Rare localizations of echinococcosis: our personal experience

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Aims and objectives

Echinococcosis (or hydatid disease) is the most widespread serious human parasitic infection in the world caused by a cestode: *Echinococcus granulosus*. It is endemic in areas with tropical or subtropical climates particularly in Mediterranean region, South America, Africa and Australia due to the close association between humans and domestic animals (1). Hydatid disease, common in Mediterranean countries, still remains endemic in Tunisia. Man is an accidental, intermediate host, and infection of humans represents a terminal event for the parasite. In fact, humans become infected through contact with definite host or consumption of contaminated food. After it enters the body, larval stage of *Echinococcus* parasite forms cysts. Since the cysts are very slowly growing, the symptoms are seen several years after primary infection.

According to the literature, hydatid disease may develop in almost any part of the body. The liver is the most frequently involved organ (75%), followed by the lung (15%) and the remainder of the body (2). Primary rare localizations of parasitosis range approximately from 5% to 30% but the exact incidence of unusual locations is difficult to ascertain as they are only reported as few sporadic cases (3). The rare localizations of hydatidosis lead to atypical clinical presentation causing difficulties in establishing the diagnosis. Definite diagnosis is mostly based on imaging techniques: ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI).

We present our experience with atypical sites of hydatidosis, including different imaging techniques for diagnostic evaluation.
Methods and materials

The case sample analyzed consisted of 9 patients (6 females and 3 males). The age ranged from 10 years to 68 years with mean age of 38 years. The mean disease duration was 10 months.

The atypical locations of the disease included: brain, orbit, neck, mediastinum, intraperitoneal cavity, pelvis, adrenal gland, psoas muscle and quadriceps muscle.

Symptoms varied according to the localization of the cyst. Palpable lump was a presenting symptom in patients with subcutaneous and muscle cysts. Four patients with atypical locations were asymptomatic, which included cases with mediastinal, peritoneal, pelvic and adrenal gland hydatid. The patient with brain hydatid presented with neurological symptoms, and exophthalmia was observed in the patient with orbital cyst.

Preoperative diagnosis was established by the history, clinical examination, complete blood counts, liver and kidney function tests, serological tests (enzyme linked immunosorbent assay and indirect hemagglutination test), X-ray chest, ultrasound, MRI and contrast enhanced CT. All patients were scanned with a 16-row multidetector CT scanner, and eight patients were explored with a 1,5 Tesla MRI using unenhanced T1-weighted sequences, T2-weighted sequences and contrast-enhanced T1-weighted fat-suppressed sequences, with T2 FLAIR sequences for cerebral location.

The treatment was surgical for 8 patients and medical for one patient. All the operated patients received preoperative and postoperative Albendazole chemotherapy.

Eosinophilia was observed in 6 patients. Diagnosis was confirmed by hydatid serology, ultrasonography, MRI and CT, with histopathological examination of the specimen for the operated patients.
Results

Hydatid disease may involve almost any anatomic site due to hematogenous dissemination. Growth of hydatid cyst in different organs of the body is variable and depends both on patient factors, parasite factors, location of organ of cyst, host reaction and presence of any complications. Compressible organs such as the brain facilitate the growth of the cyst. Free space around cyst usually makes easy growth of the cyst (4).

The diagnosis is often difficult when hydatid cyst occurs at unusual sites as the imaging appearance varies at different sites. Hydatid disease is often manifested by slowly growing cystic mass. Cysts may be single or multiple, uni- or multiloculated and thin- or thick walled. More specific signs include visualization or calcification of the cyst wall, presence of daughter cysts and membrane detachment (2). However, hydatid cysts with unusual localizations may cause serious problems in the differential diagnosis. CT and MR imaging, alone or in combination, are helpful in the diagnosis of hydatid disease. Although CT is superior in detecting calcification of the cyst wall or septa, MRI is better in detecting multiplicity and defining the anatomic relationship of the lesion with the adjacent structures and helps in surgical planning (5, 6).

A high index of suspicion, radiological investigations as well as histopathological examination are necessary in establishing the diagnosis of hydatid disease at unusual sites in the body.

