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Review Article

Hydatid Disease: A Pictorial Review of Uncommon Locations

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Abstract

Hydatid disease is a zoonotic infection caused primarily by the tapeworm parasite, *Echinococcus granulosus*. It is considered an endemic disease in the Mediterranean region. In about 90% of cases, hydatid cysts are found in the liver and lungs; however, any other organ in the body may be affected, particularly in endemic areas. When encountering cystic lesions in these areas, the physician should always keep hydatid disease as a possible diagnosis in mind. To avoid life-threatening conditions such as anaphylactic shock or pressure effect on vital organs, timely diagnosis, and proper management are critical. When a rare site is involved, hydatid disease should be diagnosed using a combination of serologic assays and imaging modalities such as ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI). These imaging modalities can also be used to determine the extent of the disease and assess possible complications. Here, we present a pictorial review of typical imaging manifestations of hydatid cysts in unusual sites. Being aware of these imaging features will assist physicians in making an accurate, timely diagnosis and subsequently, providing optimal management.

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Keywords ● *Echinococcus granulosus* ● Diagnostic imaging ● Magnetic resonance imaging ● Ultrasonography

What's Known

- Hydatid disease is a parasitic zoonotic disease, which mainly involves the liver and lungs.
- Hepatic and pulmonary hydatid cysts are easily detected on ultrasonography by typical imaging manifestations.

What's New

- In endemic areas, hydatid cysts rarely involve other organs of the body.
- Hydatid cysts found in rare sites can be misdiagnosed, leading to inappropriate treatment and life-threatening complications.
- In this paper, we describe the imaging manifestations of hydatid cysts in rare sites based on our own original cases.

Introduction

Hydatid disease is a zoonotic infection caused primarily by the tapeworm parasite *Echinococcus granulosus* larvae.[1] Cystic echinococcosis in human and animal hosts is most prevalent in temperate climate countries, including several parts of the Mediterranean region, southern and central parts of Russia, Central Asia, and China. Ingestion of food tainted with *Echinococcus* eggs or direct contact with a definitive host, usually a domestic dog, can cause humans to become incidental intermediate hosts. After migrating through the portal venous or lymphatic system, the majority of the larvae or oncospheres, released by ova in the small intestine, become trapped in the hepatic sinusoids. Since a lesser proportion of the larvae pass through the liver parenchyma and reach the lungs and systemic circulation, the liver and lungs are the most frequently involved sites.[2, 3]

Hydatid disease is highly prevalent in Iran, one of the most important endemic areas in the Middle East, and is thus considered a serious public health issue.[4] As expected, the majority of cystic hydatidosis cases reported from Iran are of the liver and lungs. However, a number of less common presentations,

including those involving the heart, orbit, brain, muscle, salivary glands, urinary tract, and pancreas have also been documented, many of which have nonspecific symptoms.[5] Familiarity with such rare hydatid disease presentations is essential, particularly in endemic regions where hydatid disease should be considered in the differential diagnosis of cystic lesions affecting any organ of the body. Since the majority of cases are asymptomatic for several years, hydatid cysts may be detected incidentally during radiologic workup.[1, 2] In some cases, distinctive imaging characteristics contribute to making a certain preoperative diagnosis, which is mandatory for optimal treatment planning. In this pictorial review, we present an overview of typical imaging manifestations of hydatid cysts in unusual locations.

Cardiac Hydatid Cyst

Cardiac hydatid cysts are extremely rare, accounting for only 0.02-2% of all reported cases of hydatidosis. Either through the pulmonary veins, lymphatic vessels, or coronary arteries, the larvae enter the myocardium.[6] Although the left ventricle is the most frequently affected site, the interventricular septum, right ventricle, and less frequently the pericardium may also be affected by the cardiac involvement. This is likely due to its greater myocardial mass and abundant blood supply.[7, 8] The signs and symptoms of cardiac hydatid cysts vary greatly depending on their location, size, and compression effects on adjacent structures. Cardiac hydatidosis might be asymptomatic and only be detected incidentally, or it might cause precordial discomfort (often vague), palpitation, dyspnea, and arrhythmia.[9] It may also cause severe life-threatening complications such as cyst rupture, which can result in acute pericarditis or cardiac tamponade, anaphylactic shock, pulmonary embolism, acute pulmonary hypertension, and systemic embolization. Since right-sided ventricular hydatid cysts are more likely to be intracavitary and subendocardial, these cysts rupture more frequently and are consequently more fatal.[10]

Transthoracic echocardiography is quite sensitive in detecting cardiac cysts.[6, 11] However, CT and magnetic resonance imaging (MRI) can provide additional information regarding the distinguishing features of hydatid cysts as well as the extent of hydatid disease (figure 1).

