

K. Abbassioun MD<sup>1</sup>.  
A. Amirjamshidi MD<sup>2</sup>.  
M. Sabouri Deylamie MD<sup>3</sup>.

## Hydatid Disease of the Central Nervous System Imaging Characteristics and General Features

**Background;** Hydatid disease primarily affects the liver and typically demonstrates characteristic imaging findings. Secondary involvement due to hematogenous dissemination may be seen in almost any locations, *e.g.*, lung, kidney, spleen, bone and central nervous system (CNS).

**Objectives:** To review the different aspects of hydatidosis of the CNS briefly and discuss the pathognomonic features and rare varieties of radiological findings useful in preoperative diagnosis of the disease in the human CNS.

**Material & Method;** In a retrospective study, the records of almost 100 cases of CNS hydatidosis were analyzed. The available images were reviewed by independent observers, either a radiologist or a neurosurgeon, and reported separately.

**Results;** In skull X-ray films, nonspecific changes denoted increased intracranial pressure, skull asymmetry and curvilinear calcification in rare instances. Computed tomography (CT) and magnetic resonance imaging (MRI) demonstrated the round or oval, well-defined cystic mass with an attenuation or signal intensity similar to that of cerebrospinal fluid, with no associated perifocal edema, and no contrast enhancement as the pathognomonic findings of brain hydatidosis. Similar findings were detected in hydatid cysts involving the orbit, spinal column and spinal cord with some variations. Such findings as mild perifocal edema, non-homogenous contrast enhancement, non-uniform shapes, calcification and multiplicity or septations have been the atypical radiological findings.

**Conclusion;** In endemic areas, familiarity with typical and atypical radiological manifestations of hydatid disease of the CNS, will be helpful in making prompt and correct preoperative diagnosis leading to a better surgical outcome.

**Keywords:** Brain, Computed Tomography, Echinococcosis, Orbit, Spine, Magnetic Resonance Imaging.

### Introduction

Hydatid disease is a worldwide zoonosis caused by the larval stage of the *Echinococcus* tapeworm. The two main types of hydatid disease are caused by *E. granulosus* and *E. multilocularis*. The former is commonly encountered in the great grazing regions of the world particularly the Mediterranean regions, Africa, South America, the Middle East, Australia and New Zealand. It is the most common type of hydatid disease in human. The classical findings in hydatid disease are well-known, however, findings related to disease complications and unusual anatomic locations and rare imaging presentations are less frequently described in the literature. In this manuscript, it has been tried to present the general features of hydatidosis of the CNS briefly and demonstrate a variety of radiological findings important in the preoperative differential diagnosis of such lesions.<sup>9</sup>

### Life Cycle

The life cycle of the *E. granulosus* involves two hosts. The definitive host may be any carnivores but usually is a dog and the adult worm of the parasite lives in the proximal small intestine of this animal and attaches by its hooklets to the

#### From:

1. Professor of Neurosurgery, Tehran University of Medical Sciences, Arad General Hospital, Somaye Ave. Tehran, IRAN.

2. Associate Professor of Neurosurgery, Tehran University of Medical Sciences, Sina Hospital, Imam Ave. Tehran, IRAN.

3. Associate Professor of Radiology, Shahid Beheshti University of Medical Sciences, Mehr General Hospital, Tehran, IRAN.

#### Corresponding Author:

Dr Amirjamshidi, Sina Hospital, Imam Ave. Tehran, IRAN.

Tel:

+9821 670 1041

intestinal mucosa. Eggs are released into the host's intestine and excreted with the feces to be ingested by the intermediate hosts such as sheep and cow. The ovum loses its protective chitinous layer as it is digested in the duodenum and the released hexacanth embryo or oncosphere passes through the intestinal wall into the portal circulation and grows into a cyst within the liver. The cycle will be completed when a definitive host such as dog, eats the viscera of the intermediate host. The human can also be the intermediate host as well through contact with a definitive host such as a domesticated dog or ingestion of contaminated water or vegetables. When the parasite passes through the intestinal wall to reach the portal venous or lymphatic systems, the liver acts as the first line of defense and therefore, it will be the most frequently involved organ.<sup>16</sup> In human, hydatid disease involves the liver in approximately 75% of the cases, the lung in 15%, brain in 2%, vertebrae in 1%, and orbit in 0.2% to 0.8%.<sup>1,7,9,10,31</sup> Once in the human liver, cysts grow to a mass 1 cm in diameter during the first six months and grows 2-3 cm annually thereafter, depending upon the tissue resistance.<sup>36</sup> In the brain, it takes 5 to 16 months for the cyst to grow to a mass 1 cm in diameter.<sup>1,9</sup> Compressible organs such as lung or brain facilitate growth of the cyst and it has been shown that brain hydatid cyst grows faster than that of liver by a ratio of 3:1.<sup>1,20</sup>

