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Case Report

The "serpent sign"—A classical sign in a nonclassical location: A case report of breast hydatid cyst ☆

Eman Abu-Mandeel, MD[a], *Safa' Al Amleh, MD*[b], *Muhannad M. Mahmoud*[c,*], *Omar Azizieh*[c]

[a] *Radiology Department, Ministry of Health, Amman, Jordan*
[b] *Pathology Department, Ministry of Health, Amman, Jordan*
[c] *The University of Jordan, Faculty of Medicine, Rafee Othman St, Amman, Jordan*

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ABSTRACT

Hydatid cyst is a rare breast pathology; however, it should be included in the differential diagnosis of any breast mass. The history and imaging findings can sometimes raise suspicion of the hydatid nature of the mass. The presence of certain imaging signs specific to hydatid cysts can further indicate this diagnosis. Nevertheless, the definitive diagnosis remains histological, and the treatment is principally surgical. Herein, we present a case of a 38-year-old female who was found to have the "serpent sign" on ultrasonography, which is pathognomonic for ruptured hydatid cysts, as part of the imaging workup for a left breast lump. This case report underscores the importance of characteristic imaging features such as the "serpent sign" in the diagnosis of hydatid disease, especially in unusual sites like the breast where the index of suspicion is particularly low. Recognition of such signs elicits special pre- and intraoperative precautions to minimize complications like local dissemination and anaphylactic shock.

Introduction

Hydatid disease is a parasitic infection caused by the larval stage of the Echinococcus cestodes [1]. Dogs typically serve as definitive hosts, while ruminants like sheep are the intermediate hosts for the infective larval stage [2]. This rare zoonosis occurs mostly in endemic areas namely, the Mediterranean regions, Central Asia, China, North and East Africa, Australia, and South America [1]. The most common form found in hu-

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* Corresponding author.
E-mail address: mohannad.mah00@gmail.com (M.M. Mahmoud).

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mans is cystic hydatid disease, which is caused by Echinococcus granulosus [3]. Humans, as accidental hosts, get infected following the ingestion of echinococcal larvae. Subsequently, the larvae penetrate the intestinal lining to reach the liver via the portal circulation. The liver serves as the first filter and is thus the most commonly involved organ (70%). The larvae may also percolate through and seed the lungs, the second most common site (20%). The remainder of cases involves other organs, such as the kidney, spleen, gallbladder, pancreas, thyroid gland, brain, and spinal cord [4,5].

Breast hydatid cyst is an extremely rare occurrence even in endemic regions, arising in less than 3 in 1000 cases [5]. Due to their similarity to other types of breast tumors, clinicians should include hydatid cysts among the differential diagnosis of any breast lump. The diagnosis of breast hydatid cysts may be obscured by a conspiracy of 2 factors: an exceedingly low index of suspicion premised on disease rarity and the resemblance to other more common diagnoses. However, highly characteristic signs may occasionally be noted on imaging. One example would be the "serpent sign," which is pathognomonic for ruptured hydatid cysts. This sign depicts detached laminated membranes following an internal (contained) cyst rupture, which appear radiologically as curvilinear structures floating within the cyst, giving the appearance of a serpent [6]. To the best of our knowledge, this is the first case report of the "serpent sign" in a breast hydatid cyst.

Case presentation

A 38-year-old previously healthy female patient presented to the clinic complaining of a palpable left breast mass gradually enlarging in size for 7 months. On examination, the mass was non-tender, firm, and mobile in the upper outer quadrant of the left breast. There were no skin abnormalities or palpable ipsilateral axillary lymph nodes. The routine laboratory tests came back normal. Mammography with craniocaudal (CC), mediolateral oblique (MLO) and spot compression views showed an oval circumscribed isodense mass at the upper outer quadrant of the left breast measuring 3.5 × 2.5 × 3 cm (BI-RADS 4a) (Fig. 1). On ultrasound, an oval circumscribed complex solid and cystic heterogeneous mass at 2 o'clock of the left breast with floating curvilinear structures representing the "serpent sign" (detached inner membranes) was seen which is indicative of a hydatid cyst (Fig. 2A). Complementary liver ultrasound was done and revealed multiple liver cysts (Fig. 2B). Echinococcal serology was not done. The patient was referred to the breast surgery department for surgical excision of the mass as a hydatid cyst was highly suspected. Computerized tomography (CT) scan of the chest and abdomen was ordered. It showed multiple non-enhancing liver lesions with fluid density, and both lungs were free (Fig. 3). Excision of the mass was done, and the histopathology examination of the excised mass showed cyst space lined by an acellular laminated membrane (Fig. 4). The diagnosis of a hydatid cyst was confirmed. Consequently, the patient was started on albendazole and had an uneventful recovery on follow-up. In addition, she was recommended to adhere to the breast cancer screening program.