Congenital cysts, aneurysms, and solid tumors (such as fibromas and myxomas) are among the differential diagnoses for cardiac hydatidosis. While MRI clearly demonstrates the exact location of the cyst and its relationship with adjacent structures, CT is the greatest tool for displaying the calcification of the cyst wall, a rather specific indication. On T1-weighted and T2-weighted MR images, a hydatid cyst is typically observed as a hypointense and hyperintense lesion, respectively. On T2-weighted images, the pericyst also appears as a low-signal intensity rim. Other specific indications that may help in the diagnosis of cardiac hydatidosis include the existence of daughter cysts and membrane detachment.[9, 10, 12]

Even in asymptomatic patients, surgical excision under cardiopulmonary bypass is still required for cardiac hydatidosis therapy.[11] This surgical procedure, followed by supplemental pharmacological treatment with albendazole, has yielded high success rates. Medical therapy should be utilized as a last resort, only when

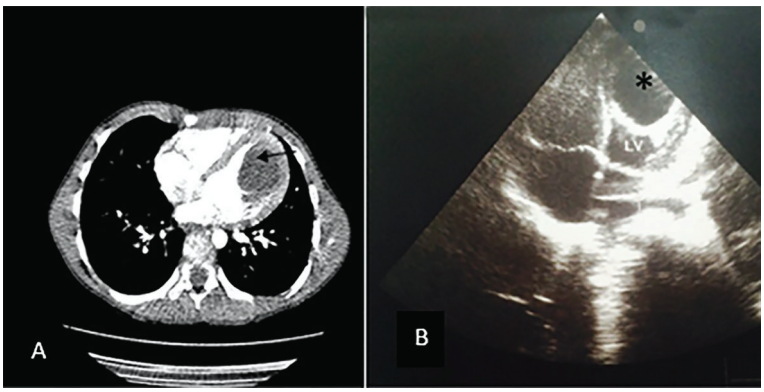

Figure 1: This figure shows cardiac hydatid disease in a seven-year-old boy with abdominal pain. Abdominal ultrasonography revealed several cysts with internal membranes in the liver resembling hydatid cysts. Similar cysts were also detected in the heart from the subxiphoid view. A. Axial contrast-enhanced CT scan of the chest shows a well-defined unilocular intraventricular cyst located at the left ventricular apex (arrow) with no involvement of any other organs including the lungs. The cyst has a low density (10 HU), which is compatible with a simple cyst. B. Echocardiography reveals a well-defined anechoic cyst (asterisk) with posterior acoustic enhancement within the left ventricular apex in the four-chamber view. During hospitalization, the patient experienced dyspnea and chest pain, and finally underwent open thoracic surgery for cardiac hydatid cyst excision, but unfortunately expired due to cyst rupture and anaphylactic shock. This figure was obtained from a patient admitted to the authors' institution and was not previously reported elsewhere.

surgery is contraindicated or not practicable. Although protoscolicidal agents such as iodine, hypertonic saline, methylene blue, or ethanol can be used to sterilize hydatid cysts prior to enucleation and pericardial cavity irrigation, none of these agents are completely safe and effective for intraoperative usage.[6, 13, 14]

Sacral Hydatid Cyst

The spine, pelvis, femur, tibia, humerus, skull, and ribs are the most commonly involved sites in hydatid disease, with a frequency of 0.5-4%.[2] There are few reports of hydatid disease with sacral involvement in the literature, most of which mentioned low back pain as the initial presenting symptom.[15]

The imaging features of musculoskeletal hydatid disease can occasionally be nonspecific and mimic those of other neoplastic or non-neoplastic pathologies, such as tuberculous spondylitis, chronic osteomyelitis, neurofibroma, metastases, giant cell tumor, meningocele, bone cyst, and also developmental cysts. Consequently, radiologists may face a diagnostic dilemma when dealing with hydatid disease of the bone.[12, 15] A number of imaging features, however, were attributed to spinal hydatid disease. These include paraspinal extension, vertebral body involvement with disk space-sparing, and absence of osteoporosis and sclerosis in the involved bone. In addition, intraosseous hydatid cysts lack the distinctive calcification that may be seen in extraosseous lesions (figure 2A). Since there are no pericysts in hydatid disease with musculoskeletal involvement, the lesion can proliferate aggressively in an irregular branching pattern. With the gradual growth of the cystic lesion over time, the cortex of the bone will be destroyed and extend into adjacent soft tissues (figure 2A). The CT scan is the preferred method for the delineation of bone destruction caused by hydatid disease (figure 2B). MRI, on the other hand, is superior in the evaluation of the extraosseous spread of the cyst to surrounding soft tissues, particularly the spinal canal (figure 3). Thus, in endemic zones, hydatid disease should be considered in the differential diagnosis of osteolytic bone lesions.[2]

In patients with sacral hydatidosis, medical treatment can reduce the size of hydatid cysts; however, the surgical excision of the cyst followed by anthelmintic therapy is the most effective treatment. After removal, protoscolicidal agents such as hypertonic saline can be administered for irrigation. Notably, formalin should be avoided due to the possibility of accidental intradural spilling and subsequent serious complications that could result. Despite treatment, disease recurrence is high in these patients, and thus, a long-term follow-up is necessary.[16]