### Structure of the Hydatid Cyst

The hydatid cyst has three layers: A) the outer pericyst, composed of modified host cells. This layer is a dense fibrous zone in the liver and other organs. In the brain, however, there is only a shiny fine gliotic layer without any remarkable resistance surrounding the primary un-ruptured cysts. B) the next layer is the middle laminated membrane which is acellular and allows the passage of nutrients, and C) the inner germinal layer where the scolices (the larval stage of the parasite) and the laminated membrane are produced. The middle laminated membrane and the germinal layer form the true wall of the cyst usually referred to as the endocyst. Daughter cysts are small spheres containing the protoscolices. The inner-most layer which is the germinative or fertile membrane performs two major functions; 1) production of the laminated membrane outward, and 2) germination of the later generation scolices inward. The formation of scolices takes place from the brood capsules attached to the germinative membrane. The pedicle of the brood cysts is very loosely attached to the germinal layer of the mother cyst. On gross examination, the vesicles resemble a bunch of grapes and may grow through the wall of the mother cyst.<sup>9,16</sup> Cyst fluid is

clear or pale yellow, has a neutral pH, and contains sodium chloride, proteins, glucose, ions and polysaccharides and is antigenic. When brood cysts and vesicles detach within the cyst, and pass into the cyst fluid, they form a white sediment known as "hydatid sand."<sup>5,9</sup>

### Central Nervous System Involvement

Pediatric population consists of about 70% to 80% of CNS hydatidosis.<sup>1,20,35,37</sup> Hematogenous spread is the most common route of dissemination of the ova to the CNS. Most cysts are acquired during childhood. Spinal hydatid disease is the most common form of the bone involvement.<sup>2,21</sup> Although hydatid disease may be located anywhere in the brain, it is most frequently located in the hemispheres (90% to 95% of the cases), particularly in the territory of the middle cerebral artery mostly in the postrolandic parts of the hemispheres.<sup>1,11</sup> The orbit,<sup>10,24</sup> hypothalamus,<sup>19</sup> pons,<sup>3,29</sup> the subarachnoid space of cerebellopontine angle,<sup>29</sup> ventricular system,<sup>1,9</sup> aqueduct of Sylvius,<sup>1</sup> and intracranial epidural space<sup>38</sup> comprise the rare locations for growth of hydatid cyst. In this article, it is not intended to discuss the clinical presentation or management of hydatidosis of the CNS. It is meant to focused only on the imaging studies and the tips for differential diagnosis of CNS echinococcosis.

### Imaging of the CNS Hydatidosis

The skull X-ray may demonstrate unilateral enlargement of the vault, suture separation (Figure 1) or erosion of the posterior clinoid process and calcifications.<sup>8,9,17,33,36</sup>

