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Diaphragmatic Hydatidosis: A Rare Location, A Case Report

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Abstract: **Case Report**

Hydatid disease, a parasitic infection caused by "Echinococcus granulosus", most commonly affects the liver and lungs. Primary involvement of the diaphragm is an exceedingly rare presentation, accounting for approximately 1% of cases, and poses a significant diagnostic challenge as it can mimic cysts originating from the hepatic dome or the lung base. This article synthesizes the crucial role of medical imaging in the diagnosis, characterization, and management of diaphragmatic hydatidosis. While chest radiography and ultrasound (US) may provide initial clues, cross-sectional imaging with computed tomography (CT) and magnetic resonance imaging (MRI) is paramount for a definitive diagnosis and pre-operative planning. We detail the specific imaging findings across these modalities, including the Gharbi classification on US and signs of complications on CT. Emphasis is placed on the key radiological signs for precise topographic localization, such as the "claw sign," the visualization of a thinned and stretched diaphragmatic muscle around the lesion, and the superior soft-tissue delineation on T2-weighted MRI sequences. A thorough understanding of these imaging features is essential for radiologists to accurately diagnose this rare entity, particularly in endemic regions. We report the case of a 67-year-old incarcerated patient admitted with chronic dyspnea of recent worsening, associated with chest pain, right hypochondrial heaviness, and distension, An initial ultrasound suggested a hydatid cyst of the hepatic dome. Computed tomography revealed a large inter-hepato-diaphragmatic hydatid cyst, very likely of diaphragmatic origin. The diagnosis was subsequently confirmed at surgery.

Keywords: Hydatid Disease, Cystic Echinococcosis, Diaphragm, Diaphragmatic Hydatid Cyst, Computed Tomography (CT), Magnetic Resonance Imaging (MRI), Claw Sign.

INTRODUCTION

Hydatid disease, or cystic echinococcosis, is a zoonotic parasitic disease caused by the ingestion of eggs from the tapeworm "echinococcus granulosus" [1-3]. Transmission to humans occurs mainly through the consumption of contaminated water or food, or by direct contact with infected definitive hosts such as dogs [1]. Endemic in livestock-raising regions and areas with low socioeconomic status, particularly the mediterranean basin, the disease most commonly affects the liver (60-80%) and lungs (10-30%) [1, 2].

Case Report

A 67-year-old incarcerated patient admitted in emergency department with chronic dyspnea of recent worsening, associated with chest pain, right hypochondrial heaviness, and distension. An initial ultrasound suggested a hydatid cyst of the hepatic dome. We performed a thoracoabdominal CT scan, which demonstrated a large inter-hepato-diaphragmatic cystic lesion measuring **16 × 17 × 22 cm** *(transverse × anteroposterior × craniocaudal)*, with a regular wall and lobulated contours, containing focal

calcifications as well as multiple septations and endocystic daughter vesicles. On axial images, there was probable extension with an exocystic vesicle into the pleural space and the right cardiophrenic angle. On sagittal and coronal reformations, the beak sign was negative with both the liver and the lung. The mass displaced the liver inferiorly and the lung superiorly, with adjacent parenchymal atelectasis. It appeared inseparable from the diaphragmatic band on sagittal images, with a claw sign visible on some slices. A loculated ipsilateral pleural effusion was also noted. Some cystic lesions were also identified in the middle and lower mediastinum, compatible with associated mediastinal involvement **(Figure 1)**. No other visceral lesions were detected, particularly in the liver, lungs, or kidneys. Subsequently, thoracic ultrasound was performed and confirmed pleural involvement, showing hydatid vesicles within the pleural space. It also supported the absence of hepatic origin and did not reveal any hepatic parenchymal hydatid lesion **(Figure 2)**. The patient underwent surgery, which confirmed the diaphragmatic location of the cyst. Complete cysts excision was performed, followed by right diaphragmatic repair.

