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Case Series

Pathology Section

Primary Hydatid Cysts in Rare Sites: A Series of Four Cases

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ABSTRACT

Hydatid disease is caused by the larval stage of the tapeworm *Echinococcus granulosus*. The definitive hosts include dogs and canines. Humans are accidental hosts and do not play a role in the biological cycle of the worm. The mode of infection is through ingesting food contaminated with dog feces containing gravid proglottids and free eggs, which hatch in the intestine and form larvae. The larvae then penetrate the intestinal wall, enter the bloodstream, and typically settle in the liver followed by the lungs. However, in rare instances, hydatid cysts can occur in various unusual sites throughout the body and even in a disseminated manner, leading to atypical clinical presentations and diagnostic challenges. Hereby, authors report a series of four cases (three males and one female) of hydatid cysts occurring at unusual sites, mainly the brain, kidney, spleen, and lung. The common age group among the observed cases was the second to fourth decade, showing a male predominance. In cases of splenic and renal hydatid cysts, patients presented with intermittent dull aching pain over the left and right flanks, respectively. In both cases, ultrasonography revealed the presence of multiple cysts in the spleen and kidney. In the case of cerebral hydatid cysts, the patient provided a history of short-term memory loss along with a few non specific symptoms, and Magnetic Resonance Imaging (MRI) revealed a hyperintense cystic lesion. In pulmonary hydatid cysts, the patient presented with symptoms of cough, dyspnoea, chest pain, and expectoration. X-ray examination supported the presence of the cyst. Surgery, preferably radical cystectomy, is considered for treatment, and the diagnosis is always confirmed by histopathology.

Keywords: Atypical presentation, *Echinococcus granulosus*, Histopathology, Radical cystectomy

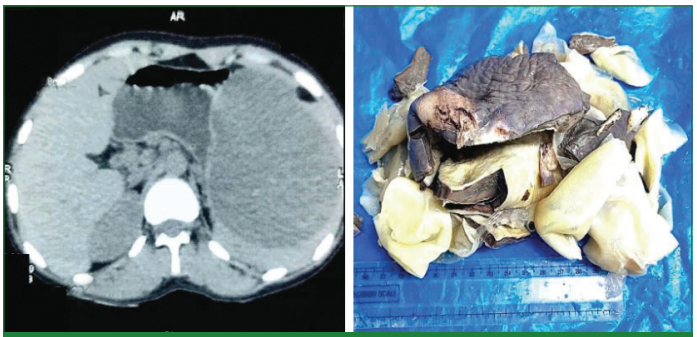
INTRODUCTION

The presentation of hydatid cysts in uncommon sites refers to their presence in locations within the body typically not associated with this parasitic infection. Hydatid disease commonly affects the liver, less frequently the lungs, and rarely the kidneys, spleen, and brain. The incidental discovery of hydatid cysts in unusual sites often occurs during routine medical investigations or diagnostic imaging studies. Therefore, hydatid cysts should be considered in the differential diagnosis of cystic lesions in various organs. Additionally, trauma or surgery at the primary site can lead to the dissemination of cysts to distant organs or tissues. The symptoms and clinical manifestations of hydatid cysts in these locations can vary widely, depending on the size, number, and location of the cysts. A case series on hydatid cysts focusing on cases in unusual locations such as the peritoneum, retroperitoneum, spleen, kidney, heart, pelvis, bladder, bone, pancreas, gallbladder, inguinal region, supraclavicular region, cerebral, spinal cord, and abdominal wall has been published in the literature [1]. Similarly, Behera BK et al., published a case series on hydatid cyst disease affecting the lung, spleen, kidney, and brain [2]. In present series, four cases of hydatid cysts involving the spleen, kidney, brain, and lung are presented with detailed clinicopathological information.

