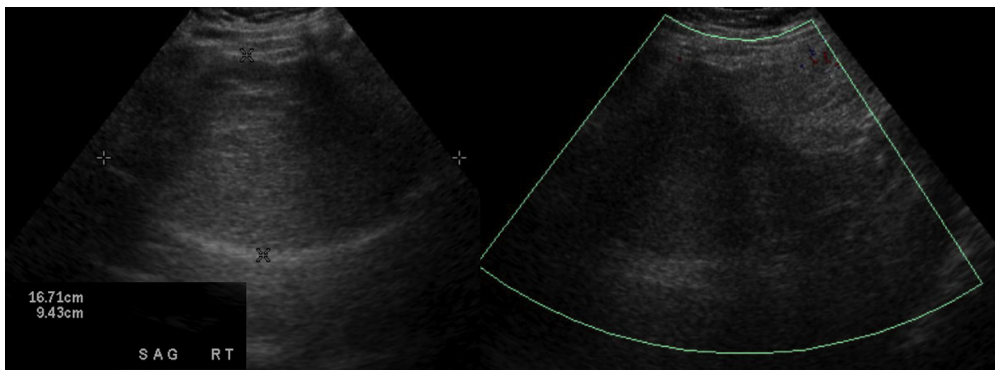

Fig. 2 – Grayscale and color Doppler ultrasound demonstrates a heterogeneous soft tissue mass without significant vascularity, overall nonspecific in appearance.

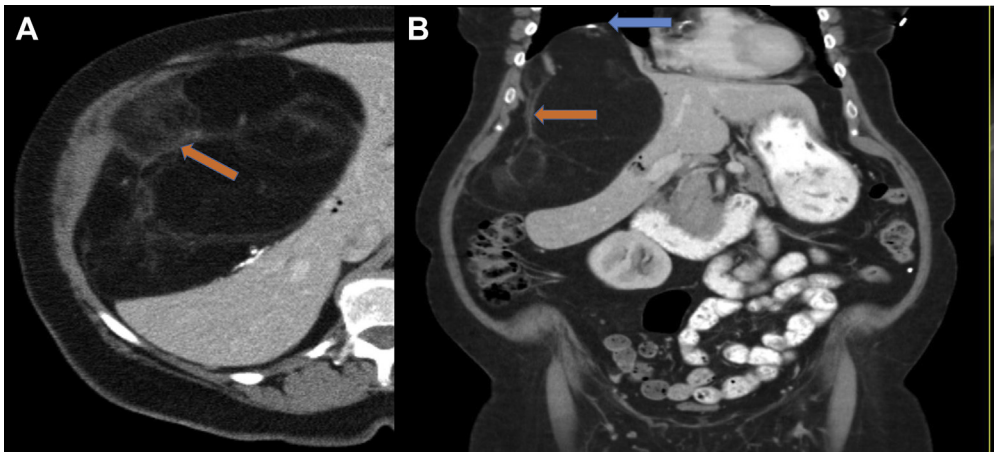

Fig. 3 – Computed tomography (CT) of the abdomen and pelvis with contrast (A) axial and (B) coronal reformats. Large, well-marginated fatty mass (>75% fat) with scattered nonlipomatous components including thick septations and/or nodules (orange arrow) and calcifications (blue arrow).

amplification may suggest that it is a different type of tumor [1]. Although up to 10% of well-differentiated liposarcomas can dedifferentiate into malignant liposarcomas, it is important to point out that liposarcomas do not arise from lipomas. Though malignant transformation of lipomas has been reported, it is unclear if this was due to sampling error and/or misdiagnosis [1,5].

Clinical manifestation of well-differentiated liposarcomas is usually nonspecific and depends on the location and size of the tumor. A painless, slow-growing mass is a common feature.

Diagnosis

Imaging with CT or magnetic resonance should be performed, which can suggest the diagnosis, provide macroanatomic localization, and evaluate for additional lesions. Well-differentiated liposarcomas typically appear as large, encapsulated, predominantly lipomatous (>75% of composition) masses with scattered nonlipomatous components [5]. These include scattered thick (>2 mm) connective tissue septations or nodules, which may enhance (therefore, increasing suspicion for malignancy). Calcifications and metaplastic ossifications are also present in 30% of cases. Ultrasound can also demonstrate a well-defined, heterogeneous mass, but is neither specific nor sensitive in confirming fatty components [5]. Image-guided core needle biopsy is also indicated before further intervention.

Differential diagnosis includes complex lipomas, other soft tissue tumors, and inflammatory pseudotumors.