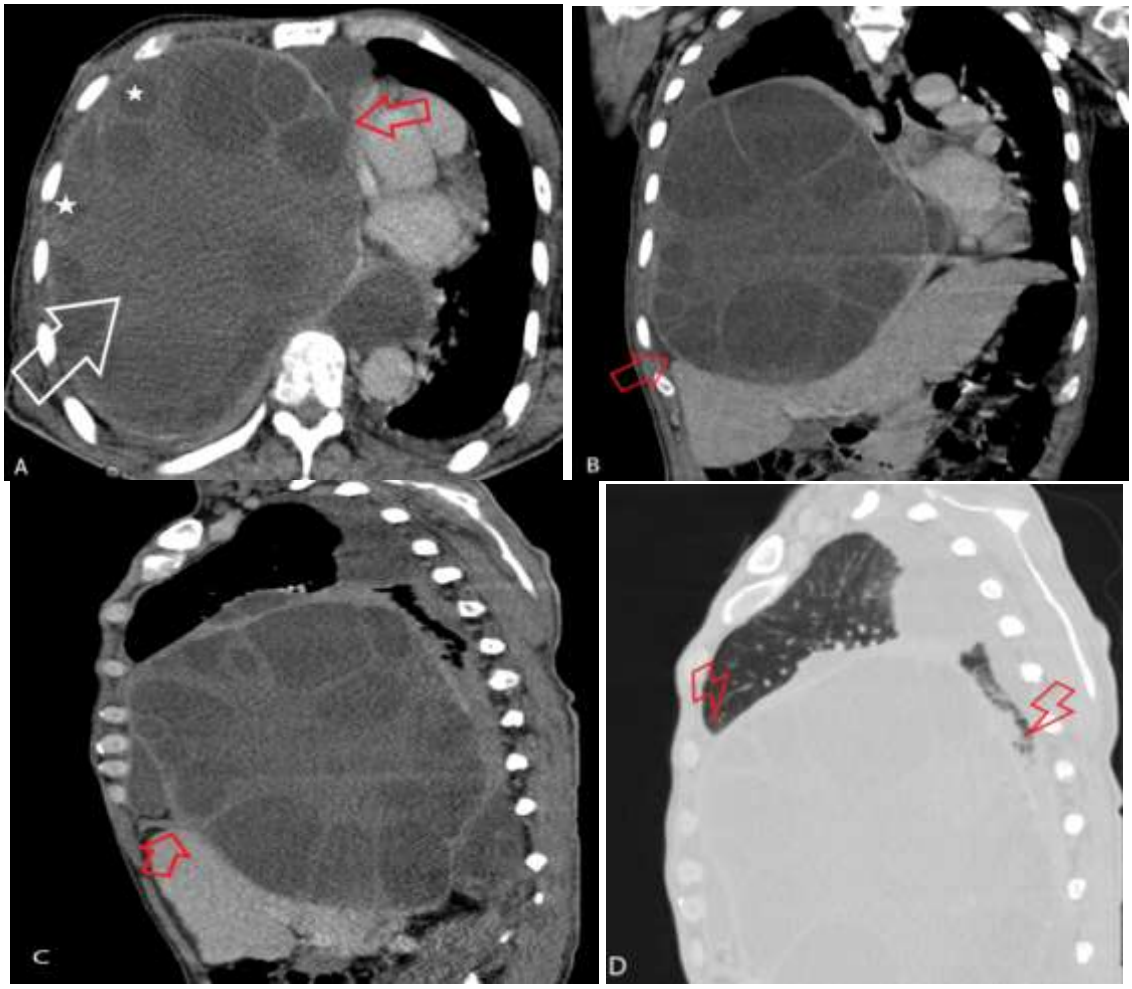

Figure 1: Contrast-enhanced thoracoabdominal CT; A: Axial image showing a cystic mass adjacent to the right hepatic dome (white arrow), containing multiple endogenous daughter vesicles/cysts (stars), with probable communication with an exocystic vesicle in the right anterior cardiophrenic space (red arrow); **B:** Coronal image showing a negative beak sign with the liver; the cyst displaces the liver inferiorly; **C:** Sagittal image showing the mass inseparable from the diaphragm (red arrow); **D:** The mass displaces the lung superiorly, with adjacent parenchymal atelectasis and a negative beak sign (red arrows)