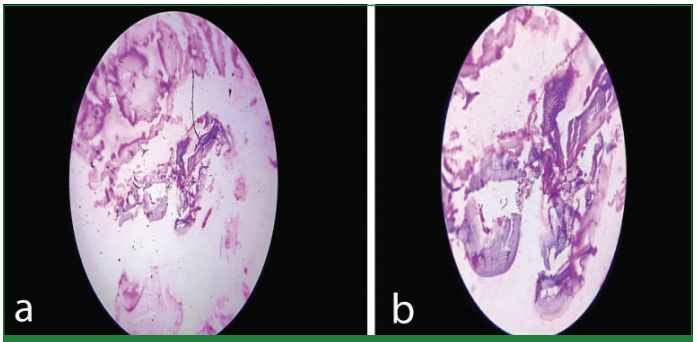
CASE SERIES

Case 1

A 26-year-old male presented with fatigue, anorexia, weight loss, and mild intermittent dull aching pain in the left flank for four months. There was no history of vomiting, fever, or trauma. On physical examination, the patient appeared pale, and a palpable mass was found in the left hypochondrium. Radiological examination (ultrasonography) revealed splenomegaly with multiple anechoic, smooth round cysts, the largest measuring 8×4×1 cc, which was confirmed on computed tomographic scanning as a benign splenic cyst [Table/Fig-1]. The patient subsequently underwent radical splenectomy. The specimen was sent to the Pathology Department in formalin for Histopathological Examination (HPE). The spleen measured 15×8×4 cc, along with multiple cysts sent separately. The largest cyst measured 10×5×1 cc, with a thin band of splenic parenchyma identified [Table/Fig-2]. On microscopic examination, the cyst wall showed an acellular, lamellated membrane with the presence of an inner germinal layer [Table/Fig-3a,b]. The differential diagnosis of splenic hydatid cyst includes splenic pseudocyst,



[Table/Fig-1]: Computed Tomography (CT) spleen showing hydatid cyst.
[Table/Fig-2]: Gross image of spleen with hydatid cyst. (Images from left to right)

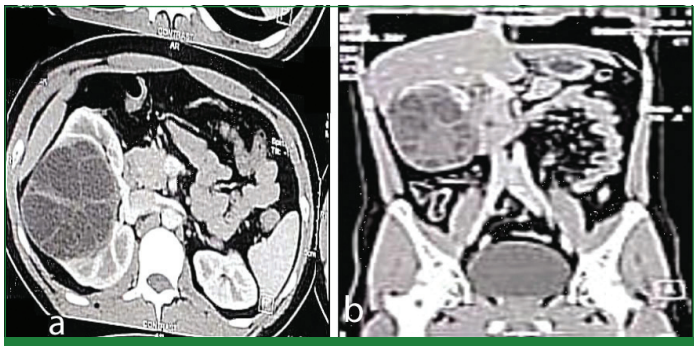


[Table/Fig-3]: a) Hydatid cyst wall from spleen (H&E 40X). b) Hydatid cyst wall from spleen (H&E, 400X).

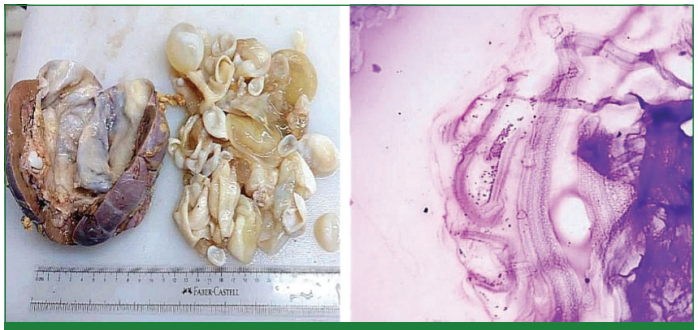
epidermoid cysts, and benign cystic lesions, which can easily be ruled out by histopathological examination. The postoperative period was uneventful, and the patient has been followed-up until now, with no complications or recurrence found.

Case 2

A 23-year-old male, non diabetic and non hypertensive, presented to the Urology Outpatient Department with complaints of fever and mild intermittent pain over the right flank for eight months. There was no history of dysuria, haematuria, or lower urinary tract symptoms. On per abdominal examination, a palpable mass was noted in the right upper abdomen. Urine microscopic examination revealed three pus cells per high power field, and urine culture did not reveal any growth of microorganisms. Ultrasonography revealed the presence of a space-occupying lesion measuring 12.4×11×9 cc in the middle and lower pole of the kidney. Contrast Enhanced Computed Tomographic (CECT) scanning of the kidney revealed a well-defined, non enhancing, thick-walled cystic lesion with multiple daughter cysts inside the interpolar region [Table/Fig-4a,b]. The patient underwent a right nephrectomy, and thereafter, the specimen was sent for histopathological examination. The right nephrectomy specimen measured 11×9×5 cc, and the attached ureter measured 7 cm in length [Table/Fig-5]. On cutting of right kidney, a large cystic area measuring 8×8×3 cc with a thin rim of normal parenchyma was observed. Multiple daughter cysts were identified altogether, measuring 100 cc, with the largest daughter cyst measuring 8×4×4 cc. Histological examination revealed an acellular lamellated membrane with an inner transparent nucleated lining representative of the germinal layer [Table/Fig-6]. Ovoid protoscolices were found attached to the germinal membrane. The outer pericyst shows an inflammatory reaction comprising fibroblasts, giant cells, and eosinophils. The distal resection margins of the ureter, Gerota's fascia, and perinephric fat were unremarkable. The postoperative period was uneventful, and the patient is being followed-up until now for any complications or recurrences.