Brain:

Hydatid disease affects rarely the central nervous system (1% of cases) and is more common in children (4, 5). Intracranial hydatid cysts are commonly solitary (Figure 1) and multiple cysts are rare. Intracranial hydatid cysts may also be classified as primary or secondary depending on whether other organs haven't / have been involved (4). The secondary multiple cysts result from spontaneous, traumatic or surgical rupture of the primary intracranial cyst (Figure 2). Patients with intracranial hydatid cysts usually present with focal neurological deficit and features of raised intracranial pressure.

The typical intracranial hydatid cysts present as a well defined solitary oval or cystic mass in the middle cerebral artery territory in parietal lobes, although they can be seen in any location including skull vault, extradural, intraventricular, meningeal, posterior fossa and brainstem (4).
CT and MR imaging demonstrate attenuation or signal intensity similar to that of cerebrospinal fluid (Figure 1, 3). Although the lesion may cause extrinsic compression of the ventricular system with subsequent hydrocephalus, there is no associated edema as is typically seen in abscesses and cystic tumors (5). Lesion does not enhance after intravenous administration of contrast material, and calcification is extremely rare. When present, edema and post contrast enhancement indicate ongoing inflammation. Presence of significant edema may indicate rupture of the cyst and may be present in post operative cases (Figure 2). Such cases are difficult to differentiate from other cystic lesions with enhancement and peripheral edema such as abscesses, large granulomas or cystic gliomas (7).

Kohli et al. (8) performed in vivo MR spectroscopy (MRS) studies in a patient with intracranial hydatid cyst and found, besides lactate, alanine and acetate, a large resonance for pyruvate. The MRS pattern appeared different from the other cystic lesions of brain and they have suggested MRS as an adjunct to imaging in the differential diagnosis of intracranial hydatids.

Surgically, intact cyst excision is the ideal treatment. Medical treatment seems to be beneficial both pre- and post-operatively (4).

**Orbit:**

Orbital involvement occurs for 0.3% of all hydatid cysts and it is more prevalent among young patients (5, 9). Typically, an orbital hydatid cyst is unilateral and occurs with or without hydatid cysts located elsewhere in the body. It tends to involve the retrobulbar tissues either within the muscle cone or outside in the superolateral or superomedial angle. The most frequent clinical findings are proptosis, chemosis, lid edema, visual impairment, and restriction of extraocular motility (10).

According to clinical and radiological findings, the differential diagnosis of orbital hydatid cyst includes any well-circumscribed, non pulsatile orbital lesions such as abscess, epidermoid-dermoid cysts, teratoma, and mucocele (11). Serological tests can be used for the diagnosis of echinococcus, but they usually give negative results.

Radiological findings play an important role for preoperative differential diagnosis. Orbital ultrasonography is a useful test when a "double wall sign" has been observed (10). MR imaging and CT are both useful techniques for preoperative diagnosis and surgical planning. CT of the orbital hydatid cyst demonstrates a hypodense, unilocular, well-defined homogenous mass with a hyperdense rim (Figure 4). MRI demonstrates a cystic lesion with a low signal intensity on T1-weighted images and high signal intensity on T2-
weighted images, without rim enhancement after contrast medium administration (Figure 5) (11).

The most appropriate treatment of an orbital hydatid cyst is surgical removal. However, dissemination of the disease due to cyst rupture during surgery is the most feared complication. Endonasal endoscopic aspiration has been described as a simple treatment with minimum postoperative reaction and complications. In addition to surgical treatment, Albendazole treatment is useful, especially if used as an adjunctive therapy to surgery (10, 11).

**Neck:**

Hydatid disease in the neck is quite rare, even in areas where the disease is endemic, with only a few cases reported in the literature (12). The diagnosis of cervical Echinococcus infection mainly depends on the clinical history of the patient, diagnostic radiological findings and serologic tests. Cervical hydatid cysts are usually slow-growing, fluctuant, painless masses.

Histopathological evaluation of the excised specimen and fine needle aspiration cytology usually leads to the diagnosis. Since puncture of the cyst may lead to an anaphylactic reaction due to spillage of hydatid fluid, the use of fine needle aspiration is controversial at present (12). Imaging modalities like Ultrasound, CT and MRI remain more sensitive than serodiagnosis. These techniques help to determine the cystic avascular nature of the lesion (Figure 6, 7). Daughter cysts, vesicles and internal septa can also be demonstrated (13).

The differential diagnosis includes branchial cleft cyst, bronchogenic, thymic, parathyroid, thoracic duct, and foregut-derived (esophageal duplication) cysts, pseudocysts or benign tumor that is congenital and acquired cystic lesions of the neck (13).