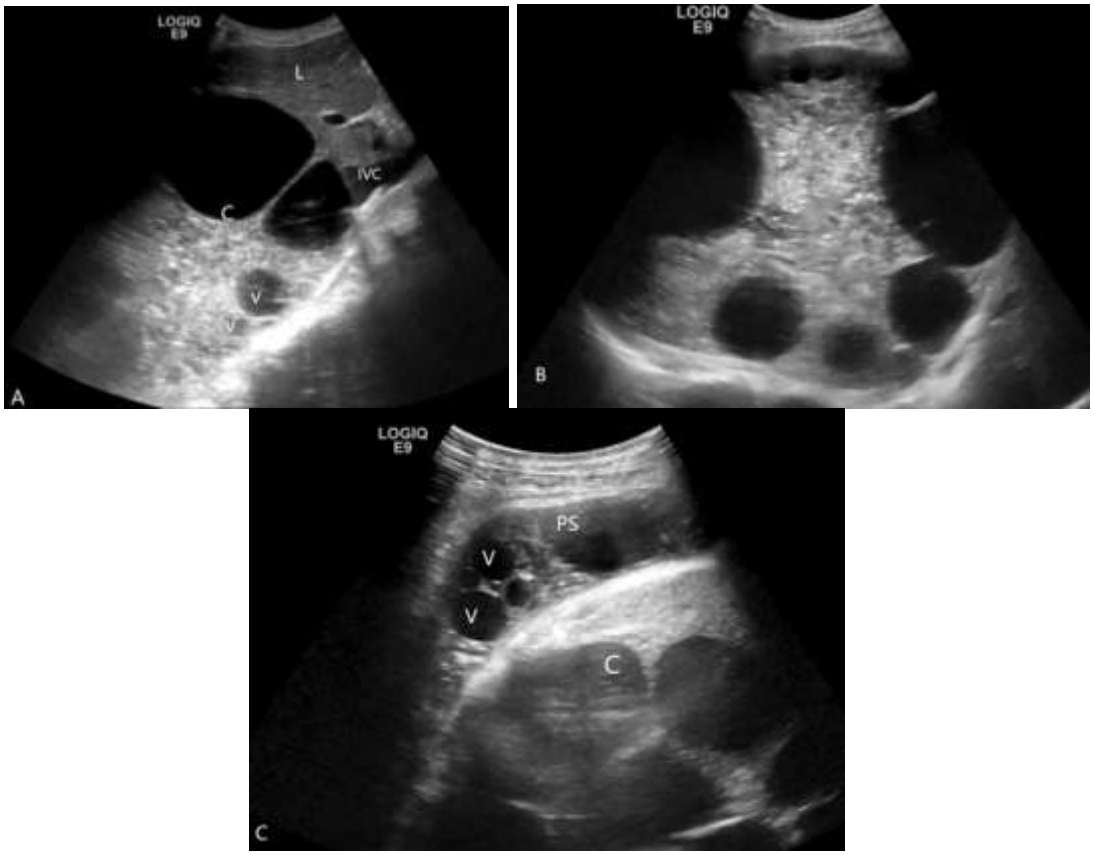

Figure 2: Ultrasonographic examination; A: Longitudinal image showing the hydatid cyst (C) displacing the liver inferiorly (L), with a negative beak sign. The cyst contains numerous endogenous daughter vesicles (V) and shows intimate contact with the inferior vena cava (IVC); **B:** Ultrasonographic appearance of the mass, showing a fairly well-defined cystic lesion with heterogeneous echogenic content, containing numerous septations and endocystic daughter vesicles; **C:** Pleural involvement confirmed by ultrasonography, with multiple hydatid vesicles (V) within the pleural space (PS); the adjacent hydatid cyst is also seen (C)

DISCUSSION

Diaphragmatic involvement is an extremely rare manifestation of this disease, accounting for only about 1% of cases [1]. The pathophysiology of this location is not fully understood, but theories suggest dissemination via lymphatic or hematogenous routes [3]. The clinical symptomatology is not specific; symptoms related to the compression of surrounding organs may be observed, such as coughing, chest pain, or dyspnea [1, 3]. The diagnosis of primary diaphragmatic hydatid disease is challenging, as cysts are often misdiagnosed as originating from adjacent structures on imaging, and their precise location is usually only confirmed during surgery [4]. Medical imaging is the cornerstone of diagnosis. Chest radiography, ultrasound (us), computed tomography (CT), and magnetic resonance imaging (MRI) are important complementary tools for diagnosis [5]. CT and MRI are often used to clarify the affected sites and facilitate surgical evaluation [2]. A chest x-ray is often the first-line examination. It can reveal indirect signs such as an elevation of the diaphragmatic dome or a well-defined, rounded opacity at the thoracic base [1]. Sometimes, a calcified sub-diaphragmatic liver cyst may be discovered incidentally on a chest film [5]. Ultrasound is a non-invasive and accessible modality that is highly effective for characterizing cystic lesions. It can clearly demonstrate the presence of floating membranes, daughter vesicles, and hydatid sand [6]. The Gharbi classification, although initially developed for liver cysts, is widely used to describe the different evolutionary stages of the cyst [7]:

Type I: a pure, anechoic, unilocular cyst with a thin wall. Type II: a cyst with a detached membrane, giving the appearance of a "floating membrane" or "water-lily sign" [6]. Type III: a multilocular cyst containing daughter vesicles, creating a "honeycomb" appearance [2]. Type IV: a heterogeneous lesion with a pseudo-tumoral aspect. Type V: a calcified cyst, often considered inactive [8]. Computed tomography (CT) plays a crucial role in diagnosis, surgical planning, and the evaluation of potential complications [9]. It offers excellent spatial resolution and allows for a detailed analysis of the cyst's morphology and its relationship with adjacent structures. Uncomplicated cysts appear as well-circumscribed lesions with a smooth wall of variable thickness and fluid density (low attenuation) [2, 9]. Wall calcifications, although rare, are a specific sign when present [2]. Magnetic resonance imaging (MRI) offers superior tissue contrast, making it an excellent modality for exploring soft tissues and the complex relationships of the cyst with neighboring organs [2]. The main diagnostic challenge of diaphragmatic hydatidosis is to confirm its strictly intramuscular location, as cysts in the hepatic dome or lung base can mimic this involvement [11]. Cross-sectional imaging, particularly CT and MRI, is fundamental for this precise topographic diagnosis [11, 12]. Several radiological signs are sought to confirm the diaphragmatic origin of the cyst: The claw sign: this is one of the most important signs. It is visible when the diaphragmatic band appears to wrap around the cystic lesion, forming a "claw" that encases it, which indicates that the lesion arises from the diaphragm itself [11]. Visualization of the thinned and displaced diaphragm: a diaphragmatic hydatid cyst will develop within the muscle fibers, stretching and thinning them. Imaging can then show the cyst embedded in the diaphragm, with a thin layer of muscle tissue visible at its periphery, clearly separating it from the liver and lung. Another sign is absence of a cleavage plane with the diaphragm: unlike a liver or lung cyst that displaces the diaphragm, a diaphragmatic cyst is intimately attached to the muscle, resulting in the absence of a fatty cleavage plane or a clear interface between the cyst and the diaphragm. MRI is particularly useful due to its excellent soft-tissue contrast [13]. On T2-weighted images, the cyst appears with high signal intensity while the diaphragmatic muscle remains with low signal intensity, allowing for clear visualization of the thinned muscle around the cyst. The presence of a peripheral low-signal-intensity T2 rim, corresponding to the fibrous pericyst, is another characteristic feature of hydatid cysts [14, 15]. For differential diagnosis, due to its sometimes atypical presentation, diaphragmatic hydatidosis can be confused with other pathologies. The differential diagnosis mainly includes [10]: benign or malignant tumors (primary diaphragmatic tumors, metastases, cavitary lung cancers), congenital cystic lesions: (bronchogenic cysts, cystic adenomatoid malformations), infectious pathologies (diaphragmatic or subphrenic abscess). In endemic areas, any cystic lesion of the diaphragm should raise the suspicion of hydatid disease [2, 10]. Although hydatid disease is a benign condition, serious complications can occur, leading to high morbidity and even death [2]. CT plays an important diagnostic role in the evaluation of complicated cases [9]. Rupture is a major complication, which can lead to parasitic dissemination and a risk of anaphylactic shock [2]. CT can reveal several signs of rupture [2, 9]: air crescent sign: the presence of air between the pericyst and the endocyst, dry cyst sign: a cyst completely emptied of its contents. Also, a perforated hydatid cyst can become infected and form an abscess, presenting a radiological pattern common to other infectious lung lesions [10].

CONCLUSION

Diaphragmatic hydatidosis is a rare entity whose diagnosis relies on a combination of clinical, biological, and especially radiological findings. Careful analysis on cross-sectional imaging is crucial to confirm the intra-diaphragmatic location. Knowledge of the typical signs and aspects of complications is essential for any radiologist,

particularly in endemic areas, to ensure prompt and appropriate management of this pathology.

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