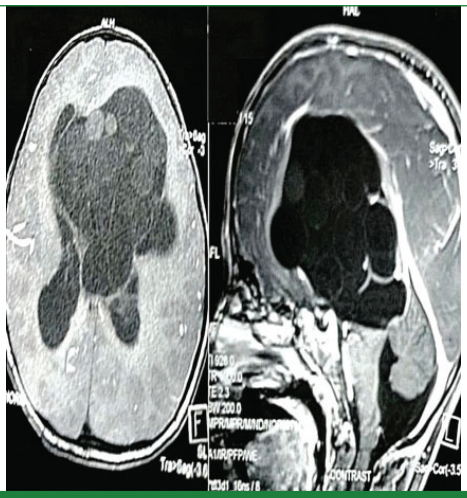
[Table/Fig-4]: a) CT image of kidney coronal view showing hydatid cyst in right kidney. b) CT image of kidney sagittal view showing hydatid cyst in right kidney.



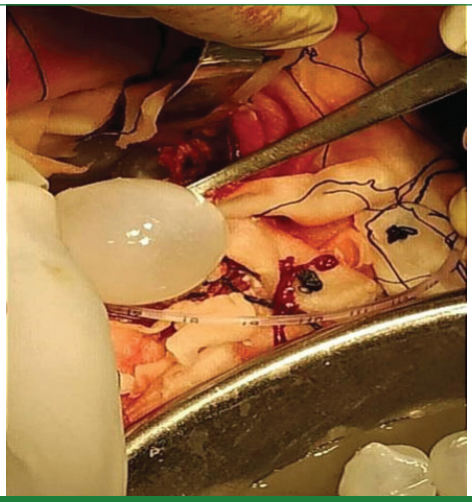
[Table/Fig-5]: Gross image of right kidney with hydatid cyst. **[Table/Fig-6]:** Hydatid cyst from right kidney showing germinal layer (H&E, 100X). (Images from left to right)

Case 3

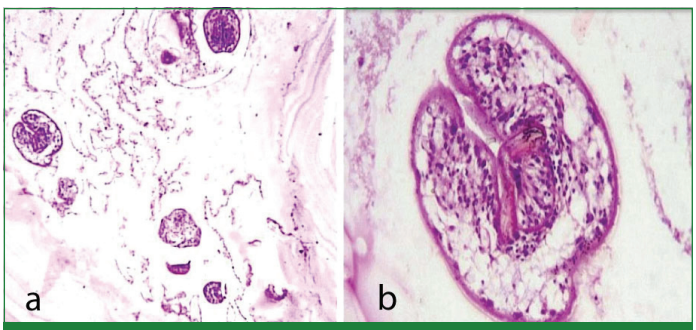
A seven-year-old girl complained of non specific symptoms like headaches, vomiting, sudden loss of balance, and short-term memory loss for six months. Conservative treatment, such as antihelmintics, was given for one month, but it proved futile as symptoms gradually intensified. MRI brain revealed a hyperintense cystic lesion involving the lateral and third ventricles [Table/Fig-7]. The postcontrast study showed no enhancement. The patient underwent surgery through Dowling's technique, and the cysts were dissected and removed. Gross examination showed multiple whitish cystic structures, the largest measuring 5×5×4 cc, and altogether measuring 100 cc [Table/Fig-8]. Microscopically, sections showed a cystic structure with an outer fibrous pericyst layer, middle laminated ectocyst layer, and inner germinative layer with brood capsules and daughter cysts [Table/Fig-9a,b], confirming primary cerebral hydatidosis. The postoperative period was uneventful, and the patient was discharged on postoperative day 10 and is being followed-up until now for any complications or recurrences.



[Table/Fig-7]: MRI image brain in coronal and sagittal view.



[Table/Fig-8]: Gross image of brain hydatid cyst.

