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Case Report

Diaphragmatic liposarcoma with gall bladder invasion: CT and MRI findings

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ABSTRACT

Primary liposarcoma originating from the diaphragm is an extremely rare case. Seventy-four-year-old male presented to emergency department with worsening right upper quadrant abdominal pain with dyspnea. Contrast-enhanced computed tomography of the abdomen demonstrated lobulated mass in right hemidiaphragm exhibiting mass effects to adjacent structures compressing gall bladder. Magnetic resonance imaging of the abdomen showed diaphragmatic mass with heterogeneous signal intensities with partial diffusion restriction. Surgical removal was performed and histology confirmed dedifferentiated liposarcoma arising from the diaphragm with gall bladder invasion.

Introduction

Liposarcoma is a mesenchymal origin malignant tumor that can arise from any fat containing region of the body [1–3]. Clinical presentation is nonspecific and variable since symptoms may develop from mass effects of the tumor depending on its size and location. Liposarcoma most commonly arises in extremities or retroperitoneal spaces where fat contents are abundant but case reports of other atypical rare locations have also been reported such as: orbit, oral cavity, esophagus, small bowel mesentery, colon, sigmoid mesentery, pancreas, and scrotum [4]. Primary liposarcoma arising from the diaphragm is an extremely rare case and only 3 case reports have been described in the literature [4–6]. From our literature review, this is the first report to describe both CT and MRI findings of diaphragmatic liposarcoma invading gall bladder.

Case report

A 74-year-old male presented to our emergency department with sudden onset of right upper quadrant abdominal pain and fever. Physical examination showed positive Murphy's sign and lab results showed increased inflammatory markers and white blood cell counts. Acute cholecystitis was suspected initially and abdominal computed tomography (CT) was taken.

Abdominal CT showed a circumscribed lobulated heterogeneous mass (approximately 12 × 9 × 5.5 cm) that was penetrating right hemidiaphragm extending towards the liver through the right subphrenic space (Fig. 1A). Mass effect was exhibited pushing the liver posteriorly and the peritoneum anteriorly. Fluid collections around the mass were also observed. The mass was composed of multiple enhancing septas

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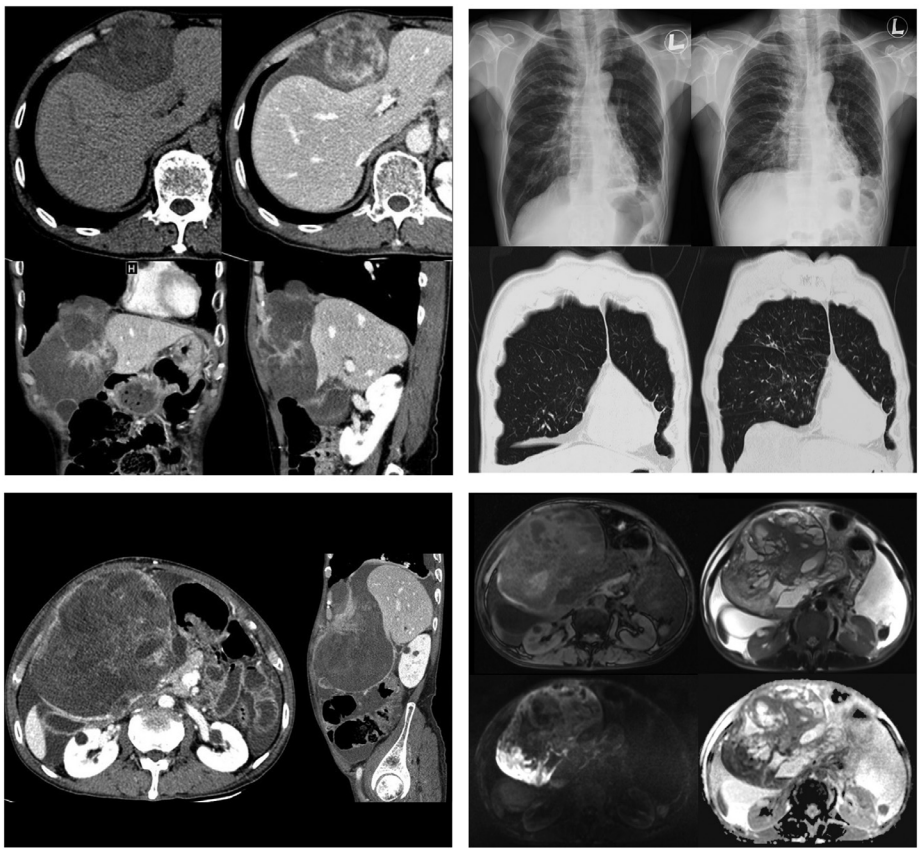

