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Pictorial review

Uncommon locations of hydatid disease: CT appearances

K. J. Gossios[1], **D. S. Kontoyiannis**[1], **M. Dascalogiannaki**[2], **N. C. Gourtsoyiannis**[2]

[1] Department of Radiology, General Hospital of Ioannina, GR-450 01 Ioannina, Greece
[2] Department of Radiology, University of Crete Medical School, GR-711 10 Iraklion, Greece

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Abstract. Hydatid disease (HD), already known by Hippocrates, is prevalent and widespread in most sheep-raising countries in Asia, Australia, South America, Near East, and southern Europe. The disease is most commonly due to *Echinococcus granulosus* and may occur in any organ or tissue. The location is mostly hepatic (75 %) and pulmonary (15 %), and only 10 % occur in the rest of the body [1]. Imaging modalities such as US, CT, and MR imaging are helpful in diagnosing the disease. The reliability of each method depends on the cyst's location in the body [2–5]. The purpose of this essay is to illustrate the use of CT in depicting some unusual locations of HD besides the liver and lung.

Key words: Hydatid disease – Computed tomography

Introduction

The main form of hydatid disease (HD) is due to *Echinococcus granulosus* and the mechanism of cyst formation is well established [1]. Humans may contract the infection either by direct contact with a dog which is the definitive host or by ingestion of foods or fluids contaminated by the eggs, which are contained in the feces of the dog. After ingestion, the eggs are freed from their coating and larvae penetrate the mucosa of the jejunum reaching through the venous and lymphatic channels to every region of the body where they transform into small cysts. The cyst wall is a laminated membrane lined by a germinal layer. Daughter cysts arise from the germinal layer probably by endoproliferation.

The growth of hydatid cysts is usually slow and asymptomatic, and clinical manifestations are caused by compression of the involved organ. The disease may mimic benign or malignant primary tumors, single or multiple metastases, cysts, abscesses, empyemas, infarcts, and other lesions [6]. However, if not detected, the cyst may become life-threatening and rupture in vital structures may occur.

Radiological signs are usually non-specific and the serologic tests (ELISA with the presence of the arc 5 of Capron) are extremely useful in the diagnosis. A "suspicious" radiologic diagnosis of HD can be strongly supported if the arc 5 of Capron is demonstrated in serological tests [7].

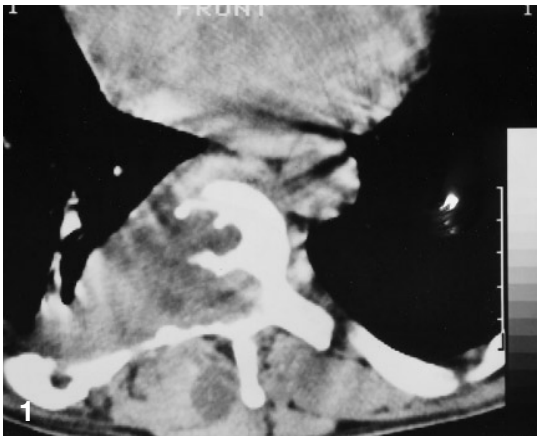
The pathological diagnosis can be made if scolices, hooklets, and/or pieces of parasitic membranes are found in sputum, stool, urine, or in the specimen which has been received by fine-needle aspiration biopsy. Their recognition is not easy; thus, the clinical and radiological examinations have an important role in the diagnosis [8].

Musculoskeletal system

Skeletal involvement by primary HD is uncommon and represents 0.5–4 % of patients. It occurs in the more highly vascularized areas of the bones. The vertebrae, long-bone epiphyses, ilium, skull, ribs, and soft tissue extension are most frequently affected. Lesions in long bones may present with pathological fractures showing no evidence of repair and spinal disease usually presents with cord compression. The appearance of HD in bone is not specific and frequently needs a differential diagnosis.