Discussion

Breast hydatid cyst can develop in isolation or more commonly, secondary to diffuse hydatid disease [7]. In the latter scenario, the breast hydatid cyst may, as in our case, be the first sign of the underlying hydatidosis. When this occurs, patients initially present with painless breast lumps that steadily grow in size without involving any local lymph nodes [8]. Although ages from 20 to 74 have been documented, 30-50 years old are the most common ages of presentation [9].

During the active stage, hydatid cysts grossly appear as 3 distinct layers: The outermost layer (pericyst) is the host's fibrous reaction that encases the parasite. The middle layer, the laminated layer (ectocyst), is the most distinctive because of its white color and relatively uniform thickness. This layer is, in turn, produced by the innermost layer, the germinal layer (endocyst) which can be demonstrated in viable cysts as a thin, translucent membrane [10]. When the cyst's internal pressure rises, it may burst and the ectocyst and pericyst may separate. The collapsed laminated membranes of the ectocyst floating within an intact pericyst will appear as discrete, curvilinear structures on ultrasound, CT, or magnetic resonance imaging and resemble a snake or serpent – the "serpent sign" [6]. This sign is exclusively seen in internally ruptured hydatid cysts. Therefore, when it was revealed on ultrasonography in our case, it prompted us to examine the rest of the body for further hydatid cysts, disclosing multiple hepatic hydatid cysts. In addition, the high degree of suspicion of the cyst's hydatid nature may have helped to avert the risk of intraoperative cyst rupture and its devastating consequences. Nevertheless, physicians are rarely fortunate enough to obtain such specific radiological signs since breast hydatid cysts often present with vague and inconclusive imaging findings leading to either a controversial fine-needle aspiration or a missed diagnosis [4,5,9,11]. Aside from the 2 most common sites for hydatid cysts, the liver and the lungs, the "serpent sign" was elicited in other atypical sites including iliopsoas muscle, skull base, pleural space, and the frontal lobe of the brain [12–17].

Ultrasonography is considered the cornerstone imaging modality to evaluate and classify hydatid cysts [18]. Ultrasonography findings fall on a spectrum and vary based on the stage of growth. However, according to Gharbi et al., hydatid cysts can be sonographically classified into 5 distinct types: Type I (unilocular, anechoic lesion), Type II (cyst with detached laminated membranes), Type III (multiseptated cyst with daughter cysts), Type IV (cyst with heterogeneous content), and Type V (solid cyst with wall calcification) [19]. Based on the World Health Organization Informal Working Group on Echinococcosis (WHO-IWGE) cyst classification, the presence of the "serpent sign" qualifies the hydatid cyst in our case to be classified as "transitional stage infection" between active and inactive disease [17].

In addition to the "serpent sign," various ultrasound signs characteristic of hydatid cyst have been evoked in the lit-

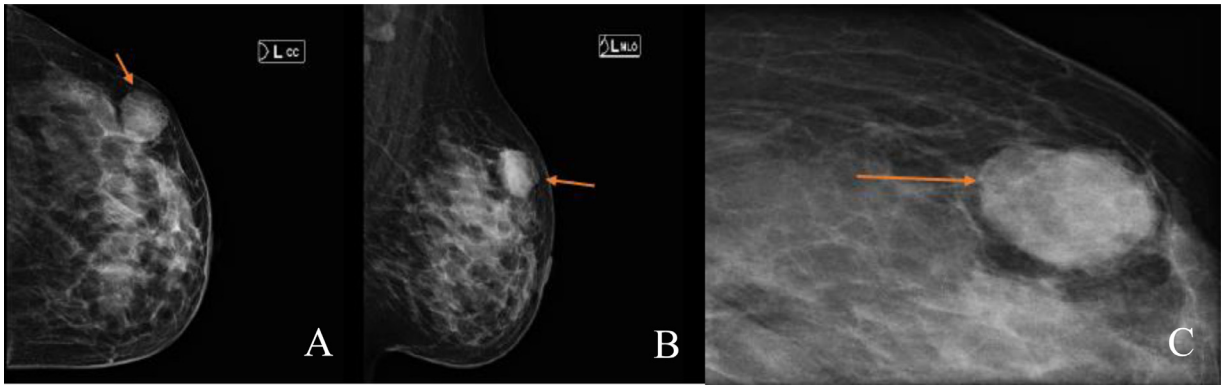

Fig. 1 – Mammography of the left breast. Craniocaudal (A), mediolateral oblique (B), and spot compression (C) views demonstrate an oval circumscribed isodense mass at the upper outer quadrant of the left breast (arrows), measuring 3.5 x 2.5 x 3 cm. There is no evidence of suspicious-looking calcifications or masses in both breasts. (BI-RADS 4a.)