Orbital Hydatid Cyst

Orbital involvement is a rare presentation of *Echinococcus granulosus* infection and accounts for less than 1% of all cystic involvements. It is mostly found in the pediatric and young adults population.[17] Unilateral and painless proptosis is the most common clinical sign in patients with orbital hydatid cysts, followed by orbital pain, chemosis, and orbital cellulitis.[18] Delayed diagnosis and treatment lead to prolonged optic nerve compression and atrophy, subsequently resulting in vision loss. The cysts are more prevalent in the retrobulbar region of the orbit and can be found within or outside the muscle cone.[17, 19] Despite being rare, a probable diagnosis of orbital hydatid cyst should always be kept in mind when encountering a patient with unilateral proptosis and orbital cystic lesion in areas endemic for echinococcosis.

Since the cysts are confined to the orbital space and there is minimal exposure of parasitic proteins to the immune system, serological tests in patients with orbital hydatidosis are

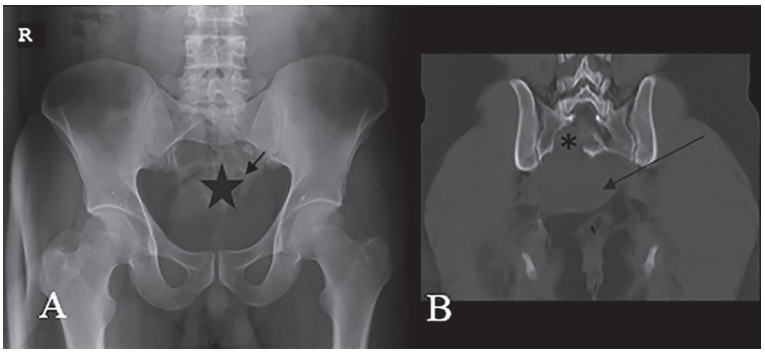

Figure 2: This figure shows sacral hydatid disease in a middle-aged man with chronic vague pelvic pain. A. Anteroposterior (AP) pelvic radiograph shows a massive destructive bone lesion in the lower segments of the sacrum without dystrophic calcification (asterisk) with faintly visible adjacent soft tissue mass (arrow). B. Coronal non-contrast enhanced pelvic CT shows a soft tissue mass with adjacent bone destruction which extends through the right sacral foramen into the spinal canal (asterisk). This figure was obtained from a patient who was admitted to the authors' institution and was not previously published.

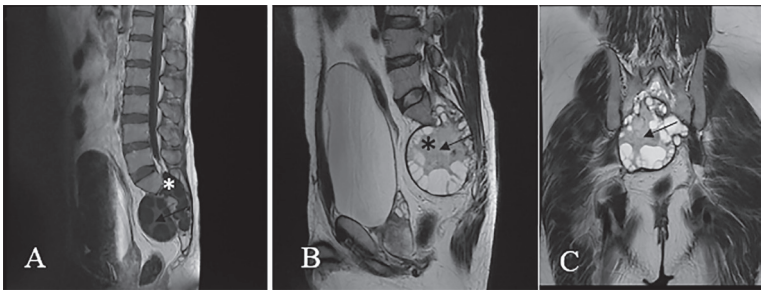

Figure 3: MRI depicts sacral hydatid disease in a 48-year-old man with chronic vague pelvic pain. A. A well-defined multiloculated cyst (arrow) with extension into the spinal canal (asterisk) is seen in a sagittal T1-weighted image. The characteristics of the water lily sign can be seen. B. Sagittal T2-weighted image obtained at the same level as (A) demonstrates the water lily sign with peripherally located high-signal-intensity daughter cysts (asterisk) and a central area with intermediate signal intensity (arrow). C. Coronal T2-weighted image of the same patient shows bone destruction and a multiloculated hydatid cyst (arrow). This figure was obtained from a patient admitted to the authors' institution and was not previously published elsewhere.

frequently negative.[17] For the primary evaluation of echinococcal-related orbital involvement, orbital CT and MRI are the preferred diagnostic modalities. Coronal CT may also be useful in determining the possibility of hydatid cyst erosion of orbital cavity walls. On a CT scan, the cyst is typically seen as a hypodense, unilocular, non-enhancing, or mildly-enhancing homogenous mass with a well-defined hyperdense border (figure 4). The lesion might also demonstrate calcification(s) and multiple septae. Furthermore, thinning and bowing of the surrounding bones can be noted occasionally.[17, 20-22] On CT images, the density of the hydatid cyst resembles that of the vitreous fluid or the cerebrospinal fluid, with a Hounsfield unit ranging from +23 to +68.[19, 23] On MRI, hydatid disease typically manifests as a simple cyst that is hypointense on T1W scans but hyperintense on T2W images. On T2W, the capsule appears as a hypointense rim surrounding the mass (figure 5).[17, 19] MRI can be a useful non-invasive method for evaluating neighboring structures as well as excluding other possible diagnoses. Other orbital cystic lesions that should be considered in the differential diagnosis of hydatid disease include abscesses, intraorbital hematomas, lacrimal tumors/cysts, mucoceles, and lymphangiomas.[24] The findings of CT and MRI scans are strongly suggestive of a hydatid cyst diagnosis when used in the proper context; however, a pathological examination is still required to confirm the diagnosis. Given the rarity of primary ocular involvement, further evaluation with abdominal ultrasound and chest imaging is recommended to determine whether there are any other hydatid cysts in the liver or lungs.[25]