The CT appearances of bone lesions are similar to those demonstrated on plain films. A well-defined, typically multiloculated, osteolytic lesion sometimes with coarse trabeculae within it is usually seen, giving a honeycomb appearance, which is accompanied by expansion of the bone and thinning of its cortex [9]. However, CT is more accurate in delineating the area of destruction while making the interpretation easier. The primary role of CT is in the recognition of the extraosseous spread of the HD within the soft tissues which may be quite variable. It may have the typical pattern of a cystic

Correspondence to: K.J. Gossios



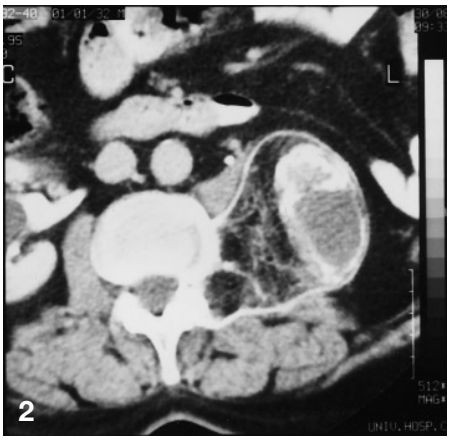
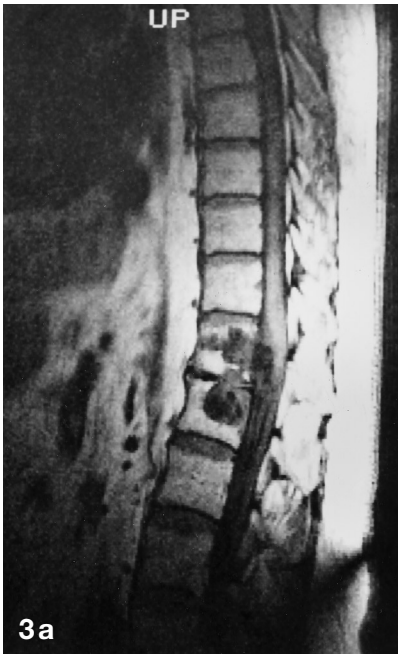
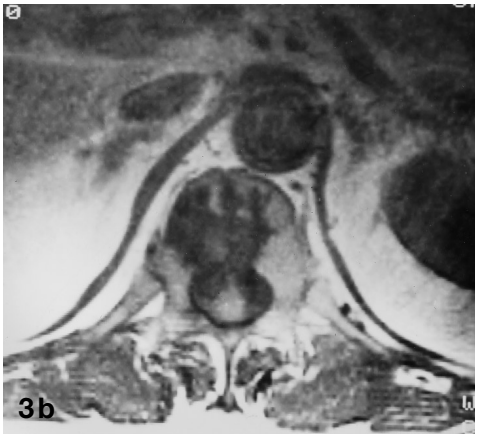


Fig. 1. Spinal hydatid disease (HD). Enhanced CT shows a soft tissue mass in T9 vertebral body which extends to the paravertebral soft tissues and infiltrates the right pedicle. The mass is heterogeneous with high CT numbers. The paravertebral component of the lesion is the most extensive and invades to the spinal canal causing paraplegia





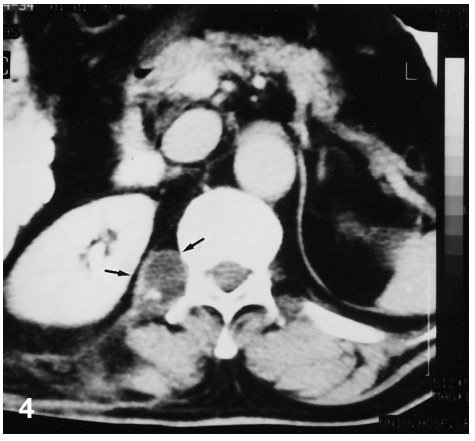


Fig. 2. Spinal hydatid disease. Enhanced CT shows a complex mass lesion, occupying and expanding the left transverse process and pedicle of the O3 vertebra. There is thinning of the cortex with good preservation of its outer margin. Coarse trabeculae are crossing the cystic part of the mass, whereas the soft tissue component is largely calcified. Ventral displacement of the left psoas muscle by the mass is also demonstrated

Fig. 3 a, b. Spinal hydatid disease. **a** Sagittal conventional spin-echo (SE) T1-weighted MR image (TR/TE = 50/17) shows a lesion involving T12 and L1 vertebrae. It has a low signal intensity and extends to the epidural space compressing the spinal cord. No abnormalities of the signal in the adjoining marrow are seen. **b** Axial conventional SE T1-weighted MR image (TR/TE = 50/25) of the same lesion