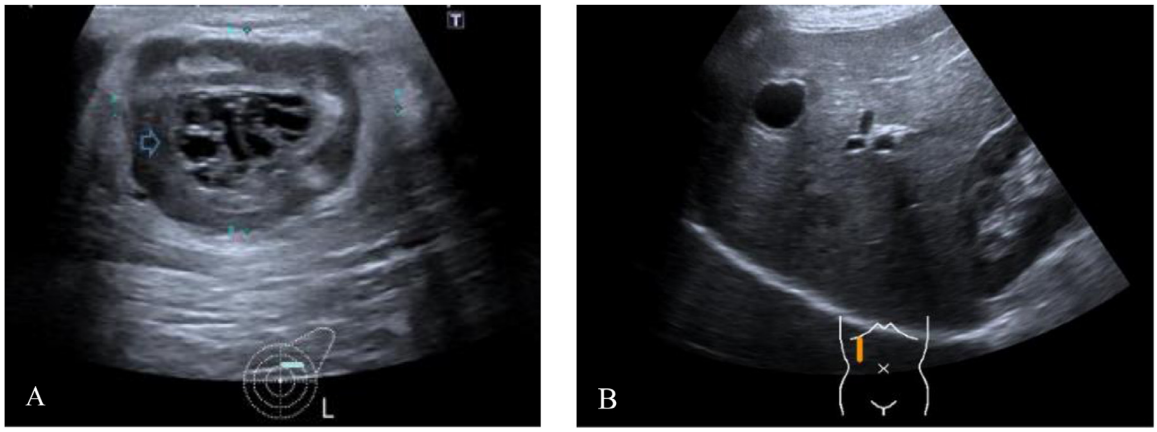

Fig. 2 – (A) Left breast ultrasound. There is an oval circumscribed complex solid and cystic heterogeneous mass at 2 o'clock of the left breast, with posterior acoustic enhancement. The mass contains floating curvilinear structures (arrowheads) representing the "serpent sign" (detached laminated membranes), suggestive of hydatid cyst. (B) Complementary liver ultrasound. Multiple, variably sized, anechoic lesions that lack soft tissue components, suggestive of cystic nature, are scattered in the liver.

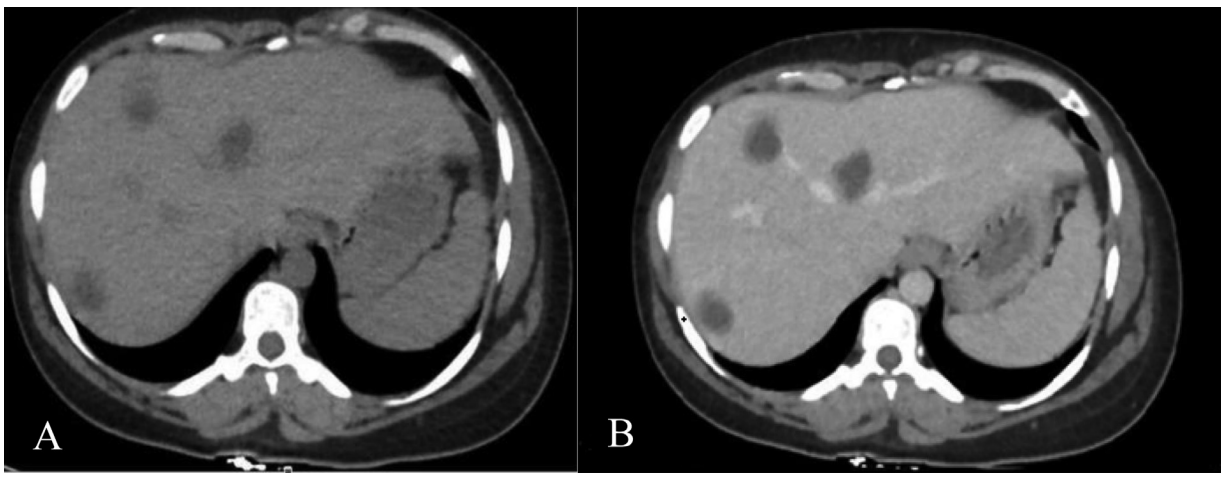

Fig. 3 – Axial CT scan of the abdomen. Pre-IV contrast (A) and post-IV contrast (B) images show multiple nonenhancing liver cysts.