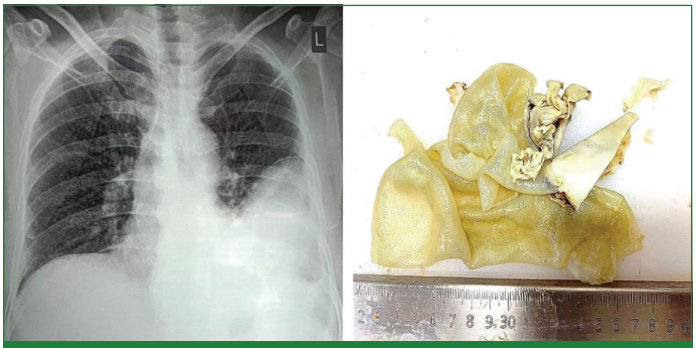


[Table/Fig-9]: a) Cerebral hydatid cyst showing daughter hydatid cysts (H&E, 100X). b) Cerebral hydatid cyst showing daughter hydatid cysts (H&E, 400X).

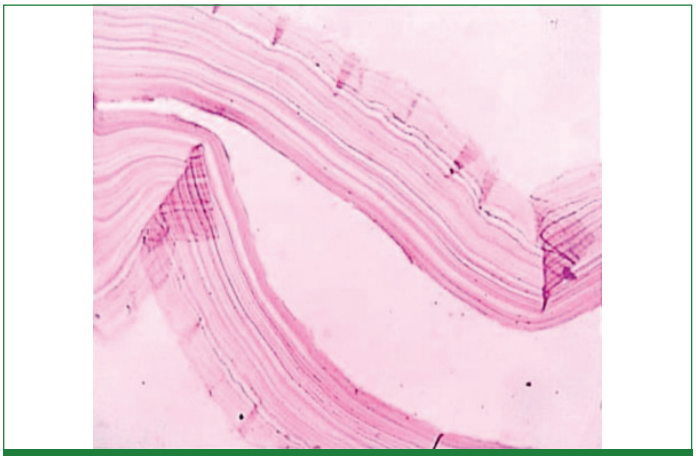
Case 4

A 52-year-old male with no significant past medical history presented to the Chest Outpatient Department with symptoms of cough, chest pain, expectoration, fever, and dyspnoea for one month. The radiological finding (X-ray) favoured a cyst measuring 6×6×3 cc

in the left hemithorax [Table/Fig-10]. On physical examination, there was decreased breath sounds in the left hemithorax. The patient underwent a left posterolateral thoracotomy through the 6th intercostal space, and the cyst along with its contents was removed. Multiple cyst walls, with the largest measuring 8×5×3 cc and the rest of the cyst walls altogether measuring 60 cc [Table/Fig-11], were submitted. Microscopy showed lung parenchyma with the presence of a cyst wall lacking an epithelial lining. The cyst wall is an acellular laminated membrane and pink. The outer pericyst shows an inflammatory reaction, with fibrinous exudates, granulation tissue, calcification, and lymphoid aggregates seen adjacent to the cyst. In the surrounding lung parenchyma, oedema and haemorrhage are observed [Table/Fig-12], confirming a pulmonary hydatid cyst. On follow-up, the patient is doing well without any complications.



[Table/Fig-10]: Chest X-ray showing space occupying lesion in left hemithorax.
[Table/Fig-11]: Gross image of hydatid cyst from lung. (Images from left to right)



[Table/Fig-12]: Lung hydatid cyst showing acellular lamellated membranes (H&E, 400X).

DISCUSSION

Hydatid cyst disease, also known as Echinococcosis, is caused by the larvae of the tapeworm Echinococcus. The disease primarily affects the liver and lungs, but it can also occur at other uncommon sites, such as the spleen, kidneys, and brain [3]. Brain involvement with hydatid disease occurs in 1-2% of all *Echinococcus granulosus* infections, and patients present with symptoms like headache, vomiting, and disorientation [4]. In this case, the patient also presented with short-term memory loss for the last six months [Table/Fig-1]. Due to the vasculature of the brain, a hydatid cyst is more common in the supratentorial region than the infratentorial region, as the middle cerebral artery supplies 75-80% of the brain. The worldwide incidence of splenic hydatid is 0.5-4%, and most cases are asymptomatic [5]. However, in present case, the patient presented with intermittent left flank pain [Table/Fig-13]. Spleen infestation generally occurs by the arterial route or via a retrograde venous path in patients with portal hypertension, bypassing the liver and lung [6].

A few cases of pulmonary hydatid cyst have also been reported in the literature. For instance, Ghallab NH and Alsabahi AA, reported a case on a giant hydatid cyst of the lung, where the patient presented with a dry cough and mild fever, without any chest pain, dyspnoea, or weight loss [6]. In the present series, the patient also presented with dyspnoea and chest pain, along with cough and fever [Table/Fig-13].