Fig. 4. Paraspinal hydatid disease. Enhanced CT shows a right paravertebral unilocular cystic mass *(arrows)* without signs of osseous destruction or spinal canal encroachement

lesion shown as a rounded or oval area containing fluid, with sharp and thin margins, exhibiting no contrast enchancement. Alternatively, it may mimic a pattern similar to an abscess or a tumor which is difficult to differentiate from a malignant bone tumor (Figs. 1–4) [2]. Signs meaningful for a differential diagnosis include lack of

Fig. 5. Hydatid disease of bone. Unenhanced CT shows an extensive destruction of the left iliac bone by a homogeneous soft tissue mass which also involves the gluteus medius and iliopsoas muscles

Fig. 6. Hydatid disease of soft tissue. Enhanced CT shows abdominal wall location of a mass in a patient with multiple occurrence of HD. The lesion is heterogeneous with high CT numbers and septae within it

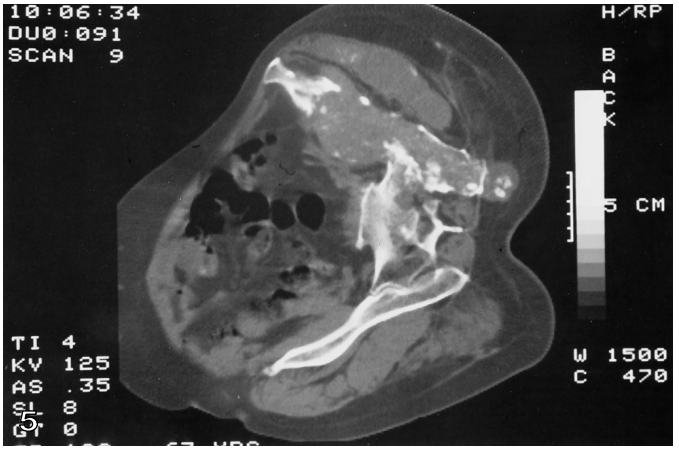
Fig. 7 a, b. Hydatid disease of muscle. **a** Unenhanced CT shows a multivesicular cyst involving the left adductor magnus muscle with multiple low-attenuation daughter cysts. **b** Longitudinal ultrasonography of the same patient shows a large cystic lesion containing multiple, rounded, and anechoic smaller cysts

Fig. 8. Hydatid disease of the kidney. Unenhanced CT shows a large cyst in the right kidney with low-attenuation daughter cysts and mural calcification

Fig. 9. Hydatid disease of the urinary bladder. Enhanced CT shows a large cyst in the urinary bladder with mural thickening, partial calcification, and enhancement corresponding to a hydatid cyst

Fig. 10. Hydatid disease of the spleen. Enhanced CT shows a large splenic hypodense mass with broad mural calcification corresponding to a hydatid cyst

Fig. 11. Hydatid disease of the spleen. Unenhanced CT shows multiple, rounded, hypodense splenic lesions, with mural calcification, corresponding to multiple hydatid cysts