Surgical removal is the most effective treatment of hydatid cyst. The cyst should be excised as a whole without being ruptured to prevent any complications.

**Mediastinum:**

Primary mediastinal hydatid cysts are quite rare. They can be found at various locations within the mediastinum. Symptoms of mediastinal hydatid cysts depend on their size and location and whether or not they exert any compression on the neighboring structures.
The most serious complications include leakage of the hydatid material to the neighboring vessel, causing embolization and/or anaphylaxis with possible fatal consequences (14).

Diagnosis can be obtained through the combined assessment of clinical, radiological, laboratory, and historic data of patient. Hydatid serology is the only biologic aid to preoperative diagnosis. Its negativity does not exclude the diagnosis (15).

Chest radiograph, Ultrasound, CT and MRI facilitate diagnosis. Chest radiograph oriented the diagnosis by showing a mediastinal water tone, often rounded or oval (16). Thoracic ultrasound allows confirmation of the diagnosis when the lesion is accessible. Thoracic ultrasound also reveals the fluid character of the opacity and, in many cases, the proliferous membrane, pathognomonic of hydatid cysts (15). CT is considered essential and is important for displaying morphology, density, and limits of these lesions. It often accurately defines the relationship of the lesion to the adjacent structures. The most common CT finding of a mediastinal hydatid cyst is a homogenous mass with fluid (Figure 8). The presence of peripheral calcifications supports diagnosis (14). MR imaging, with its superior soft tissue resolution, can also provide us with a clear delineation of the cyst.

The differential diagnosis includes other mediastinal cystic masses, such as enteric cyst, cystic lymphangioma, pleuroperticardial cyst and bronchogenic cyst (15).

Surgery is the principal method of treatment: cystotomy and aspiration are performed. When localization of the cyst and invasion to vital structures prevent the total excision, partial pericystectomy is the treatment of choice (14).

**Intraperitoneum and pelvis:**

Peritoneal hydatidosis is almost always secondary to hepatic disease, although some unusual cases of primary peritoneal hydatidosis have been described (2). The overall frequency of peritoneal disease in cases of abdominal echinococcus is approximately 13% (17). Peritoneal involvement is usually undetected unless cysts are large enough to cause symptoms. Most of the cases of peritoneal hydatid disease are secondary to previous surgery for liver hydatidosis.

Accurate and rapid diagnosis of peritoneal hydatid disease is possible because of the availability of modern imaging techniques, and the surgical procedures are decided on radiological findings (Figure 9, 10). Ultrasound and CT scan are the radiological methods of choice for assessing the number of hydatid cysts in the abdomen and assessing the changes in size, number and density of lesions in response to drug therapy (18).
Ultrasound is particularly useful for detection of cystic membrane, septa, and to look for hydatid sand. CT scan best demonstrates cyst wall calcification and cyst infection, and enables better delineation of extent of disease (Figure 9). Thickening, calcification of wall, reduction in size and number of cysts are taken as therapeutic response on follow up (17).

Serological tests are being carried out for the diagnosis, screening and post-operative follow up for recurrence.

The treatment of choice for localized hydatid cysts in liver or lungs is principally surgical while the therapy for disseminated peritoneal hydatidosis remains medical (18).

Although primary peritoneal and pelvic involvement of the hydatid cyst generally arise with the second inoculation, the distinctive feature of pelvic and peritoneal mass should be considered for regions where hydatidosis is endemic. It could be diagnosed missing as ovarian or peritoneal carcinoma at the pre-operative period.

**Adrenal Gland:**

Hydatid cyst of the adrenal gland is rare as this entity has been reported in only 7% of all adrenal cysts and constitutes less than 1% of all cases (19). Adrenal hydatid cysts usually form in association with generalized echinococcosis. On the other hand, primary hydatid cysts of the adrenal gland are extremely rare.

Most adrenal cysts are asymptomatic, they are usually found as incidental findings on imaging studies or incidentally during surgery. When symptoms are present, most are related to local visceral compression. Rarely, endocrine abnormalities like arterial hypertension are seen in adrenal hydatidosis (4). The complications of an adrenal gland hydatid cyst include rupture in the peritoneum or retroperitoneum, local infection, fistula, hemorrhage, or compression of adjacent tissues (9).