Treatment is surgery to achieve oncologically appropriate margins [16]. If complete resection (R0) is achieved, no

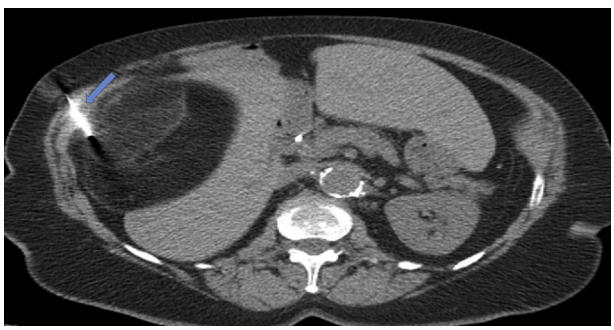

Fig. 4 – CT-guided core needle biopsy of the area with septations and/or nodules (blue arrow).

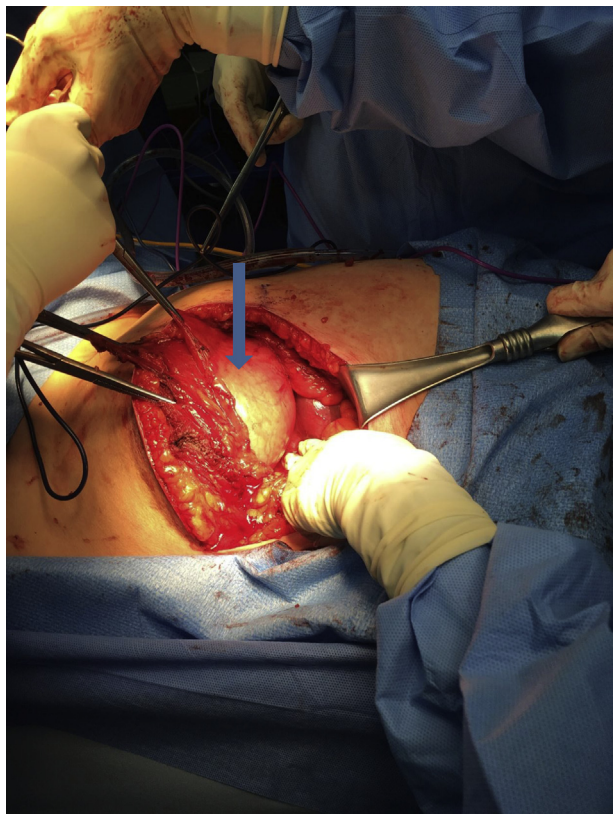

Fig. 5 – Right lateral subcostal approach was used to access the mass (blue arrow).

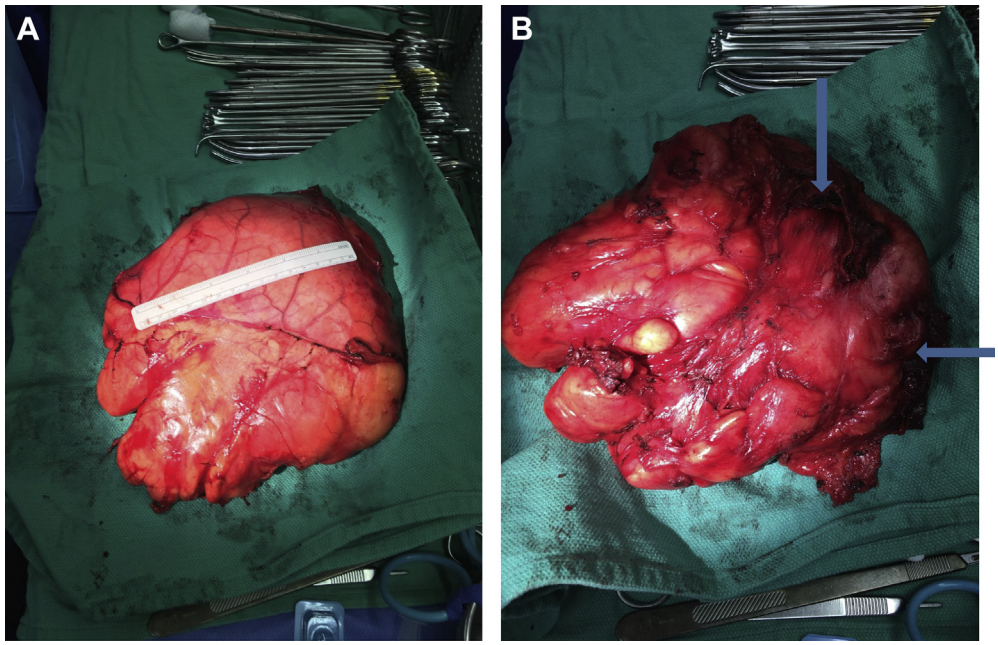

Fig. 6 – (A) Large resected tumor with glistening capsule. (B) The lobulated diaphragmatic surface of the mass is inseparable from the right hemidiaphragmatic muscle fragment, which was resected en bloc with the mass (blue arrows).

further therapy is necessary, although intermittent monitoring with chest, abdomen, and pelvic CT is suggested. For grossly positive margins (R2), re-excision is usually recommended. If excision with negative margins cannot be achieved because of location, or unacceptable functional sequela, radiation therapy may be offered. If the tumor seems unresectable, downstaging with preoperative radiation treatment or a combination of radiotherapy and chemotherapy is an option. Adjuvant postoperative chemotherapy has shown marginal benefit in recurrence-free survival for extremity sarcomas. Chemotherapy with single or combination agents has been used for advanced, unresectable, or metastatic tumors [16].