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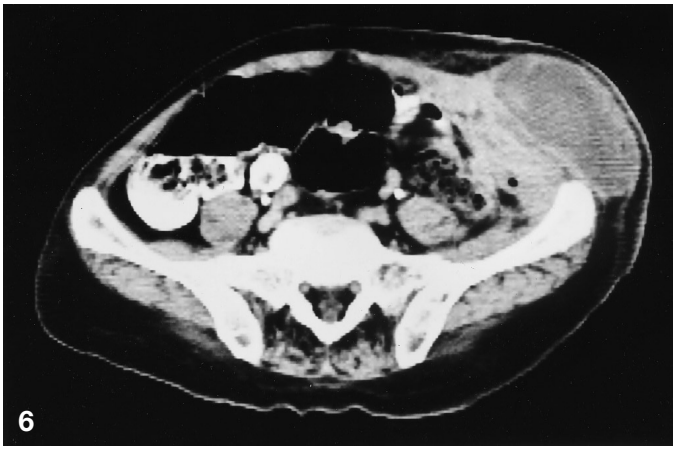
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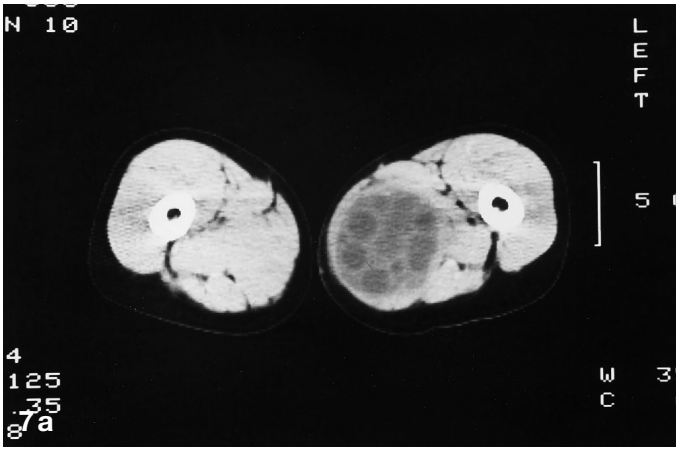

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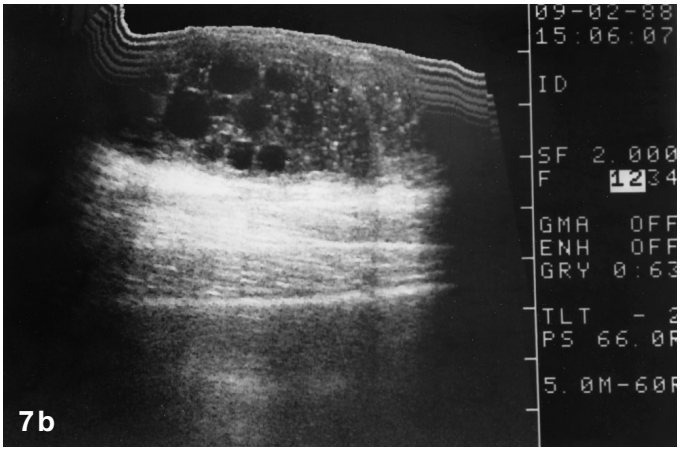

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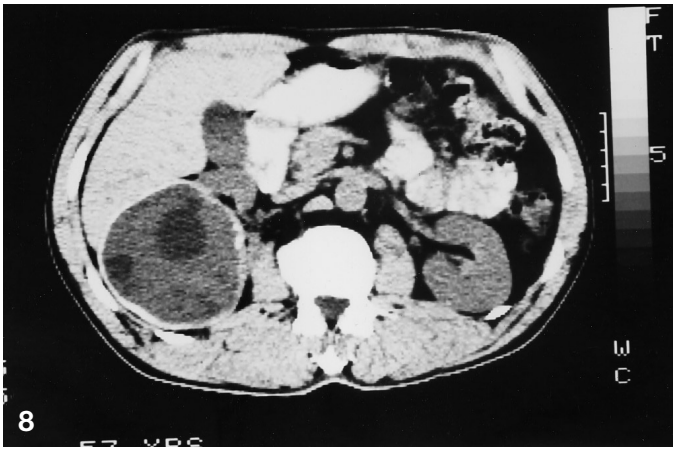

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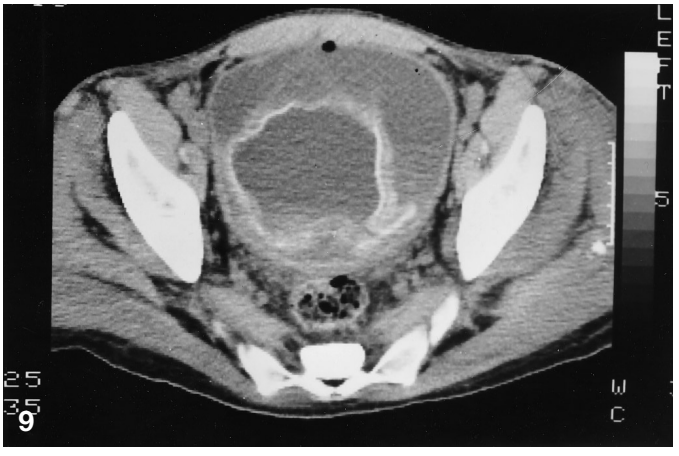