Preoperative treatment with antihelminthic medications such as albendazole and mebendazole, if continued for at least two months, can reduce the size of the orbital cystic lesion. Additionally, to destroy the germinal layer

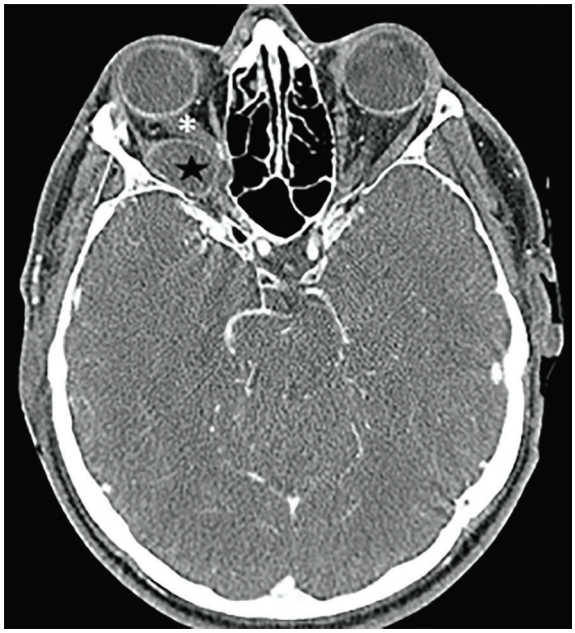

Figure 4: This figure demonstrates orbital hydatid disease in a 62-year-old man with right-sided proptosis, ocular pain, and decreased visual acuity from three months ago. Axial contrast-enhanced CT scan of the orbit displays an intraconal, unilocular, well-defined cyst (black star) with peripheral rim enhancement adjacent to the lateral rectus muscle (white arrow) that is suggestive of orbital hydatid cyst. As shown, a significant pressure effect caused the medial displacement of the right optic nerve (white asterisk). The patient underwent surgical excision of the cyst, and histopathologic examination confirmed the diagnosis of hydatid disease. Following cyst excision, the patients' post-operative visual acuity improved significantly. This figure was obtained from a patient admitted to the authors' institution and was not previously published elsewhere.

and scolices, cytocidal agents such as hypertonic saline or ethanol can be injected into the cyst. Nevertheless, intact cyst excision without cystic rupture is the gold standard treatment, which can help in the complete removal of the lesion.[26]

Mediastinal Hydatid Cyst

Mediastinal hydatid cysts are quite rare, even in endemic areas. The mediastinal hydatid

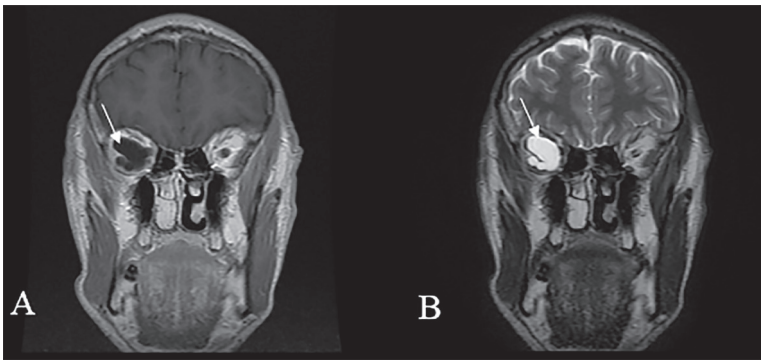

Figure 5: This figure shows orbital hydatid disease in a 62-year-old man with right-sided proptosis, ocular pain, and decreased visual acuity from three months ago. Coronal MR imaging at the orbital apex level shows a well-demarcated simple cyst with a hypointense signal on T1W and a hyperintense signal on T2W. The cystic lesion is located in the posterior aspect of the orbit, compatible with a diagnosis of hydatid cyst (arrow). The patient underwent surgical excision of the cyst and histopathologic examination confirmed the diagnosis of hydatid disease. The patients' post-operative visual acuity improved significantly following cyst excision. This figure was obtained from a patient admitted to the authors' institution and was not previously published elsewhere.