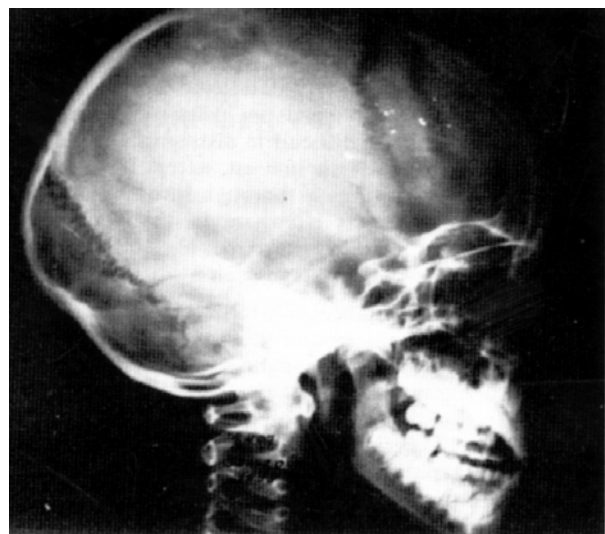
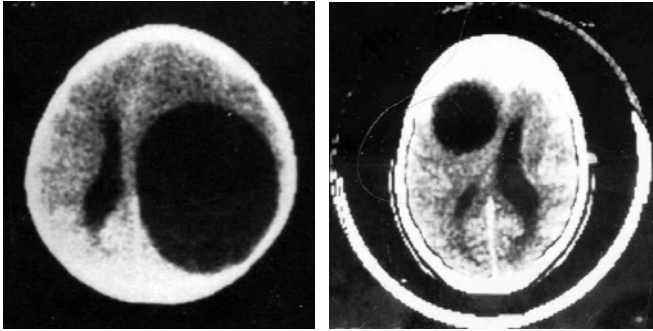
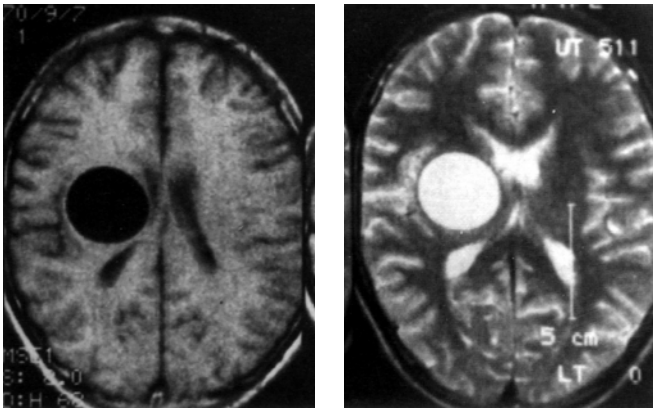


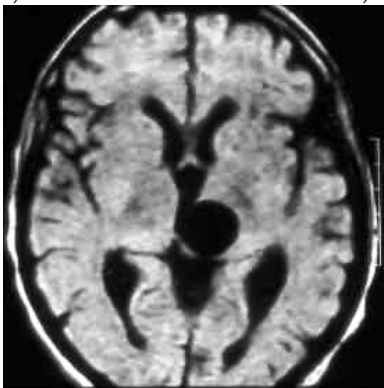
Figure 1: Plain skull X-ray demonstrating the asymmetric skull expansion with suture departure and bone erosion in the left frontal region



**Figure 2:** Contrast enhanced CT scan showing a large oval-shaped low density lesion involving most of one hemisphere causing midline shift and contra-lateral ventricular dilatation.



**Figure 3:** T1 low intense, T2 hyperintense round cystic lesions without peripheral edema



c)



d)

CT (Figure 2) and MRI (Figure 3 a to d), both demonstrate a well-defined oval or cystic mass with an attenuation or a signal intensity similar to that of cerebrospinal fluid. The lesions do not enhance after injection of contrast medium.