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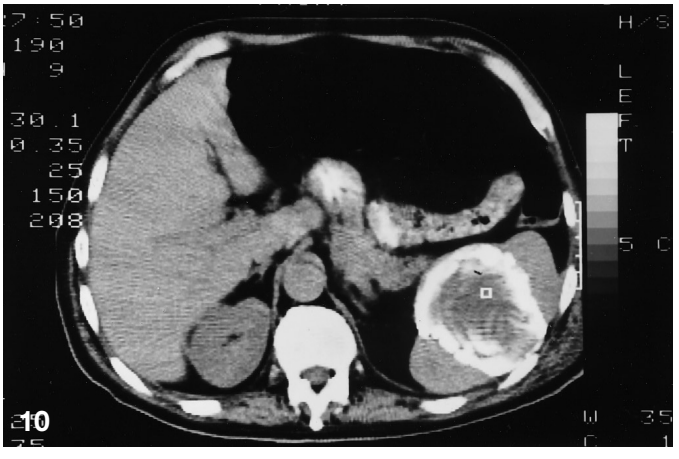

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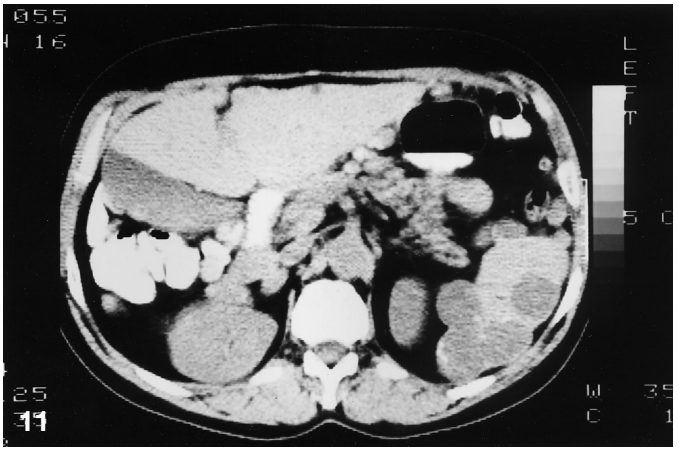