cyst can affect people of all ages; however, it is mostly seen in young patients.[5, 27, 28] Mediastinal involvement can be either primary or secondary to coastal, vertebral, pulmonary, or hepatic hydatid disease.[29] Primary mediastinal involvement is extremely rare and accounts for less than 0.1% of all hydatid disease cases.[27] It is assumed that primary mediastinal disease is caused by the localization of the parasite following lymphatic dissemination or hematogenous distribution via a branch of the thoracic aorta.[30] Secondary mediastinal hydatidosis is more frequent than primary involvement, and the main pathogenesis of secondary mediastinal involvement is mediastinal contamination caused by asymptomatic or post-traumatic micro-rupture of the primary hydatid cyst, and echinococcal fluid leakage during surgery. Hydatid cysts may be solitary or multiple and can infect the anterior, middle, or posterior mediastinal compartments. Overall, the posterior mediastinum is the preferred site for the echinococcal parasite localization; nevertheless, primary hydatid disease is mostly seen in the anterior mediastinum.[27, 31]

Mediastinal hydatid cysts complications and symptoms are typically caused by the involvement of adjacent structures and thus vary based on the size and location of the cysts. Dyspnea and cough, retrosternal chest pain, dysphagia, dysphasia, back pain, and superior vena cava syndrome are only a few of these symptoms.[32] Due to the presence of multiple vital organs within the mediastinum and consequently, the risk of developing life-threatening complications, timely diagnosis is crucial.

Imaging modalities such as plain radiography and CT are beneficial as primary tools for detecting cysts. There is no pathognomonic sign for mediastinal hydatidosis, and imaging manifestations may vary.[33] Unilocular, multilocular, complicated, and calcified round cysts are among the various imaging presentations. Besides, mediastinal widening was reported.[33, 34] The presence of cystic wall calcification and/or a high amount of air fluid provides additional clues to the diagnosis of the mediastinal hydatid cyst.[27] Abdominal ultrasonography and thoracic MRI, respectively, should be carried out if there is any indication of primary hepatic or spinal involvement.

In endemic areas, mediastinal hydatid cysts should always be kept in mind, especially when accompanied by mediastinal compression signs (figures 6A and B).[35] Depending on the mediastinal compartment that is involved, other cystic lesions such as meningocele, mature cystic teratoma, thymic lymphangioma, thymoma, mediastinal carcinoma, and bronchogenic, pleuropericardial, or neurenteric benign masses should be taken into consideration in the differential diagnosis.[32]

After three months of medical treatment with antihelminthic medications, the patient's symptoms improved, and the size of the cysts significantly decreased.

Soft Tissue Hydatid Cyst

Even in endemic areas, soft tissue involvement is a rare clinical entity of echinococcal infection. It accounts for about 2-3% of all hydatidosis in these areas.[36, 37] Furthermore, the head and neck seem to be one of the rarest primary sites of soft tissue involvement.[38] The most common manifestation of soft tissue hydatid disease is a slow-growing painless mass. In case of rupture of the cyst(s), edema and acute inflammation may also occur.[34] The sensitivity and specificity of the enzyme-linked immunosorbent assay (ELISA) for the diagnosis of hydatid disease in organs other than the liver are comparatively low.[38]

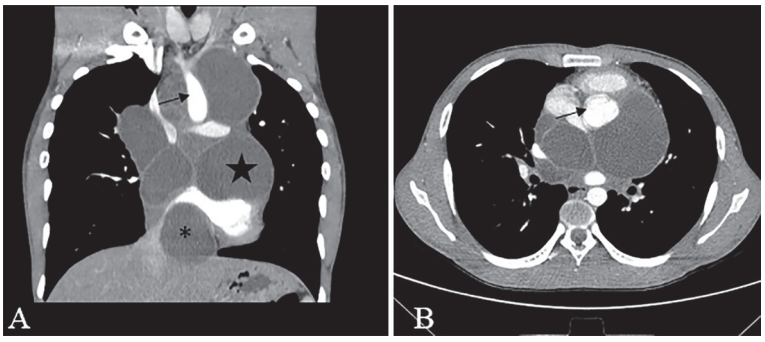

Figure 6: This figure demonstrates mediastinal hydatid disease in a 36-year-old man with persistent fever, cough, weight loss, and dyspnea. Chest radiography showed both mediastinal widening and multiple lobulated mediastinal opacities. A. Coronal contrast-enhanced CT scan of the chest shows several cysts of various sizes within the mediastinum as well as encasement of mediastinal vessels without evidence of thrombosis (arrow). Simple-appearing cysts are also noted within the heart at the junction of the IVC and right atrium (asterisk), as well as the atrioventricular junction (star). B. Axial CT scan of the same patient at the carina level indicates multiple mediastinal cysts pushing the aorta anteriorly (arrow) and causing severe vascular encasement. After three months of medical treatment with antihelminthic medication, the patient's symptoms improved, and a significant decrease in the size of the cysts was observed. This figure was obtained from a patient who was admitted to the authors' institution and was not previously published elsewhere.

