

## ORIGINAL ARTICLE

## PRIMARY INTRACRANIAL AND SPINAL HYDATIDOSIS: CT AND MR IMAGING FINDINGS WITH PATHOLOGICAL CORRELATION

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## ABSTRACT

**Background:** Primary cerebral and spinal hydatidosis are very rare accounting for 2 and 1 percent of hydatidosis respectively. The aim of this study is to describe the MRI and CT imaging features and the pathological correlates of primary cerebral and spinal hydatidosis and discuss the differential diagnosis.

**Materials and Methods:** The MRI and CT images of assorted seven cerebral and three spinal primary hydatidosis who were operated on in teaching and tertiary referral hospitals between 2006 and 2016 and had histological confirmation were analyzed after exclusion of primary visceral hydatidosis using chest x-ray and abdominal ultrasound.

**Results:** There were seven cerebral and three spinal primary hydatidosis. Their median age was 11.5 years with female to male ratio of 1.5:1. Cerebral hydatidosis presented with focal neurological deficits, seizure and blurred vision and the spinal cases presented with paraparesis/incontinence. The cerebral hydatids were intra-axial with an average size of 8 cm and located in the temporoparietal and parietooccipital lobes, and of the three spinal hydatids, two were located in the thoracic and one in the sacral region. The latter cases involved the spinal canal and paravertebral regions compressing the spinal cord and the dura and destroying the vertebrae.

**Conclusion:** Primary central nervous system (CNS) hydatidosis is being observed in a relatively increased frequency in the Ethiopian setting after the recent introduction of MRI and CT imaging in teaching tertiary referral hospitals. Therefore, awareness should be raised in the differential diagnosis of cystic CNS mass lesion especially in pediatric and young adult patients from rural Ethiopia to improve treatment outcomes.

**Key words:** CNS hydatidosis, CT/MRI imaging

## INTRODUCTION

Human echinococcosis is caused by infection with larval forms (metacestodes) of echinococcus tape worms found in small intestine of carnivores, and is considered one of the most geographically widespread zoonoses in the world. It remains highly endemic in pastoral communities, particularly in Africa, Middle East, Asia and Latin America, and is rare in Western Europe and North America (1-4). It is not uncommon and endemic in Ethiopia, causing significant clinical and surgical morbidity and mortality as reported by different authors (5-8). The major organs to be affected by hydatidosis are the lungs (7-43%) and the liver (50-70%), whereas hydatid cysts of the brain and spine are rare and account for approximately 2% and 1% of all echinococcosis infections, respectively (9). Most of the central nervous system (CNS) hydatidosis are primary. Secondary hydatidosis is extremely rare, occurs as a result of ruptured visceral hydatidosis with hematogeneous spread, and is multiple in contrast to solitary primary

hydatidosis (10). Although spine and brain hydatidosis brief case reports were made in the past (10, 11), in this study we report a study of assorted cases over a 10-year period. This includes review of the CT and MRI findings of operated and histopathologically confirmed primary hydatidosis involving the brain and the spine in tertiary referral and teaching hospitals.

## PATIENTS AND METHODS

The MRI and /or CT findings of 10 patients who were operated at Tikur Anbessa and Zewditu Hospitals, teaching and tertiary referral hospitals, for assorted primary hydatidosis cases from 2006 to 2016. The cases were retrieved from the neuroradiology and neurosurgical, and neuroradiology, neurosurgery and neuropathology joint sessions held every week and every fortnight, respectively. Clinical data, radiological investigations, treatments, treatment outcomes and pathology results were retrospectively analyzed. All cases had undergone preoperative neu-

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rological and neuroimaging evaluation; six patients had MRI, 5 patients had CT examinations and one patient had both CT and MRI. Postcontrast studies were performed when there was perilesional edema, and cystic tumors were suspected. Surgical specimens were obtained for each case for histopathological confirmation. Preoperative abdominal ultrasonography and chest radiographs were performed for all of the patients for visceral localization and hence exclusion of secondary hydatidosis.

## RESULTS

There were seven cases of cerebral and three cases of spinal primary CNS hydatidosis, surgically and pathologically proven, which had no visceral abdominal or pulmonary localization on ultrasound and

chest x-ray (CXR), respectively (Table 1). The cases comprised six females and four males with an age range of 5-29 years and median age of 11.5 years. All of the cerebral hydatidosis presented with focal neurological deficits such as hemiparesis, headache, visual disturbance and/or seizure with average duration of 6-9 months. The primary spinal hydatidosis presented with incontinence and paraparesis with average duration of one year. CT and MRI showed intracranial and intra-axial cysts measuring 5-11 cm in size with average size of 8 cm, and mass effect and midline shift which were seen all cases. The cysts were anatomically located in the frontoparietal (3), frontotemporal (2), and parietooccipital (2) areas (Figure 1).