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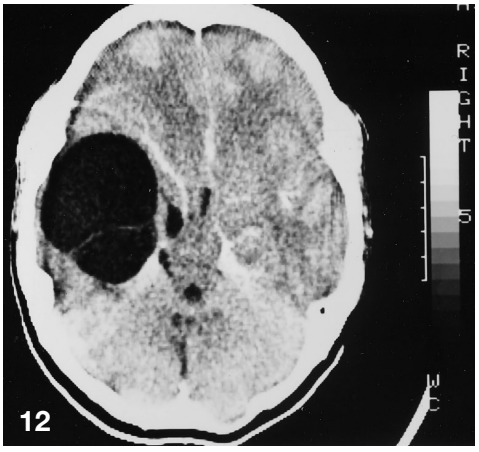
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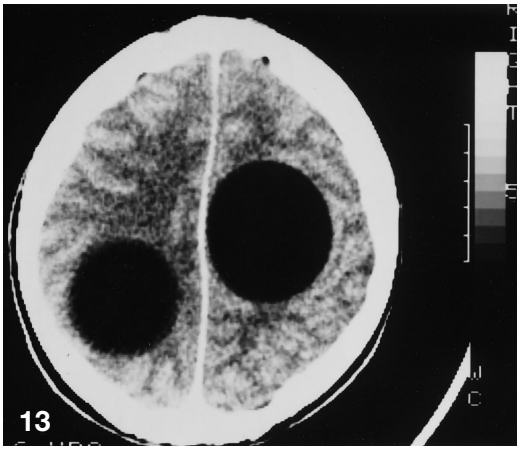
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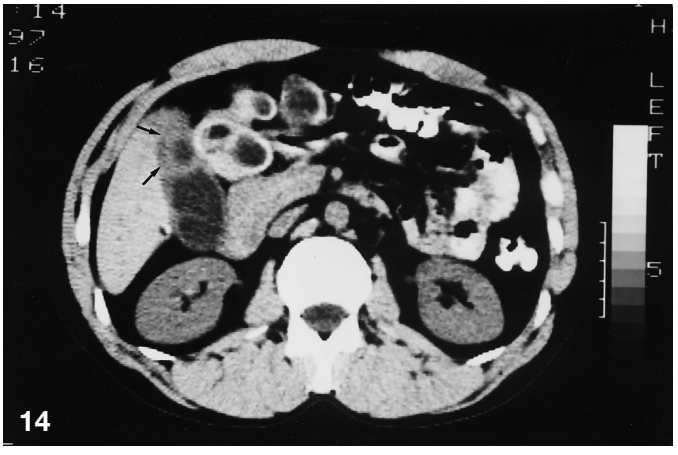
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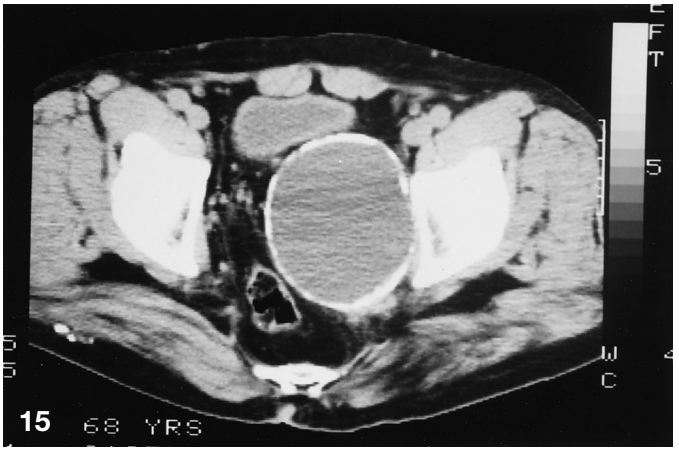
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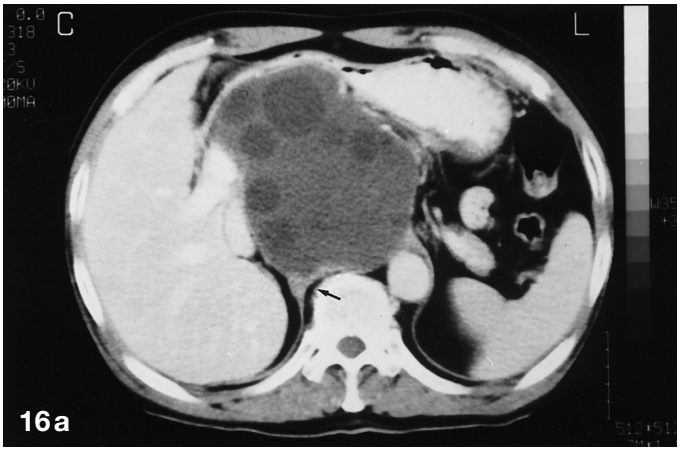
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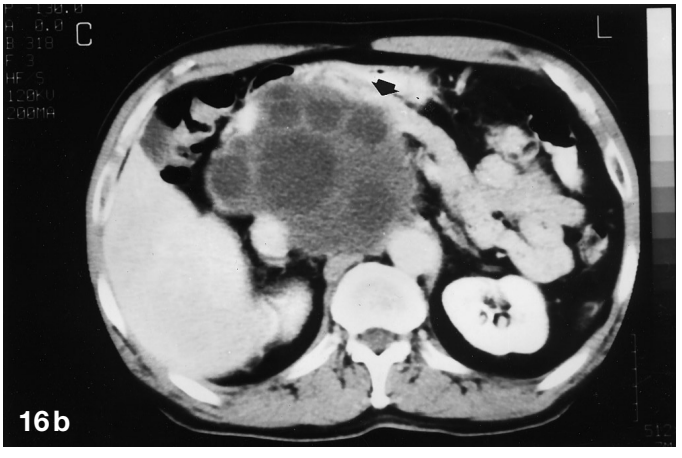
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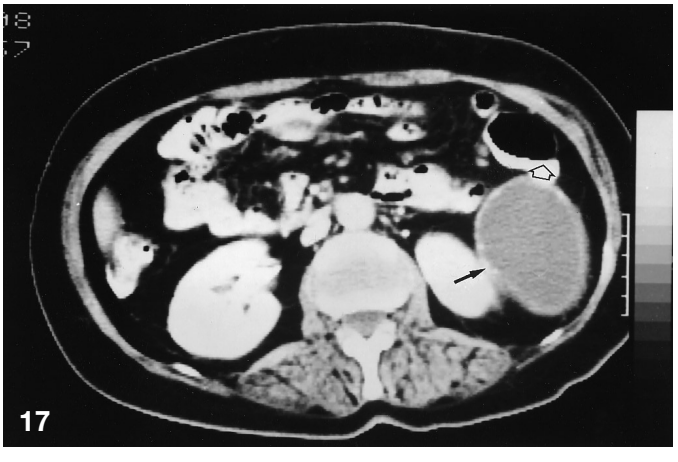
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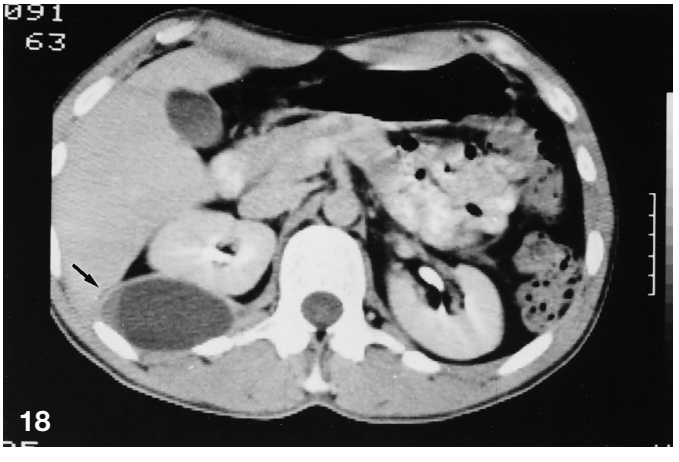
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osteoporosis and bone thickening in the host bone and the presence of intralesional calcifications [2]. The association of bone lesions with soft tissue calcification allows a reliable diagnosis of HD [9].