Serological studies may assist in diagnosis, but lack both sensitivity and specificity (19).

Radiological imaging techniques provide an important aid in the diagnosis and follow up. Hydatid cyst identification in the adrenal gland is based mainly on ultrasound and CT scan (Figure 11, 12). MRI shows the characteristic low signal intensity rim of the hydatid cyst on T2-weighted images. Both MR and CT images are able to show the exact anatomic extent, size, volume and position of the mass, the number of cysts, the relationship to other organs and possible complications (9). The presence of calcifications in the adrenal
mass very much supports a diagnosis of hydatidosis. However, the definitive diagnosis is made by macroscopic and microscopic examination of the cyst.

The differential diagnosis of an adrenal cyst include an endothelial cyst, a pseudocyst due to infarction or hemorrhage, a cystic neoplasm like lymphangioma, a posttraumatic cyst, a cystic phaeochromocytoma, an abscess and other congenital or acquired cysts (19).

Treatment of hydatid disease of adrenals is mostly surgical and the operation of choice is removal of cyst preserving ipsilateral kidney and, if possible, the remaining adrenal gland to provide adequate haemostasis (9, 19). Secondary infestation can be prevented by intraoperative injection of concentrated saline into the cyst.

**Muscle:**

Primary muscular hydatid cysts are rare, accounting for 3% of all patients with hydatidosis (4, 9). Muscular hydatidosis usually occur as isolated lesions without hepatic or pulmonary lesions.

The pathogenesis of muscular localization remains ill understood; but most authors believe embryo can reach the muscles from the systemic circulation after leaving the intestine and passing through two filters: the liver and the lungs (20). However, the primary muscular hydatid cyst is extremely rare even in endemic zones. This infrequency of muscular locations is explained by continual muscular contractions and the production of lactic acid which prevent scolex. These muscular cysts are revealed as a painless enlarging soft tissue masses.

The tropism to the muscles of trunk and the root of the limbs can be explained the increased vascularization and the decreased muscular activity in these areas (21). Primary HCs of the lower extremity muscles are usually solitary and unilocular or multilocular (Figure 13).

Psoas muscle is an unusual location for hydatid cyst accounting for only 1 to 3% of cases and can be unilateral or bilateral (22). Hydatid of psoas muscle can be isolated (Figure 14, 15) or associated with hydatid disease elsewhere in body. Because of location, hydatid of psoas muscle usually remains asymptomatic and is found incidentally or an enlarging cyst can compress adjacent structures.

Serology may not always be helpful in diagnosing primary muscle hydatidosis. A negative test does not rule out the diagnosis of echinococcosis.
Intramuscular infestation may mimic a soft tissue tumor leading to inappropriate cyst rupture with the attendant risks of anaphylaxis and dissemination to other organs, so a preoperative correct diagnosis is important. Ultrasound is a useful tool for orienting the diagnosis of any tumefaction of the soft tissues, showing the size, localization and type of the cyst (23). CT is especially valuable for surgical planning because of the three-dimensional information regarding localization of the cyst. On CT scan they appear as a well-defined cystic lesion with daughter cysts that may contain wall calcification, septae or debris without the enhancement on intravenous contrast (Figure 14) (21).

MRI is the examination of choice in case of suspicion of hydatid disease of muscle, due to its ability to adequately demonstrate most features of hydatid disease, with the exception of calcifications (9). The multiplanar imaging and the excellent soft tissue contrast provide valuable information on the extent of the disease. In addition, MR imaging is superior to ultrasound and CT in depicting an exact compromise of adjacent soft tissues. The classic MRI findings include a multivesicular cyst, a low-intensity rim ("rim sign") on T2-weighted images or a detached membrane (Figure 13, 15). The most pathognomonic sign is that of daughter cysts within larger cysts (23).

The differential diagnosis of a cystic soft tissue mass includes abscess, chronic hematoma, synovial cyst and necrotic malignant soft tissue tumor (2). The clinical history often suggests the diagnosis.

The treatment of choice is surgical excision (pericystectomy), potentially combined with antihelminthic medication. Postoperative medical treatment reduces recurrence rate. Percutaneous aspiration, infusion of scolicidal agents and reaspiration (PAIR) under imaging (ultrasound or CT) guidance can be used as alternative to surgery in inoperable cases (23).
Fig. 1: Cerebral CT: axial (a), sagittal (b) and coronal (c) images after injection of contrast: Left temporo-parietal lobulated mass, unilocular, presenting a thin and regular rim with homogeneous density close to that of CSF, unmodified after injection of contrast, compressing the left lateral ventricle and driving back the midline structures to the right.