Fig. 1 – Dedifferentiated liposarcoma arising from diaphragm in a 74-year-old man. (A) (left upper) Axial CT of the abdomen and pelvis shows lobulated heterogeneous mass (approximately 12 x 9 x 5.5 cm) in right hemidiaphragm with some low density components and some muscle like density components. (right upper). Axial CT of the abdomen and pelvis with contrast shows enhancement of thick septations and nodular components. (left lower) Coronal and (right lower) Sagittal CT of the abdomen and pelvis with contrast shows displacement of liver, gall bladder, and peritoneum due to mass effects. (B) (left upper) Plain chest radiograph showing COPD lung with Lt lower lobe atelectasis filled with bronchiectasis (6 months ago) (right upper) Plain chest radiograph showing elevated Rt diaphragm (when patient presented for the first time) (left lower) Coronal CT of the chest (6months ago) (right lower) Coronal CT of the chest (when patient presented for the first time). (C) (left) Post 1 month follow up axial CT of the abdomen and pelvis with contrast shows interval increase in size of diaphragmatic mass with exacerbation of mass effects compressing gall bladder and inferior vena cava. (right) Post 1 month follow up sagittal CT of the abdomen and pelvis with contrast shows interval increase in size of diaphragmatic mass with exacerbation of mass effects compressing and displacing gall bladder. Gall bladder appears collapsed. (D) (left upper) Axial MRI T1 weighted image shows region of high T1 signal intensity in posterolateral region of the mass which may suggest hemorrhagic components (right upper) Axial MRI T2 weighted image shows multiple low T1 high T2 cyst like structures around peripheral region of the mass (left lower) Axial MRI diffusion restriction and (right lower) ADC image shows increased diffusion restriction in posterolateral region of the mass (E) Axial MRI enhanced image shows enhancement of septas and central soft tissue region. (F) Gross pathologic specimen shows 24 x 16 x 12 cm, lobulated mass composed of hemorrhagic necrosis with central grayish fibrous mass with multifocal cystic cavities. (G) Dedifferentiated liposarcoma of the diaphragm stained with hematoxylin and eosin (H&E) stain (left) Peripheral region shows well differentiated fatty cells with myxoid materials (original magnification x 200) (right) Central region showed atypical malignant cells (original magnification x 200). CT, computed tomography; MRI, magnetic resonance imaging; COPD, chronic obstructive pulmonary disease; ADC, Apparent diffusion coefficient.

with nonenhancing low density components (about 20 HU) and heterogeneously enhancing higher density components (about 40 HU). Retrospectively, raised right hemidiaphragm on the plain chest radiograph was also noted compared to 6 months ago (Fig. 1B). Ultrasound guided biopsy was suggested but the patient refused and was discharged.

The patient returned 1 month later with worsened abdominal pain and dyspnea. Another CT was taken and over 1 month the mass approximately doubled in size (approximately 23.6 × 17 × 13.3 cm) exhibiting greater mass effects compressing the adjacent gall bladder and the inferior vena cava in addition to the liver (Fig. 1C).

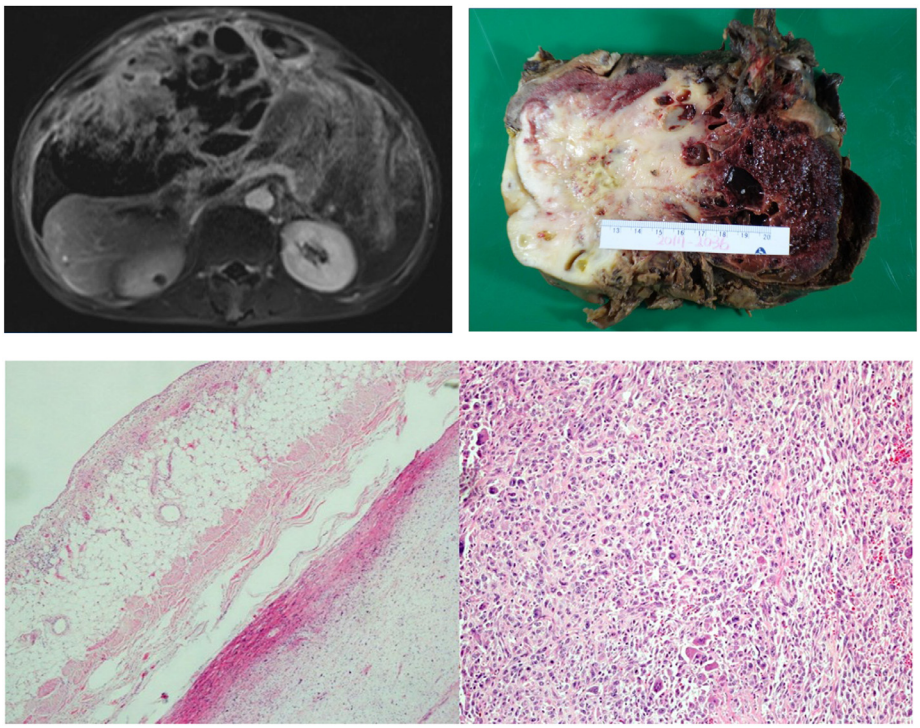

Fig. 1 – Continued

Magnetic resonance imaging (MRI) showed no evidence of fatty component in the mass. Low density region on the CT showed high T2 signal intensity suggesting cystic nature and higher density region on the CT showed isointense signal to the muscles suggesting soft tissue nature (Fig. 1D). Enhancement patterns were similar to the CT where the septa and suspected soft tissue regions were enhancing brightly (Fig. 1E). Hemorrhage or necrosis was also suspected in posterolateral region of the mass from its high T1 signal intensity (Fig. 1D). Diffusion restriction was also observed in the posterolateral soft tissue components (Fig. 1D).

