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Case Report

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

Károly András Virágh MD[a,*], *Sergey Cherneykin MD*[b], *Roy Oommen MD*[c], *Zahra Shafaee MD*[c]

[a] *Department of Radiology, Columbia University, NewYork-Presbytarian/Lawrence Hospital Center, 55 Palmer Road, Bronxville, NY 10708, USA*

[b] *Department of Pathology, Columbia University, NewYork-Presbytarian/Lawrence Hospital Center, Bronxville, NY, USA*

[c] *Department of Surgery, Columbia University, NewYork-Presbytarian/Lawrence Hospital Center, Bronxville, NY, USA*

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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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* *Corresponding author.*
E-mail address: kav2107@columbia.edu (K.A. Virágh).

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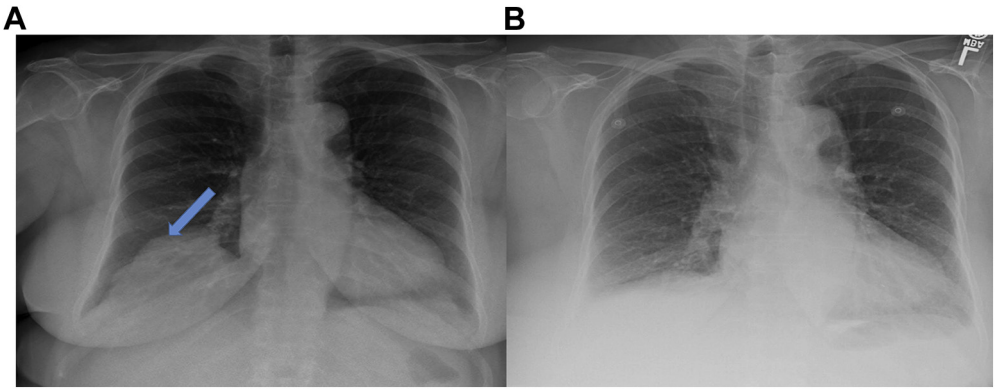

Fig. 1 – Frontal chest radiograph. (A) Right hemidiaphragmatic elevation and/or eventration (blue arrow), otherwise no acute process. (B) Postoperative normal appearance of the right hemidiaphragm.

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

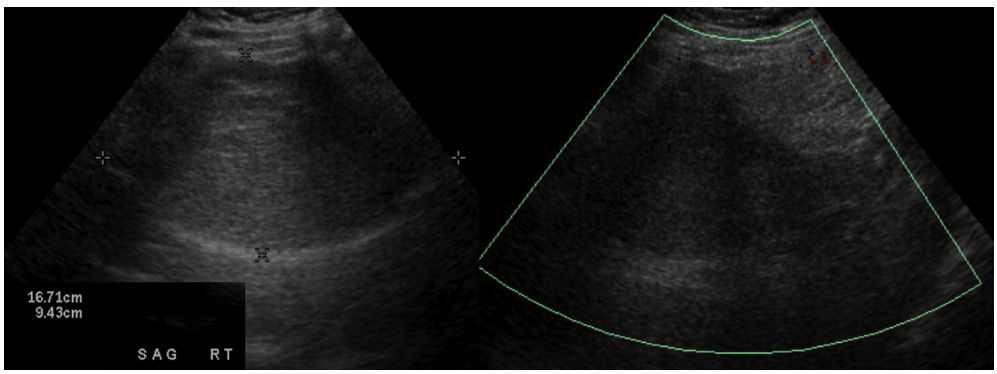

Fig. 2 – Grayscale and color Doppler ultrasound demonstrates a heterogeneous soft tissue mass without significant vascularity, overall nonspecific in appearance.