In spinal HD the most common differential diagnosis problem is tuberculous spondylitis. The absence of damage of the disc surfaces of the vertebral bodies and the spread of the disease through a subperiosteal and subligamentous path are typical of vertebral HD [2].

Hydatid disease presenting in the soft tissues occurs in 0.5–4.7% of patients. A palpable mass is the most constant clinical or exploratory finding of HD affecting soft tissues. The clinical manifestations are caused by compression of the involved organ. The CT findings are variable including the unilocular cyst, the multivesicular lesion, and the atypical complex or solid lesion. The multivesicular lesion is characteristic of HD and presents with multiple daughter cysts within the parent cyst, whereas complex or solid lesion is the result of inflammatory changes mimicking a tumor (Figs. 5–7). In that case MR and US could be useful in the diagnosis [4].

Urinary tract

Renal hydatidosis represents 2–3% of patients. It may be asymptomatic, whereas in the majority of patients clinical presentation includes a flank mass, renal colic, persistent fever, hematuria, dysuria, pyuria, renal stones, or hypertension, symptoms which are by no means specific to reliably establish the diagnosis [5].

The CT findings consist of unilocular or multilocular cysts usually polar with well-defined walls that enhanced with contrast medium. Mural calcification and the presence of daughter cysts, which are lower in density, within the large parent cyst, often coexist [10, 11]. The presence of these two findings help in differentiating the lesion from simple renal cyst, renal abscess, infected cyst, and necrotic neoplasm (Fig. 8) [5].

Urinary bladder hydatidosis is extremely rare and we are not aware of any previous radiological description of such an involvement. It appears as a large unilocular lesion with mural thickening and calcification, enhancement in the contrast medium, and no obvious septae or daughter cysts within it. The disease had a multiple location in other organs, thus making the diagnosis easier (Fig. 9).