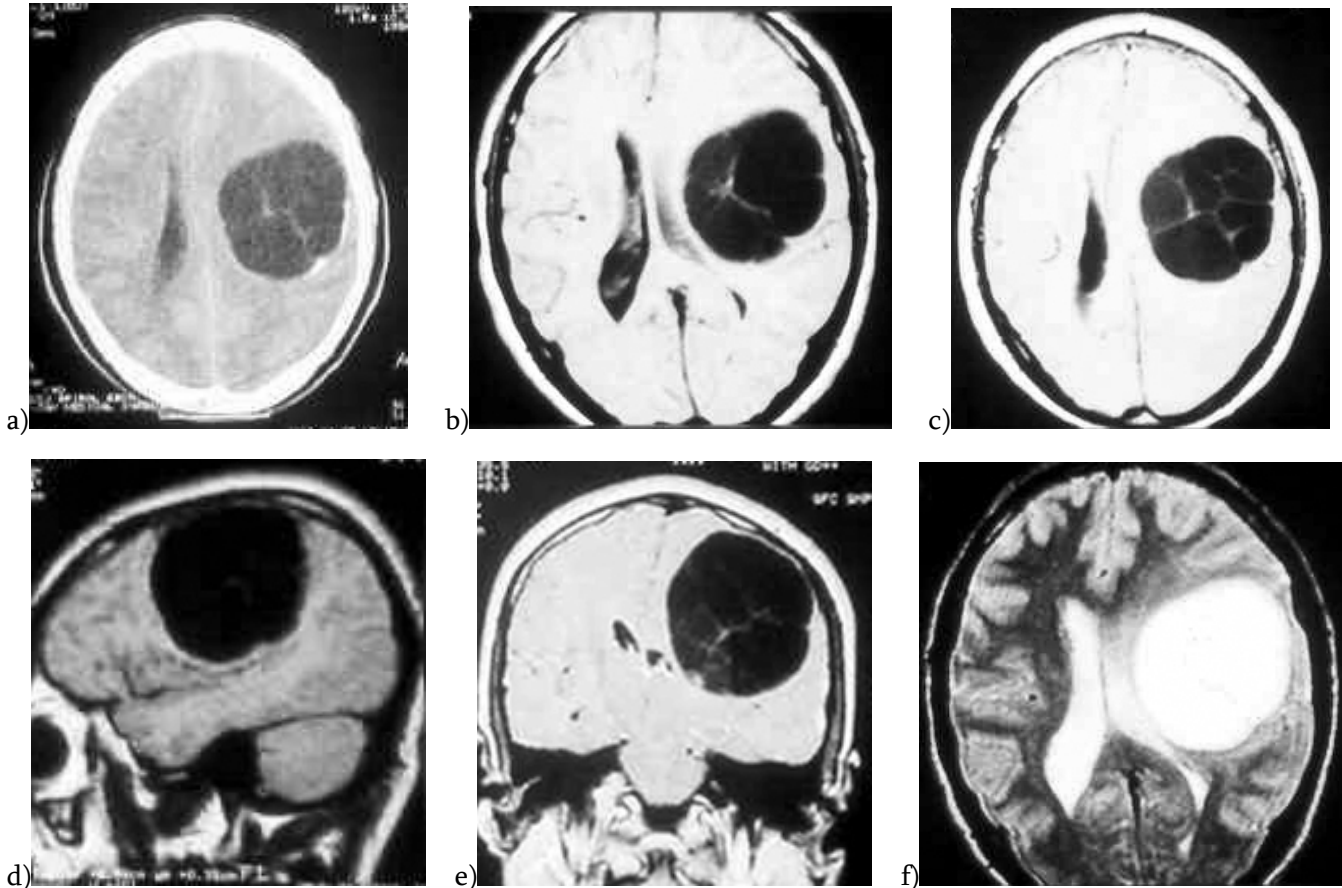
Although the lesion may cause extrinsic compression on the ventricular system with subsequent development of hydrocephalus, there is no associated pericystic edema as is typically observed in abscesses and cystic tumors on rare occasions.<sup>4,9,18,25,39,41</sup> The rim of these cystic lesions may occasionally enhance after injection of contrast medium and may show perifocal edema.<sup>1,7,23,37,39,42</sup>

The cysts are usually single. However, there may be multiple cysts primarily lodged within the CNS or multiple cysts secondarily developed after rupture of a primary or mother cyst.<sup>6,32</sup> The multiple cysts appear as small and large multilocular cystic lesions on CT and MRI (Figure 4). The cysts are ovoid or polygonal resembling a bunch of grapes. There may be perifocal edema or even a fine enhancement after injection of the contrast medium, both on CT and MRI.<sup>1,14,15,23,29,32,42</sup> The cyst may be hyperdense on un-enhanced CT (Figure 5) with a calcified ring surrounding the cyst cavity,<sup>55</sup> or it may be septated.<sup>41,42</sup> Partial or complete calcification and internal partitioning are other rarely described features of hydatid cysts of the CNS, anecdotally reported in the literature as CT or MRI findings.<sup>8</sup>

Calvarium is another rare site of involvement by hydatid disease.<sup>1,22</sup> It grows as an expansile intradiploic lesion (Figure 6) and should be differentiated from all other cystic intradiploic pathologies such as cysticercosis and leptomeningeal cysts. CT using bone window density and MRI, both can be complementary studies for differentiation of other possible intra-diploic lesions.<sup>1,40</sup>

### Orbit

A young patient from rural areas, presenting with painless unilateral exophthalmos. CT and/or MRI showing single or multiple homogenous retrobulbar masses with regular contour and fine rim enhancement after injection of contrast material (Figure 7) can be characteristic of orbital hydatidosis. Destruction of the wall of the orbit visible both on plain skull X-ray and CT is another finding compatible with gradual and slow growth of the lesion.<sup>1,10,31,38</sup>



**Figure 4:** a) CT scan with contrast demonstrating multiloculated hydatid cyst with mild enhancement of the cyst wall., b-f ) MRI T1, low intense and T2, hyperintense multiloculated hydatid cyst.