Table 1. CT/MRI findings and clinical presentation of primary intra cranial and spinal hydatidosis

Case, age (yrs) sex	Clinical signs & symptoms	Domicile	Type of cyst	CT/MRI findings	Intraoperative and post operative outcome
C1, 8, M	progressive visual impairment, seizure disorder	Arsi	Cyst with internal septation	6x5 cm cyst with detached membrane (CT), left frontoparietal; obstructive hydrocephalus and mid line shift	Cyst residual cavity gliosis.
C2,10,F	Visual impairment, seizure, hemiparesis	Arsi	Simple cyst with mural vesicle (developing daughter cysts)	Right frontoparietal 7x8 cm cyst with mural vesicles (CT) developing daughter cysts with obstructive hydrocephalus, midline shift	Ruptured cyst but removed enblock . Long term outcome unknown
C3,10,F	Progressive visual impairment seizure, hemiparesis	Bale	Cyst with daughter cysts	Right frontoparietal and tempoparietal cyst 6x8 cm(CT/MRI), obstructive hydrocephalus, midline shift	Complete removal with residual gliosis
C4, 29,M	Progressive visual impairment, hemiparesis	Dire Dawa, used to live in Bale during childhood	Cyst with internal septation	Left tempoparietal 6x5 cm cyst with perilesional edema and showing pericystic contrast enhancement (MRI) possibly infected	Complete cyst removal with residual gliosis.
C5, 13,F	Progressive hemiparesis and visual impairment	Arsi	Simple cyst	Right parieto-occipital simple cyst measuring 6x5 cm with inadvertent insertion of cystoperitoneal shunt catheter detaching the inner germinal layer (CT) with obstructive hydrocephalus, midline shift	Complicated cyst with peritoneal hydatidosis because of the peritoneal shunt detected on abdominal ultrasound. Long term outcome unknown

C6,5,F	Progressive visual impairment and seizure disorder, hemiparesis	Arsi	Cyst with daughter cyst	Left frontoparietal cyst with daughter cyst 8x9 cm (MRI), obstructive hydrocephalus, midline shift	Complete cyst removal. Long term outcome unknown.
C7,7,F	Progressive visual impairment with hemi paresis and seizure	Arsi	Simple cyst	Left parietooccipital measuring 7x11 cm (CT, MRI), obstructive hydrocephalus midline shift	Incomplete removal cyst ruptured with sub dural collections
C8,23,F	Paraparesis and double incontinence to stool and urine	Bale	Multiple para spinal cysts	T2 and T3 vertebral body destruction with paraspinal and intraspinal cystic extension sparing the posterior elements mimicking TB spondylitis and inadvertently treated for TB (MRI)	Complete removal of the cyst. Long term outcome not known
C9,14,M	Para-paresis with back pain and double incontinence	Bale	Multiple para spinal and epidural cysts	T10, T11 right pedicle erosion and para spinal mass with epidural extension compressing the thoracic cord. (MRI)	Complete removal of the cyst. Regained power and bladder and anal continence
C10,15,M	Double incontinence to stool and urine	Bale	cyst with daughter cyst	10 cm size pre-sacral pelvis and sacral spinal canal cyst with daughter cyst with para spinal and intra spinal epidural extension and sacral bone destruction. (MRI)	Complete removal. Long term outcome unknown