Although positive serology has a high diagnostic value, a negative result does not exclude the possibility of echinococcal infection.[37]

Since soft tissue hydatidosis may resemble soft tissue tumors, radiographic evaluation is essential to avoid unnecessary invasive procedures such as biopsy. In addition, in the case of hydatidosis, a biopsy may cause cystic rupture and fluid leakage, which can lead to anaphylaxis or systemic infection.[36] In general, radiologic diagnosis of soft tissue hydatid cysts is difficult and poses a challenge to the radiologist. Furthermore, the diagnosis is more challenging in non-endemic areas due to the rarity of soft tissue involvement. Soft tissue hydatid cysts may present as unilocular, multilocular, with daughter vesicles or detached membranes, or complicated cystic lesions with or without calcification. The differential diagnosis of soft tissue involvement includes a wide range of benign and malignant disorders including abscess, hematoma, synovial cyst, necrotic lymphadenopathy, necrotic soft tissue tumors, and lymphangioma.[12]

A primary ultrasound and/or CT examination is a useful diagnostic procedure that helps display daughter cysts, internal membranes, and calcifications in cystic masses (figure 7).[1, 37] Further evaluation with MRI is usually performed to assess the multiplicity and extent of the lesion, its relationship with adjacent tissues, and its differentiation from other soft tissue masses.[12] MRI is also considered the gold standard imaging modality for evaluating complicated or solid-cystic lesions.[39, 40] On T2-weighted MR imaging, a low signal intensity mass with a hypointense rim is considered a typical sign of soft-tissue hydatid disease.[41] This non-specific imaging feature is uncommon in hydatid cysts located elsewhere in the body.[34]

As previously stated, since primary liver or lung involvement soft tissue hydatid disease is extremely rare, chest and abdominal imaging should be performed. The preferred treatment is complete surgical excision without rupture. Preoperatively, daughter cysts and scolices

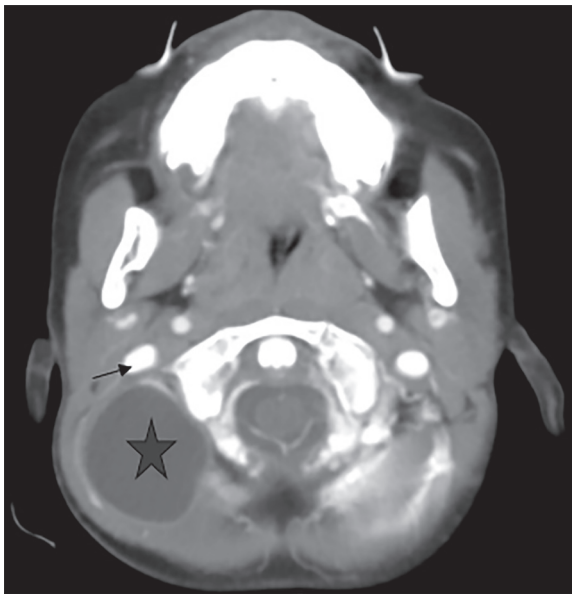

Figure 7: Soft tissue hydatid disease is depicted in a six-year-old boy with a non-tender, well-defined, mobile subcutaneous mass on the right side of his neck. Ultrasound was suggestive of an anechoic thin-walled cyst with posterior acoustic enhancement and no internal echogenicity or abnormal vascularity. Axial contrast-enhanced CT of the neck demonstrates a unilocular, well-defined cyst (star) with peripheral rim enhancement in the right posterior triangle of the neck, posterior to the carotid sheath (arrow), causing anterior displacement of the sheath. There is no evidence of severe inflammation in the adjacent soft tissues. With the suspicion of a branchial cleft cyst, the patient underwent surgical excision; however, a subsequent histopathologic examination was in favor of soft tissue hydatid disease. This figure was obtained from a patient who was admitted to the authors' institution and was not previously published elsewhere.

should be inactivated by injecting 20% hypertonic saline, 5% silver nitrate, or formalin into the cyst. To avoid recurrence, postoperative medical treatment with albendazole or mebendazole is recommended.[38, 42]

Renal Hydatid Disease

Although kidney involvement in hydatid disease is uncommon, it has been observed in up to 3% of all hydatid cases. The majority of the time, renal hydatidosis symptoms are nonspecific and do not often develop early in the course of the condition. The most common presenting symptoms of patients with renal hydatid cysts include flank discomfort, lumbar or abdominal mass, subcostal pain, vomiting, fever, dysuria, and hematuria. In a minority of patients, in about 20% of cases, hydatid cysts may rupture into the collecting system and cause hydatiduria. Renal hydatid cysts are typically unilateral, solitary, and located within the lower or upper pole of the kidney. Similar to other hydatid cysts, integration of clinical, radiological, and serological examinations could yield a diagnosis of renal hydatid disease before surgery; however, imaging is the primary diagnostic tool.[43, 44]