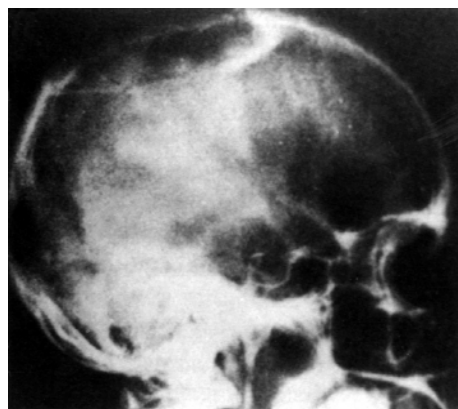
### Structure of the Spinal Hydatid Cyst

Spinal hydatid disease is the most common form of bone involvement accounting for more than 50% of cases of hydatid disease of the bone.<sup>1</sup> Some imaging characteristics are typical for spinal hydatid disease. These include lack of osteoporosis and sclerosis in the

host bone, absence of damage to the disc space and spread of the disease either via hematogenous route to the more vascular areas of the vertebra, or via a subperiosteal and subligamentous path to the paraspinal region that results in involving a contiguous rib and destructing the adjacent costo-chondral junction.<sup>12,30</sup>



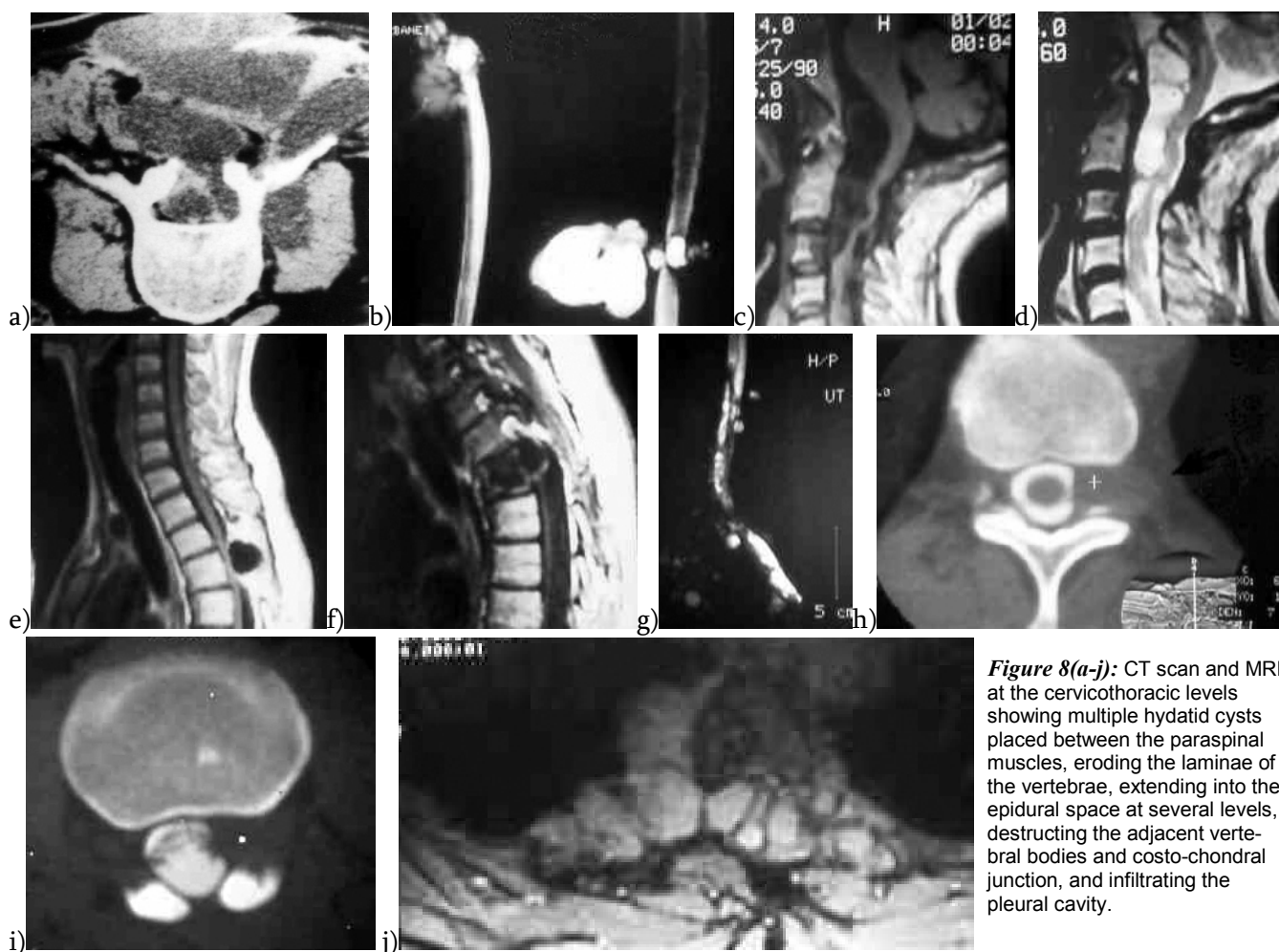
**Figure 5:** CT scan demonstrating a rather completely calcified hydatid cyst.