Spleen

Primary splenic involvement by HD is rare and accounts for less than 2% of patients. Secondary HD of the spleen may be due to rupture of abdominal cysts with dissemination to the spleen. The clinical manifestations, which are not specific, include abdominal pain, enlarged spleen, and fever.

On CT the cysts usually exhibit a homogeneous fluid content showing water attenuation values, but the presence of intracystic debris, hydatid sand, and inflammatory cells are presumed to cause high CT values. Mural calcification may occur and multiple daughter cysts within a large cyst may also be present (Figs. 10, 11).

Differential diagnosis of splenic HD includes epidermoid cyst, pseudocyts, solitary abscess or hematoma, intrasplenic pancreatic pseudocyst, and cystic neoplasm of the spleen. The diagnosis is easier when the lesion has multiple location involving other organs or when daughter cysts are present [3, 12].

Brain

Cerebral HD represents only 2% of all cerebral space-occupying lesions even in the countries where the disease is endemic. It is seven times more common in children than in adults. The clinical manifestations include headache, vomiting, papilledema, or diminution of vision. It is more frequently supratentorial, involving the middle cerebral arterial territory. The parietal lobe is the most common site of HD development.

The typical CT appearance is a large, well-defined, spherical or oval, nonenhanced, unilocular intraparenchymal lesion containing fluid similar in density to cerebrospinal fluid (Figs. 12, 13). There is usually neither calcification (1%) nor perifocal edema, although there is considerable mass effect and there may be hydrocephalus [13–17].

Lesions to be differentiated from hydatid cyst include arachnoid cyst, porencephalic cyst, cystic tumor, and abscess. The arachnoid cyst is not round or surrounded by brain tissue, as the HD while the poren-

Fig. 12. Cerebral hydatid disease. Unenhanced CT shows a large, well-defined, unilocular cystic mass in the right hemisphere with septae and without perifocal edema corresponding to a hydatid cyst

Fig. 13. Cerebral hydatid disease. Enhanced CT shows multiple, large, and rounded cysts in the hemispheres, without enhancement and with diffuse edema, corresponding to hydatid cysts

Fig. 14. Hydatid disease of the gallbladder. Unenhanced CT shows a small cyst with mural thickening located within the gallbladder *(arrows)*. Some other calcified cysts in the peritoneum coexist

Fig. 15. Hydatid disease of the pelvis. Enhanced CT shows a large cyst with mural calcification and without enhancement located on the left of the minor pelvis. Because of its size, the urinary bladder and rectum are compressed

Fig. 16 a, b. Hydatid disease of the retroperitoneum. **a** Enhanced CT shows a lobulated retroperitoneal cystic mass containing multiple low-attenuation daughter cysts. Because of its size and location, inferior vena cava *(arrow)* is displaced ventrally and to the right. **b** Enhanced CT of the same patient shows ventral displacement of the pancreas *(arrow)* by the mass

Fig. 17. Hydatid disease of the retroperitoneum. Enhanced CT shows a large cyst with mural thickening and calcification, without enhancement located in the left anterior pararenal compartment *(arrow)*. Because of its size, the descending colon is displaced ventrally (*open arrow*)

Fig. 18. Hydatid disease of the retroperitoneum. Enhanced CT shows a cystic lesion, without enhancement, located in the right posterior pararenal compartment and compressing the kidney *(arrow)*

cephalic cyst usually communicates with the ventricle. Cystic tumor usually has a soft tissue component which enhances after intravenous contrast, whereas an abscess usually shows contrast enhancement and perifocal edema.

Peritoneal and retroperitoneal hydatidosis

Peritoneal hydatidosis, although rare, can be situated anywhere and is usually the result of traumatic or surgical rupture of a hepatic, splenic, or mesenteric cyst. The frequency of a liver HD rupturing within the peritoneum is reported in the range of 7%, whereas that of HD rupturing in the biliary tract is 9%.

The cysts may cause abdominal distension or obstruction (Figs. 14, 15). Primary hydatidosis in the retroperitoneum is very rare and can be considered primary only when no other cysts of the same etiology are present (Figs. 16–18) [18]. Mural calcification, which is the hallmark of the disease, and the cystic nature of the lesion can be reliably detected by CT.

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