Positron emission tomography scan showed no evidence of distant metastasis. Ultrasound guided biopsy was performed and the pathology initially reported giant cell tumor. Subsequently radical resection of the tumor and diaphragm was performed. According to the operation note, the mass was easily detached from the liver and the anterior peritoneal wall but adhesion to the gall bladder was noted and cholecystectomy was additionally performed.

On gross examination, the mass comprised of cystic cavities, hemorrhage, and necrotic components on peripheral region and more central solid soft tissue mass which all correlated well to our CT and MRI findings (Fig. 1F). Histopathologic evaluation showed central atypical malignancy cells surrounded by myxoid materials and benign appearing well differentiated liposarcoma like fatty cells (Fig. 1G). Gall bladder wall was also infiltrated with malignant cells. Dedifferentiated liposarcoma originating from the diaphragm invading gall bladder was the final diagnosis.

Discussion

All reported diaphragmatic liposarcoma cases describe similar clinical features to our case including abdominal mass, indigestion, dyspnea, epigastric, or chest pain [4–6] and perhaps can be mistaken for acute cholecystitis. Major dilemma when encountered with the diaphragmatic mass on imaging is whether it is truly a diaphragmatic mass rather than adjacent thoracic or abdominal origin especially when the mass is already large and extensive on first imaging [5,7]. In our case diagnosis was rather easy as it first presented as typical diaphragmatic hump which developed to rapidly growing mass over 1 month.

CT is helpful to evaluate mass and its surrounding anatomy such as penetration of diaphragm, sharp border to the liver, and adjacent structures which can suggest diaphragmatic origin [4–7]. Conventional chest radiography may show raised hemidiaphragm as of our case [4].

On CT and MRI, the appearance of diaphragmatic liposarcomas, as of the liposarcomas of other parts, varies depending on its pathologic subtypes [4–6,8]. MRI can provide more information about various different components inside the mass and can reflect histologic heterogeneity of liposarcoma and thus allows predicting pathologic subtypes. In this case, MRI suggested high T2 signal myxoid components, high T1 signal hemorrhagic component, and diffusion restriction of solid components which all correlated well with our postsurgical specimens. Only mismatching part was that the MRI showed no evidence of fat components in the mass, but histology

showed benign appearing fatty cells on the peripheral region of the mass. This may be due to minimal amount of fatty cells that failed to produce significant signal on MRI.

Dedifferentiated liposarcoma is known to be much more aggressive than well-differentiated type and can invade local adjacent structures prior to distant metastasis [2,9]. In this case, local invasion to the gall bladder by diaphragmatic liposarcoma was present. There was no evidence of liver or lung invasion despite abutment and extent of the mass which may suggest anatomic susceptibility of the gall bladder for invasion due to its lack of submucosal layer.

It is also interesting in our case that initially it was diagnosed as giant cell tumor on ultrasound guided needle biopsy. Dedifferentiated liposarcoma often contain primitive stem cells that may lead to misdiagnosis if only focal needle biopsy is taken. Since liposarcomas are composed of various different type of cells, diagnosis should be made on the basis of examining the whole mass after surgical excision or alternatively perform multiple biopsies when surgical resection is not possible [6].

Management of liposarcoma is primarily surgical resection [1,2]. Some may consider adjuvant or neoadjuvant chemotherapy for masses greater than 2 cm or in high-grade pathologic subtypes. Diaphragmatic liposarcoma growing towards intrathoracic direction seems to have poorer prognosis than the mass growing in inferior direction into intra-abdominal space. This is because mass can compress the heart and mediastinum causing cardiopulmonary complications and making surgical resection impossible [6].

In conclusion, CT and MRI play a vital role in diagnosis of liposarcoma providing information about its origin site, extent, invasion, metastasis and can even suggest pathologic subtypes prior to operation. Discriminating diaphragmatic tumor from other thoracic and abdominal neoplasm remains challenging but careful radiological examination can aid in determining true origin site. We herein describe CT and MRI findings of rare case of diaphragmatic liposarcoma. Despite its rarity, radiologists should consider possibility of liposarcoma as one of differential diagnosis when encountered with mass in the diaphragm that exhibits mass effect to adjacent organs.

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