**Figure 6:** Plain skull X-ray showing expansion of the parietal parasagittal bone due to an intradiploic hydatid cyst



**Figure 7:** CT scan showing a retroglobally located single hydatid cyst with enhancing wall after injection of contrast medium.



**Figure 8(a-j):** CT scan and MRI at the cervicothoracic levels showing multiple hydatid cysts placed between the paraspinal muscles, eroding the laminae of the vertebrae, extending into the epidural space at several levels, destructing the adjacent vertebral bodies and costo-chondral junction, and infiltrating the pleural cavity.

CT scan allows a precise assessment of osseous lesions and can clearly depict hydatid cysts or the microvesicular polycystic vertebra as observed in the last stage of involvement of the vertebral body—before the affected vertebra collapse. MR imaging, however, is superior to CT since it can demonstrate involvement of the neural structures.<sup>12,28,30</sup> The X-ray findings are mostly nonspecific whereas, CT and MR findings are almost characteristic, showing the cysts which are usually in an alveolar and/or a diffuse form typically compressing the spinal cord posteriolaterally or posterolaterally.<sup>27,30</sup> Pure spinal cord hydatid cysts can be extradural, subdural, subarachnoidal or intramedullary.<sup>12,28</sup> MRI (Fig 8) is the diagnostic tool of choice for depicting such lesions.<sup>21,33,34</sup>

## Discussion

Even though CT and MRI findings of hydatidosis can be pathognomonic, the physicians practicing in endemic areas must be familiar with specific clinical and paraclinical features of the illness and look for the systemic involvement of the disease while referring the patients to a neurosurgical department. Considering liver as the most common organ in-

involved by hydatid cysts, abdominal CT scan is the diagnostic tool of choice with a higher rate of accuracy than ultrasonography for detecting the visceral cysts.<sup>36</sup>

In the brain, differentiation of the suspected lesions from arachnoid cysts, brain abscesses and cystic brain tumors are of major importance. Absence of perilesional edema and rim enhancement are valuable findings on CT and MRI, nevertheless, clinical findings such as a young age at presentation, asymmetrical head configuration, signs of chronically increased intracranial pressure, epilepsy and recent appearance of neurological deficit are important clues tending the history of the illness towards a more benign acquired illness.<sup>1,7,9,13,35</sup> Considering the reports about the presence of rim enhancement and pericystic edema in cerebral hydatidosis, possibly due to the leakage of the cyst material through the cyst wall that induces reaction of the surrounding glial tissue, some difficulty and pitfalls may happen in preoperative differential diagnosis of such cystic lesions if the diagnosis is made solely based on the imaging findings.<sup>1,36,37</sup> Proton magnetic resonance spectroscopy (MRS) has been performed in some cases of intracranial hydatidosis. Besides lactate, alanine, and acetate,

a large resonance of pyruvate was observed in these cases. These findings were further confirmed by *ex-vivo* high-resolution NMR spectroscopy of the evacuated cyst fluid, as well as the fluid aspirated from a cyst in the liver of the same patient. The MRS, therefore, may be used as an adjunct to imaging for the diagnosis of intracranial hydatid cysts. It may also be used for monitoring the response to drug therapy.<sup>26</sup>