Prognosis is good in general, but directly depends on completeness of resection and achieving microscopically negative margins. Well-differentiated liposarcoma can recur locally if resection is incomplete, but there is no metastatic potential (unless dedifferentiation occurs).

In conclusion, liposarcomas are common tumors that typically originate in the retroperitoneum and extremities, but may arise in unexpected locations. The extremely rare primary diaphragmatic liposarcoma presented here is such an entity. Imaging is an important part of the diagnostic workup that can demonstrate the lipomatous nature of the tumor. Surgical excision with oncologically appropriate margins is the gold standard of treatment.

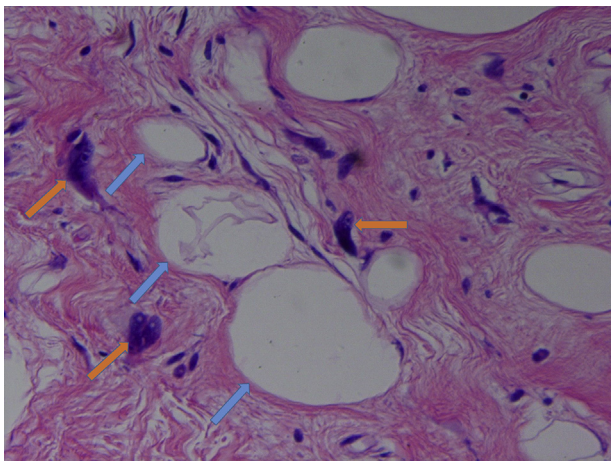

Fig. 7 – Well-differentiated liposarcoma. Adipocytes show variation in size and shape (blue arrows). Enlarged bizarre hyperchromatic stromal cells are present (orange arrows). (Hematoxylin-eosin staining, ×400).

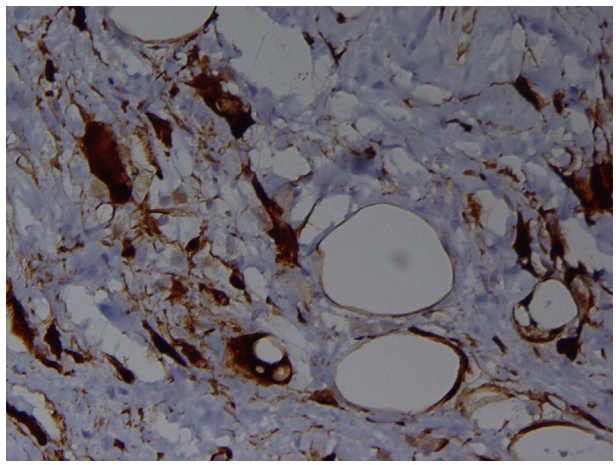

Fig. 8 – Well-differentiated liposarcoma. CDK4-positive immunohistochemical staining confirms the diagnosis; CDK4 is amplified in well-differentiated liposarcoma (×400).

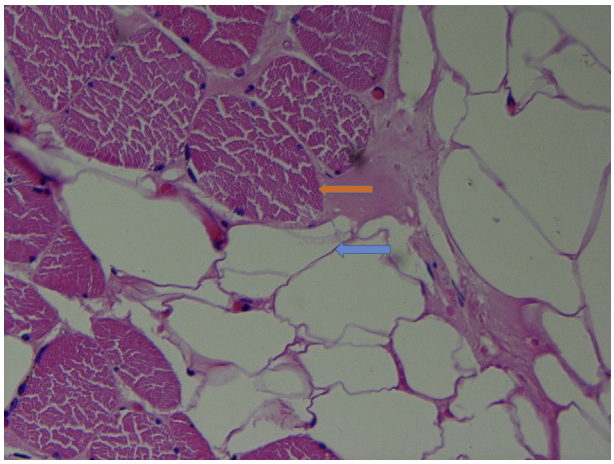

Fig. 9 – Well-differentiated liposarcoma. Adipocytes (blue arrow) intermixed with diaphragmatic skeletal muscle fibers (orange arrow) demonstrate that the liposarcoma originated from the diaphragm. (Hematoxylin-eosin staining, original magnification ×200).

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