Since ultrasound is readily available, inexpensive, safe, and highly reliable, it is the most valuable imaging modality in the detection and differentiation of renal cystic lesions. The ultrasonographic features of renal hydatidosis are similar to those of hydatid cysts located elsewhere. These features include the snowstorm sign seen in cysts with multiple echogenic foci due to hydatid sand, a fluid-filled cystic lesion with floating membranes caused by the detachment of the endocyst from the pericyst, a cyst wall depicted by double echogenic lines separated by a hypoechogenic layer, and the wheel-spoke or racemose pattern.[2, 45] If ultrasonography cannot confirm a diagnosis of hydatid disease, a CT scan should be performed (figure 8). On imaging, renal hydatid disease might seem like other renal cystic lesions, such as cystic nephroma, simple renal cyst, necrotic renal cell carcinoma, and renal abscess.[1, 44] Although ring-like or curvilinear calcification corresponding to a pericyst can suggest the diagnosis of hydatid disease, it is not simply specific to hydatid disease. Up to 5% of benign simple cysts may demonstrate peripheral calcification on imaging.[44]

Various therapeutic approaches have been investigated for renal hydatidosis, including medical therapy with mebendazole or albendazole followed by percutaneous drainage with the PAIR technique (puncture, aspiration, injection, and re-aspiration of protoscolicidal agents) or surgery. Surgical techniques include cystectomy combined with partial pericystectomy, and nephrectomy, either partial or complete, via laparoscopy or laparotomy, depending on the size, location, and presence of superimposed infections and/or daughter cysts. In addition, postoperative medical therapy can reduce the risk of disease recurrence. During surgery, spillage caused by cyst rupture must be avoided, as it can result in subsequent anaphylactic shock. Since hydatid disease is benign in nature, total nephrectomy should only be performed in patients with non-functioning kidneys. Importantly, early detection of renal hydatid disease allows for the best possible treatment with renal-conservation surgeries.[44-46]

Hydatid Disease of the Central Nervous System

Preoperative diagnosis of central nervous system (CNS) echinococcosis, which accounts for approximately 1-3% of cases, is not always

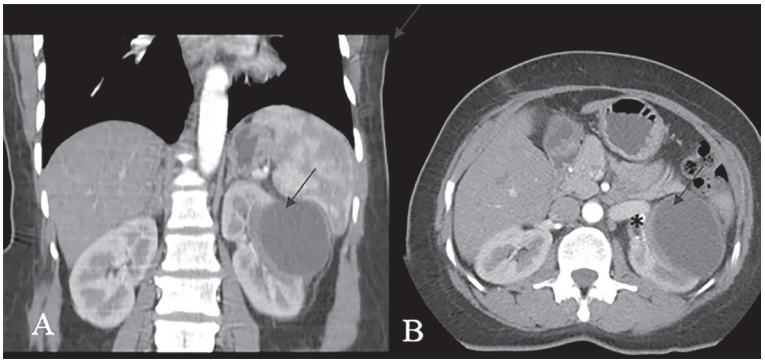

Figure 8: Renal hydatid disease is shown in a 54-year-old woman with flank pain and intermittent hematuria lasting for several months. Ultrasonography revealed a thin-walled cyst with no interior solid component in the mid-pole of the left kidney. Coronal (A) and axial (B) contrast-enhanced abdominopelvic CT images show a unilocular thin-walled cyst in the interpolar region of the left kidney (arrow) with no enhancing solid components in the arterial phase, compatible with Bosniak type I renal cyst. Ultrasound-guided percutaneous drainage of the cysts was performed, and aspirated fluid serology was positive for *Echinococcus granulosus*. This figure was obtained from a patient admitted to the authors' institution and was not previously published elsewhere.

possible.[2, 47] In patients with cerebral hydatid disease, the involvement of other organs is frequently observed. Patients with increased intracranial pressure frequently experience signs and symptoms such as headache, vomiting, and papilledema. Peripheral neuropathy, skull deformities, mental changes, and seizures may also occur. Intracranial hydatid cysts are most commonly supratentorial and involve regions of the parietal and frontal lobes within the middle cerebral artery territory. Ventricular involvement is uncommon in these patients. Multiple cysts are rare and are mainly caused by the spontaneous, traumatic, or surgical rupture of a solitary lesion.[48] MRI is the best imaging modality for identifying intracranial hydatid cysts. On MRI, cysts present as thin-walled, non-homogeneous space-occupying lesions with no intraluminal degradation products. While T2-weighted scans of these lesions show hyperintensity, T1-weighted images show iso- or hypointensity. Brain CT can also be used to evaluate cerebral hydatid disease, particularly when there is bone involvement. Following intravenous contrast injection, cysts appeared on CT as well-defined, thin-walled unilocular lesions with peripheral enhancement.[47, 49] Brain abscesses, intracranial arachnoid cysts, and tumors are all possible differential diagnoses. Unlike hydatid disease, perilesional edema is typically found in cystic tumors and abscesses. The standard of care for those with cerebral hydatid disease is the surgical removal of the cyst, which is mainly performed using the Dowling-Orlando procedure. Cysts may be more easily located prior to surgery with the help of radiologic examinations. Medical treatment with albendazole may be helpful for patients who are unable or unwilling to undergo surgery.[48]