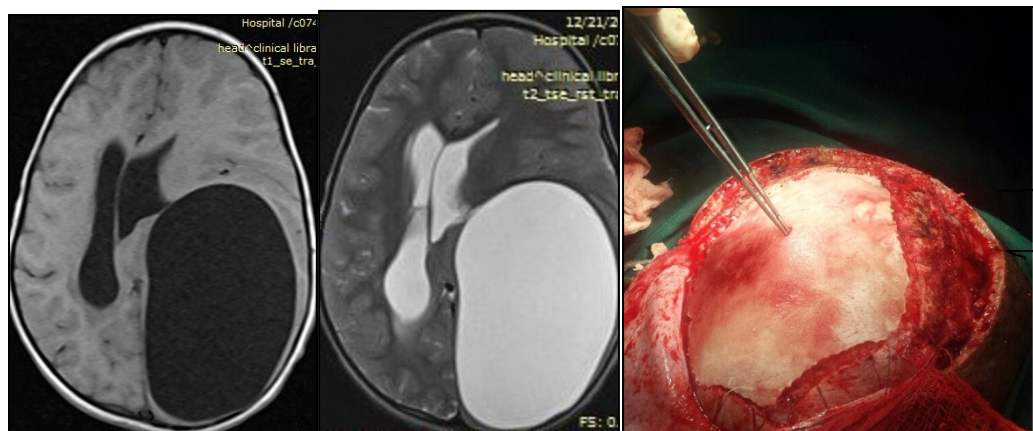


Figure 1. Left frontoparietal giant cyst with mass effect and midline shift. MRI Axial T1W (a) and T2W (b) images. Intraoperative image of the cyst (c).

Morphologically, two cases of cerebral (Figure 2) and one case of spinal hydatid (Figure 3) showed daughter cysts. Two cases showed multiseptated cysts. One was spontaneously and the other post-operatively infected, as assessed intraoperatively and from histopathology. An abscess was drained from the postoperatively diagnosed infection. Both multi-

septated cysts showed rim contrast enhancement and perilesional edema and detached floating membranes.

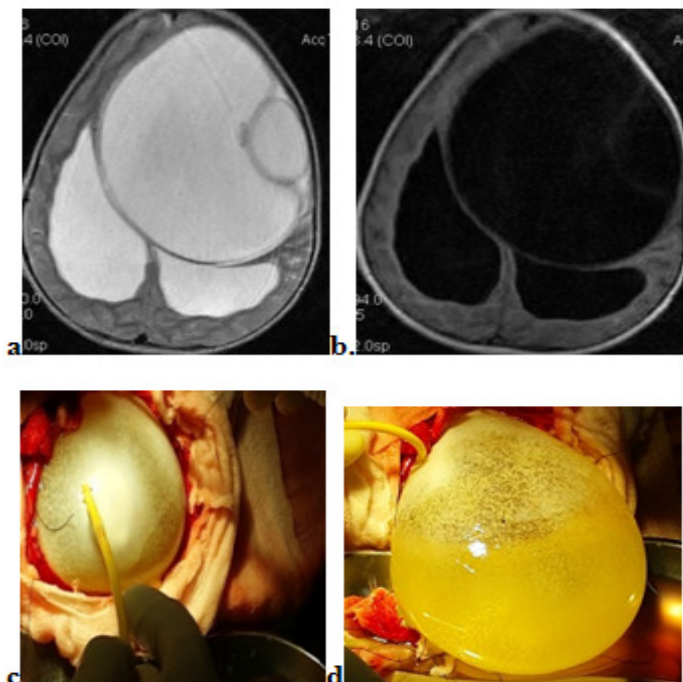


Figure 2. Axial T2W (a) and T1W (b) MRI images showing left frontoparietal giant cyst with mural daughter cyst and causing significant mass effect. Intraoperative findings (c) and postsurgical specimen of the cyst after total and intact removal of the cyst (d).

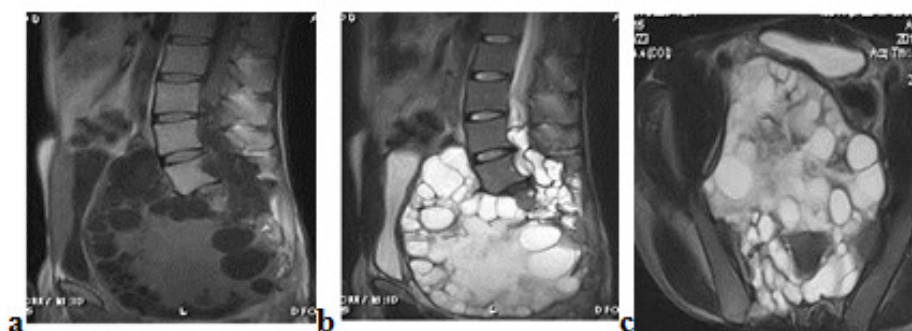


Figure 3. Large sacral cystic mass with multiple daughter cysts destroying the sacrum and extending into the sacral spinal canal. Sagittal T1W (a) and T2W (b) images, and T2W coronal images (c).