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Fig. 2: Cerebral CT: axial images (a, b) after injection of contrast: Postoperative oedemato-hemorrhagic alterations associated with daughter cysts in the left temporal lobe.

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Fig. 3: Cerebral MRI: (a) axial unenhanced T1-weighted image, (b) axial T2 FLAIR -weighted image, (c) axial and (d) coronal contrast-enhanced T1-weighted images: pure fluid mass with low-signal-intensity on T1 and high signal-intensity on T2, canceling completely on FLAIR, surrounded by a thin rim discreetly hyperintense on T1 and hypointense on T2, unmodified after injection of Gadolinium and associated with perilesional edema. This lesion excerts a mass effect on the median structures. Furthermore, extra-dural left frontal abscess with fluid collections of the scalp.

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Fig. 4: Orbito-cerebral CT: axial images before (a) and after (b) injection of contrast: left intra-orbital hypodense, unilocular and homogenous mass with a hyperdense rim, unenhanced after injection of contrast.

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Fig. 5: Orbital-cerebral MRI: (a) axial and (b) coronal fat-saturated T2-weighted images, axial (c) unenhanced and (d) contrast-enhanced T1-weighted images: left intra-orbital cystic lesion with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, without rim enhancement after injection of Gadolinium.

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Fig. 6: Ultrasound axial image: left basi-cervical cystic mass, rounded, heterogeneous and avascular on Doppler, measuring 10 cm, with posterior enhancement of echos.

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Fig. 7: Cervical CT: axial images before (a) and after (b) injection of contrast: subcutaneous cystic mass, thin-waled, unenhanced after injection of contrast and contenting a serpiginous structure realizing the aspect of a detached membrane.

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Fig. 8: Thoracic unenhanced CT: axial images (a, b): homogenous thin-waled médiastinal mass with fluid density and thin parietal calcifications, coming into contact with the right edge of the heart.

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**Fig. 9:** Contrast-enhanced abdominal CT: axial images (a, b, c, d): multiple cystic masses, multi-compartmentalised, unmodified after injection of PDC and localized in the mesentery (a, b, c) and the pelvis (d). Note the presence of a thin parietal calcification (a).

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**Fig. 10**: Pelvic MRI: (a) axial, (b) coronal and (c) sagittal T2-weighted images: Cystic pelvic mass, multi-compartmentalised with high signal intensity on T2-weighted images, and presenting a thin hypointense rim.

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**Fig. 11:** Ultrasound axial image: multi-compartimentalised anechoic mass with posterior enhancement of echos.

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**Fig. 12:** Contrast-enhanced abdominal CT: axial images (a, b): large thin-walled cystic mass, with homogeneous content, driving back the right hepatic lobe.

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**Fig. 13:** MRI of the thigh: coronal unenhanced T1-weighted image (a) and fat-saturated T2-weighted image (b), axial T2-weighted (c) and contrast-enhanced T1-weighted (d) images: a solitary muscular cystic mass, hypointense on T1-weighted images and hyperintense on T2-weighted images with a low-intensity rim on T2-weighted images and a detached membrane, without enhancement after injection of Gadolinium.

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Fig. 14: Contrast-enhanced abdominal CT: axial images (a, b): oval and thin-waled cystic lesions, multi-vesicular, with heterogeneous density, located in the right psoas muscle.

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Fig. 15: MRI of the psoas muscles: coronal (a) unenhanced and (b) contrast-enhanced T1-weighted images, (c) coronal and (d) sagittal fat-saturated T2-weighted images: multiple multi-vesicular cystic lesions, hypointense on T1-weighted images and hyperintense on T2-weighted images, without enhancement after injection of Gadolinium, located in the right psoas muscle. Note the hypointense signal on T1- and T2-weighted images of the internal partitions and the rim.

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Conclusion

Hydatid disease should be considered in the differential diagnosis of the all cystic masses in all anatomic locations, especially when they occur in areas where the disease is endemic. The combination of clinical history, imaging finding, and serologic test results usually help the diagnosis.
References