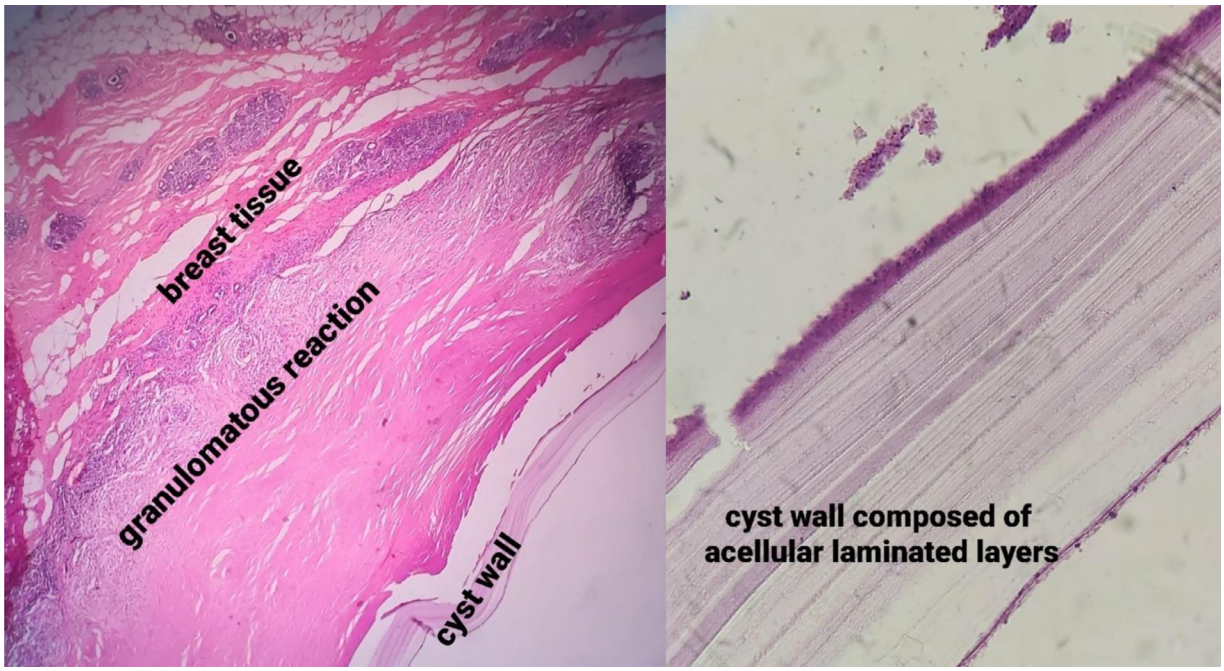

Fig. 4 – Cyst wall composed of acellular laminated layers and fibrous tissue, surrounded by a dense granulomatous reaction within the breast tissue.

erature, including a double-layered wall appearance of the cyst, the "double line" sign [20]. The movement of hydatid sand within the cyst produces a second sign, the "snowstorm sign [21]. Another strongly indicative sign of hydatid cysts is the "congealed water lily sign," which is characterized by an immobile germinal membrane and enhanced echogenicity of cystic fluid as the typically watery fluid becomes viscid [22].

Imaging modalities other than ultrasonography can also be used in the evaluation of breast masses, including mammography, CT, and magnetic resonance imaging. Hydatid cyst findings are nonspecific on mammography, which typically shows, as in our case, well-circumscribed, homogenous, isodense lesions. On the basis of such features, the differential diagnosis would include cyst, fibroadenoma, phylloides tumor, and, rarely, well-circumscribed carcinoma [23].

Conclusion

Owing to its rarity, hydatid disease is easily overlooked in the differential diagnosis of a breast mass, even in endemic areas. The appearance of particular hydatid cyst imaging indicators, such as the "serpent sign," can alert clinicians to the possibility of this unusual diagnosis, precluding risky operations such as fine-needle aspiration and necessitating special precautions during surgery.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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