One case showed a large cyst with a mural nodule or vesicle. One case was mistaken preoperatively for an arachnoid cyst and had inadvertent cystoperitoneal shunt catheter insertion; later follow up CT revealed a detached germinal layer (Figure 4a, b, c) which prompted the postoperative consideration of hydatidosis. This patient had subsequently presented with peritoneal hydatidosis detected by ultrasound.

There were three cases of primary spinal hydatidosis, two of thoracic (Figure 5) and one sacral origin (Figure 3), all presenting with progressive paraparesis and incontinence. One had been mistaken for a spinal arachnoid cyst. One was misdiagnosed for tuberculous (TB) spondylitis and treated for TB, and subsequently

operated on due to a failed response to anti-TB treatment. MRI for this case showed multiple paraspinal and epidural spinal canal hydatid cysts, the sacral being a giant one measuring 11-12 cm with daughter cysts (Figure 3). All of them were associated with bony destruction relatively sparing the disks in the thoracic region while destroying that of the body and disk in the sacrum.

Nine cases came from rural Ethiopia, the South East Oromia Regional State, in particular Arsi/Bale zone. One case was from Dire Dawa but had lived as a child in Bale/Arsi. All had served or were serving as shepherds in their childhood with close contact to pet animals.

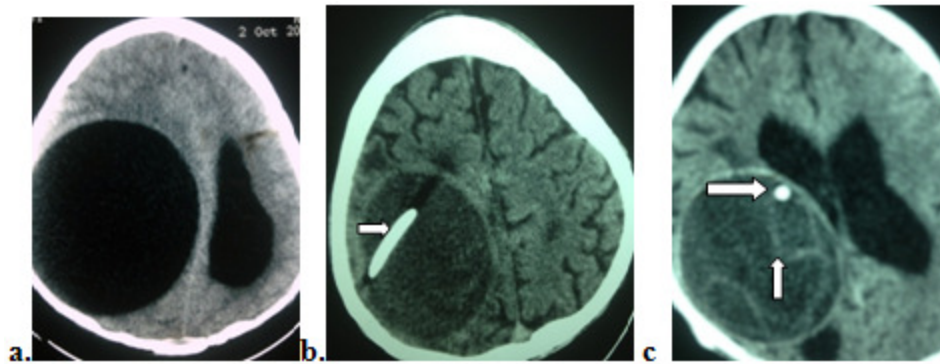


Figure 4. Preoperative CT image (a), post-operative cystoperitoneal shunt catheter inserted mistakenly placed for misdiagnosed arachnoids cyst with catheter inside the cyst (arrow) (b) and a floating membrane, depicted by vertical arrow and VP shunt catheter tip by horizontal arrow (c).



Figure 5. Sagittal T1W(a) and T2W(b) images showing destruction of the T2 and T3 vertebral bodies with parasagittal collection or mass. T1W and T2W axial images shown in (c) and (d), respectively.

## DISCUSSION

Although radiological features of CNS hydatidosis are very characteristic, definitive diagnosis of hydatidosis is based on histopathological examination of a surgical specimen. On imaging both CT and MRI demonstrate a spherical and well defined thin walled

and smooth homogeneous cystic mass with fluid density or intensity similar to that of CSF with significant mass effect and midline shift, and effacement of the lateral ventricles. It is often solitary and large sized, located in the parietal and temporal regions supplied by the middle cerebral artery, with average diameter of greater than 5-6 cm, the largest measuring 12 cm with an average doubling time of 4-5 months (13). Clinically it shows various forms of neurological dysfunction, often hemiparesis and

signs and symptoms of increased intracranial pressure (ICP). Cerebral hydatid disease accounts for 2% of all hydatid disease. The majority of the patients with primary intracranial hydatid disease in our study were children and young adults with a median age of 11.5 years, findings consistent with other reports indicating the majority (60-93%) of cases were acquired in childhood (13, 14). In contrast, in a 14 year review of primary hydatidosis Luo et al reported 21 cases of primary CNS hydatidosis of which 20 were brain with a median of 26.5 years of age, and only one case was spinal (15). These differences could be due to earlier diagnosis in our setting. Otherwise the imaging features and clinical presentations and the geographic distribution are similar.