Splenic Hydatid Cyst

Splenic hydatidosis affects approximately 2% of all echinococcosis cases and nearly 4% of patients with abdominal hydatid disease.[50, 51] The most common pathology of parasitic splenic cysts is *Echinococcus granulosus*, to the point where, in endemic areas, hydatidosis causes 50-80% of cystic lesions of the spleen.[52] The major routes for splenic infestation are thought to be systemic dissemination and/or intraperitoneal spread from a ruptured liver cyst. Splenic hydatid disease is generally asymptomatic and is usually detected incidentally as an abdominal mass, mainly in the left hypochondrium and less commonly in the epigastrium.[53] If the cyst grows in size, patients will experience non-specific symptoms such as abdominal pain and fever, as well as a painful mass in the left hypochondrium.[50] In rare cases, the pressure effect may cause dyspnea, dyspepsia, colon fistula, and perforation into the diaphragm or bronchial tree.[54] Similar to hydatid cysts of other organs, mismanagement of splenic hydatidosis can result in secondary infection, cyst rupture, and even anaphylactic shock.[51]

Epidermoid cysts, pseudocysts, solitary abscesses, cystic tumors, or hematoma are among the differential diagnosis of splenic cystic lesions.[54] On imaging, splenic hydatidosis looks extremely similar to other splenic cystic lesions, posing a diagnostic challenge. The presence of daughter cysts within a larger cystic lesion or concurrent cystic lesions in other organs, particularly in the liver, should favor the diagnosis of splenic hydatidosis. Nevertheless, in endemic areas, echinococcosis should be the primary suspicion in patients with splenic cysts, unless proven otherwise.[55]

Plain radiography may reveal a marginal egg-shell calcification in the left hypochondrium, however, CT and ultrasound represent the most useful imaging modalities for the diagnosis and assessment of the splenic lesions.[56] In general, a splenic hydatid cyst can present as a purely cystic to a completely solid lesion. In detecting splenic hydatid cysts, ultrasound has a sensitivity of nearly 90-95%. The most common finding on ultrasound is an anechoic smooth, round cyst, which resembles most benign cysts. Furthermore, the presence of membranes can cause mixed echoes, making it difficult to differentiate from an abscess or neoplasm.[51, 54] Nevertheless, internal septations, separation of the hydatid membrane from the wall of the cyst, and hydatid sand can all be the signs of hydatidosis.[57] The hydatid cysts can be differentiated from simple cysts, cystic tumors, pseudocysts, and metastases using the pathognomic snake/serpent sign, water-lily sign, and double-line sign.[58, 59] After medical therapy, ultrasound can also be used to monitor response to therapy by showing detached membranes within the degenerated cyst.[60]

When compared to ultrasound, CT is a superior modality, with sensitivity rates as high as 95-100%.[61] CT is more effective than radiography or ultrasound in demonstrating cyst wall calcification, which is usually curvilinear or ring-shaped.[51] Due to intra-cystic debris, hydatid sand, and inflammatory cells, CT may display the typical high-attenuation linear wall as well as daughter vesicles within the cyst.[62] The density of daughter cysts ranges from 0 to 15 H, which is lower than the mother cyst fluid, which has a density of 30-40 H.[54] However, MRI can be helpful in situations where ultrasonography and

CT findings are unclear, and serology results are negative. The “low-signal intensity rim”, which is best depicted on the T2-weighted sequence of MRI, is helpful for hydatid cyst diagnosis.[63]

Surgery, either full or partial splenectomy, is typically used to treat splenic hydatidosis. Treatment with albendazole is recommended during the postoperative follow-up phase.[50]

Conclusion

Hydatid disease has the potential to involve any organ and thus presents with a wide spectrum of indications. Timely diagnosis and proper management are required to prevent life-threatening conditions such as anaphylactic shock or pressure effect on vital organs. When encountering cystic lesions at any anatomical site in endemic areas, the radiologist should be aware to include hydatid disease in the differential diagnosis. Besides the essential role of imaging in directing accurate diagnosis, it is also beneficial for determining the extent to which adjacent structures are involved and for guiding treatment planning strategies.

Author’s Contribution

N.K, P.I, N.K, and S.H: Conception/design of the work, acquisition, analysis, interpretation of data, drafting and critical revision; All authors have read and approved the final manuscript and agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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