## Conclusion

The typical appearance of hydatid cyst of the CNS is a large non-enhancing cyst of CSF density in CT and MRI with no pericystic edema and no rim enhancement after injection of the contrast medium. There may be however, occasional atypical appearances worthy to be aware of. These include irregularity of the cyst wall contour, enhancement of the surrounding rim, isodensity or heterogeneity of the cyst content, surrounding brain edema and globular or curvilinear calcification. Awareness of such atypical findings on CT or MRI can prevent misdiagnosis and inappropriate premature puncture of the lesion during surgery that may result in subsequent catastrophe.

Of clinical importance is that a great liver hydatid cyst may be asymptomatic and discovered incidentally by sonography or CT scanning. The majority of them will undergo spontaneous abortion and do not need any active intervention. This is not the case for the hydatid cyst of the CNS. Because of the unpredictable growth pattern of CNS cysts, until a proper medical therapy is discovered, they must be surgically removed at almost any stage they are diagnosed.

## References

- Abbassioun K. Amirjamshidi A. Diagnosis and management of Hydatid cyst of the central nervous system. Part 1. *General considerations and Hydatid disease of the brain. Neurosurgery Quarterly*, 2001; 11:1-9.
- Abbassioun K. Amirjamshidi A. Diagnosis and management of Hydatid cyst of the central nervous system. Part 2. *Hydatid cysts of the skull, orbit and spine. Neurosurgery Quarterly*, 2001; 11:10-16.
- Abbassioun K. Amirjamshidi A. Moinipoor MT. Hydatid cyst of the pons. *Surg. Neurol.* 1984; 26:297-300.
- Abbassioun K. Rahmat H. Ameli NO. Tafazoli M. Computed tomography in hydatid cyst of the brain. *J. Neurosurgery*, 1978; 94: 408-11.
- Abbassioun K. Sabouri-Deylami M. Rahmat H. Diagnostic preoperative des kystes hydatiques cerebraux par la tomodynamometrie et son importance pronostique. *Neurochirurgie*, 1982; 28: 21-24.
- Abdulla K. Tapoo AK. Agha HS. Ruptured cerebral Hydatid cyst. Case report. *J. Tropical Med. Hyg.* 1988; 91:302-305.
- Altinrs N. Bavbek. Caner HH. et al. Central nervous system hydatidosis in Turkey. A cooperative study and literature analysis of 458 cases. *J. Neurosurgery*, 2000; 93:1-8.
- Alvarez F. Blazquez MG. Oliver B. et al. Calcified Hydatid cyst. *Surg. Neurol.* 1960;17:163-4.
- Ameli NO. Abbassioun K. *Hydatid disease of the central nervous system.* Kitab Sara Co. 1995, Tehran, Iran. Chap. 2-6, pp. 21-141.
- Ba'assiri A, Jabi A. Computed tomography in orbital echinococcosis. *Brit. J. Radiol.* 1984, 57:270-272.
- Begg NC, Begg AC, Robinson RG. Primary Hydatid disease of the brain. It's diagnosis, radiological investigations, treatment and prevention. *NZ. Med. J* 1957; 56:94-98.
- Berk C, Ciftci E. Erdogan A. MRI in primary intraspinal extradural Hydatid disease; a case report. *Neuroradiology* 1998; 40:390-2.
- Bonakdarpour A. Echinococcus disease. Report of 112 cases from Iran and a review of 611 cases from the United States. *AJR* 1967; 99:660-7.
- Coates R. MR imaging of cerebral Hydatid cysts. *AJNR* 1990; 11:1249-50.
- Chang KH. Hann MH. MRI of CNS parasitic diseases. *J. Magn. Reson. Imaging* 1998; 8:297-307.
- Crag & Faust's *Clinical parasitology*. 8th.ed.1970. Lea & Feibieger, Holland.