On imaging by MRI or CT, cerebral hydatid cysts usually show a rim of low signal intensity on both T1 and T2 weighted images. The T2 weighted images are preferable because the low signal intensity cystic wall contrasts well with high signal intensity fluid. In non-contrast enhanced CT, the cyst wall is isodense or hyperdense to the brain tissue (15, 16). If daughter cysts and mural vesicles are noted on imaging it may establish the diagnosis; this was true for two of the cases in this study. With both CT and MRI, surrounding edema and rim enhancement may be noted in postsurgical or superinfected cases. Rim enhancement and detached floating membranes were seen in two of the patients in this study who were either surgically complicated or were initially superinfected. This has also been reported in other studies (16). Primary cerebral hydatid cysts are often solitary, in contrast to secondary cysts, and they are often large. When symptomatic, their size ranges from 5-11 cm and are located supratentorially in the arterial supply zone of the middle cerebral artery, i.e. temporoparietal region. These well described characteristics were also observed in our study.

The differential diagnoses of primary cerebral hydatid cyst include porencephalic cysts, arachnoid cysts, necrotic tumors and brain abscesses. In contrast to hydatid cysts, porencephalic cysts and arachnoid cysts are not spherical in shape and not surrounded entirely by brain substance. Arachnoid cysts are extraaxial and hydatid cysts often intra-axial. Porencephalic cysts result from insults to normal brain tissue and are lined by gliotic white matter which can easily be demonstrated with MRI. Cystic tumors of the brain can be differentiated by enhancement of the mural nodule, if present, at the periphery of the tumor. When pyogenic abscess shows cystic like central necrosis the rim enhancement and peripheral vasogenic edema is always present, but in contrast to this infected hydatid cysts may show floating membranes, easily demonstrated on both CT and MRI. Even with advanced CT and MRI techniques

imaging diagnosis of hydatid cysts may be difficult and may vary from classic images when they are infected and especially in alveolar echinococcosis (2). In such instances MR spectroscopy may help and may show lactate, acetate and succinate elevated resonance peaks in hydatid disease, small and reduced lactate resonance in arachnoid cysts, and increase in choline with decrease in N-acetylaspartate resonance in necrotic tumors (17, 18).

In the Ethiopia context, the preoperative clinical presentation of signs of raised intracranial pressure (ICP), focal neurological deficit (hemiparesis), seizures and visual disturbances in a patient coming from a rural setting, whose livelihood is based on animal husbandry, and are or were shepherds, combined with very specific imaging findings of a solitary large cyst causing significant mass effect should strongly favor the diagnosis of cerebral hydatidosis.

Spinal hydatidosis is less common than cerebral hydatidosis and accounts for only 1% of primary CNS hydatidosis (9). Clinically spinal hydatids may present with non-specific signs of cord and nerve root compression. Radiologically, spinal hydatids may present with non-specific features, and hence prompt diagnosis may be difficult. They may easily be mistaken for arachnoid cysts or paravertebral tuberculous abscess; this occurred in two of the cases in this study. Spinal hydatids lack the typical well formed cyst wall seen in cerebral cases, and are less spherical and often erode and thin the vertebral bones with easy extension into the less resistant paraspinal and intraspinal soft tissue. Occasionally features such as a mother cyst with multiple daughter cysts may be seen on cross-sectional imaging, as we observed in the lumbosacral hydatid cyst in this study. These features increase preoperative diagnostic accuracy.

The distribution of spinal hydatidosis comprises thoracic 50%, lumbar (20%), sacral (20%) and cervical (10%) regions. It is further classified into five groups: Intramedullary, intradural, extramedullary, extradural intraspinal hydatid cysts of the vertebrae and paravertebral hydatid disease (19). The first two groups are rare with few cases reported. In this study all the hydatids corresponded to the last three groups. Two were thoracic and one was lumbosacral in location. All of them had involved the vertebral bones with extension to the intraspinal canal and paravertebral spaces.

Despite modern surgical and pharmacological therapies, hydatid disease of the CNS remains difficult to cure and patient outcomes are less favorable due to high incidence of postoperative recurrence, which is reported in 20-40% of patients, especially in surgically ruptured cerebral hydatid disease cases (20-22).



Postoperative spinal hydatosis recurrence is not documented to date in the literature.

**Conclusion:** Although separate and brief case reports of cerebral and spinal hydatidosis have been published previously, a comprehensive descriptive study of assorted cases is reported. We consider it likely that we may see more of such cases as diagnostic and

treatment facilities expand. Therefore, the inclusion of hydatid disease of the CNS in the differential diagnosis of children and young adults with cerebral and spinal cysts should be emphasized. This can expedite early diagnosis and treatment and avoid morbid and lethal consequences of misdiagnosis and inappropriate procedures such as VP shunt catheter insertion.

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