- Demir K. Karsli AF. Kaya T. et al. Cerebral Hydatid cysts. CT findings. *Neuroradiology* 1991; 33: 22-24.
- Draouat S. Filali N. Namous H. et al. X-ray computed tomography in cerebral Hydatid disease in children. *J. Radiol.* 1986; 67:179-184.
- Erashin Y. Mutluer S. Dermirts E. et al. A case of thalamic Hydatid cyst. *Clin. Neurol. Neurosurg* 1995; 97: 321-3.
- Ersahin Y. Mutluer S. Guzelbag E. Intracranial Hydatid cysts in children. *Neurosurgery* 1993; 33: 219-224.
- Ferris BD, Scott JE, Utterley D. Hydatid disease of the cervical spine. *Clin. Orthop.* 1986; 207:174-177.
- Gokalp HZ, Kanpolat Y, Gokben B. Hydatid cyst of the skull. *Neurosurg.* 1979; 4: 431-433.
- Gonzalez-Ruiz CA. Isla A. Perez-Higueras A. et al. Unusual CT image of a cerebral Hydatid cyst. *Ped. Radiol.* 1990; 20: 283-4.
- Hamza R. Tonibi S. Jamoussi M. et al. Intracranial and orbital Hydatid cysts. *Neuroradiology* 1982; 22: 211-4.
- Karak PK. Mittal M. Bhatia S. et al. Isolated cerebral Hydatid cyst with pathognomonic CT sign. *Neuroradiology* 1992; 34: 9-10.
- Kohli A. Gupta RK. Poptani H. In vivo proton magnetic resonance in a case of intracranial Hydatid cyst. *Neurology* 1995; 45: 562-4.
- Law WB. Extradural spinal hydatid cyst. *Aust. New Zeal. J. Surg.* 1965; 34:215-217.
- Ley AJ, Marti A. Intramedullary hydatid cyst. *J. Neurosurg.* 1970; 33:457-459.
- Mascalchi M. Ragazzoni A. Dal Pozzo G. Pontine Hydatid cyst in association with an acoustic neurinoma. MR appearance in an unusual case. *AJNR* 1991; 12: 78-9.
- Mikhael MA, CiricIJ, Taskington JA. MR imaging in spinal echinococcosis. *J. Comp. Assist. Tom.* 1985; 9:308-400.
- Morales AG, Croxatto JO, Croveto L, Ebenes R. Hydatid cysts of the orbit. (A review of 35 cases). *Ophthalmology*, 1988; 95:1027-1032.
- Nurchi G. Floris F. Montaldo C. et al. Multiple cerebral Hydatid disease: Case report with magnetic resonance imaging study. *Neurosurgery* 1992; 30: 436-8.
- Ozer AF. Ozek MM. Pamir MN. et al. Magnetic resonance imaging in diagnosis of spinal Hydatid cyst disease. *Paraplegia* 1993; 31: 338-40.
- Pau A, Simonetti G, Tortori-Donati P, Turtas S, Viale GL. Computed tomography and magnetic resonance imaging in spinal hydatidosis. *Surg. Neurol.* 1987; 27:365-369.
- Pearl M. Kosilimbos DG. Lehrer HZ, et al. Cerebral Echinococcosis, a pediatric disease. Report of 2 cases with one successful five year survival. *Pediatrics* 1987; 61: 915-920.
- Pedrosa I. Saiz A. Arrazola J. et al. Hydatid disease; Radiologic and pathologic features and complications. *Radiographics* 2000; 20:795-817.
- Peter JC. Domingo Z. Sinclair-Smith C. et al. Hydatid infestation of the brain. Difficulties with computed tomography diagnosis and surgical treatment. *Pediat. Neurosurg* 1994; 20: 78-83.
- Pirouz MS. Hydatid disease of the orbit 7cases. *Orbit* 1983; 2: 65-68.

39. Rudwan MA, Khaffaji S. CT of cerebral Hydatid disease. *Neuroradiology* 1988; 30: 498-9.
40. Teymourian GA, Bagheri F. Hydatid cyst of the skull. Report of four cases. *Radiology* 1976;118:97-110.
41. Tuzun M, Hekimoglu B. Hydatid disease of the CNS. Imaging features. *AJR* 1998;171:1497-1500.
42. Wada H, Takahashi S, Higano S, et al. Ct and MRI findings in a case of alveolar echinococcosis masquerading as brain tumor. *Nippon Igaku Hoshasen Zasshi* (English abstract) 1996; 56:608-9.