Hydatid cyst disease is rarely presented in the kidneys, and isolated renal occurrence is estimated to be about 2-4% of all cases [7]. Similarly, a case report of primary renal hydatid cyst has been reported by Choi H et al., where the patient presented with left flank pain, haematuria, and dysuria [8]. A CT scan revealed a cystic lesion in the left kidney, which was initially regarded as a renal malignancy, and radical nephrectomy was performed to remove the renal mass. The final pathological diagnosis was a renal hydatid cyst [9]. In the present study, the patient with renal hydatidosis did not have haematuria or dysuria, and the Computed Tomography (CT) scan revealed multiple daughter cysts [Table/Fig-13]. The lungs and liver, by virtue of their capillary beds, filter out the majority of *Echinococcus Granulosus* larvae and are the most common sites of disease manifestation [10]. The occurrence of hydatid cysts at uncommon sites can present diagnostic and therapeutic challenges. Hence, hydatid cysts should also be considered in the differential diagnosis of cystic lesions in solid organs such as the brain, kidneys, spleen, and adrenals [11]. Correct diagnosis is imperative to start preoperative albendazole and take measures to prevent

Diseases	Age (years), gender	Radiological findings	Gross findings	Microscopic findings
Splenic hydatidosis	26, male	Ultrasonography whole abdomen revealed splenomegaly with multiple anechoic, smooth round cysts largest measuring 8×4×1 cc which was confirmed on computed tomographic scanning as benign splenic cyst.	Cut open spleen received measuring 15×8×4 cc. along with multiple cyst walls sent separately. Largest cyst wall measure 10×5×1 cc and second largest measure 5×5×1 cc.	Cyst wall showed histology of acellular lamellated membrane with presence of inner germinal layer with normal splenic parenchyma. Minimal tissue reaction was noticed.
Renal hydatid cyst	23, male	Ultrasonography revealed presence of space occupying lesion measuring 12.4×11×9 cc in the middle and lower pole of the kidney. Multiple small cysts were noted present peripherally. Contrast enhanced computed tomographic scanning of kidney revealed a well-defined non enhancing thick walled cystic lesion with multiple daughter cysts inside interpolar region.	Right Nephrectomy specimen measured 11×9×5 cc and attached ureter measured 7 cm in length. On cutting open the right kidney large cystic area was identified measuring 8×8×3 cc with a thin rim of normal parenchyma. Multiple daughter cysts were identified altogether measuring 100 cc and largest daughter cyst measuring 8×4×4 cc.	Cyst wall show histology of acellular lamellated membrane with inner transparent nucleated lining representative of the germinal membrane. Ovoid protoscolices were found attached to the germinal membrane. The outer pericyst shows inflammatory reaction comprising of fibroblast, giant cells and eosinophils. Ureter, perinephric fat and fascia of gerota were unremarkable.
Primary cerebral hydatidosis	7, female	MRI brain revealed hyperintense cystic lesion involving lateral and third ventricles. Postcontrast study showed no enhancement.	Multiple whitish cystic structures received largest measuring 5×5×4 cc and altogether measuring 100 cc.	Microscopically, sections showed cystic structure with outer fibrous pericyst layer, middle laminated ectocyst layer and inner germinative layer with brood capsules and daughter cysts.
Pulmonary hydatid cyst	52, male	X-ray revealed a cyst measuring 6×6×3 cc in the left hemithorax.	Multiple cysts walls received largest measuring 8×5×3 cc and rest other cyst walls altogether measured 60 cc.	Microscopically sections showed lung parenchyma with presence of cyst wall lacking epithelial lining. The cyst wall is acellular laminated membrane and pink. The outer pericyst shows inflammatory reaction. Fibrinous exudates and granulation tissue were seen adjacent to cyst.

[Table/Fig-13]: Summary of the present study cases.

spillage. Complications like a severe allergic reaction and parasitic dissemination are rare with the use of scolicidal agents [12]. Currently, the most effective treatment for hydatid disease is still surgery. The main purpose of surgery is to prevent complications such as compression of surrounding structures, infection, or cyst rupture.

A similar case has been reported in the literature by Garg M et al., where ultrasonographic examination revealed a heterogeneous solid cystic lesion in the left suprarenal region measuring 15×12 cm^2 and it was proven to be splenic hydatidosis on histopathological examination [13]. Similarly, Dilli A et al., has reported a case of splenic hydatid disease where the MRI revealed a multivesicular cystic mass near the splenic hilus. The patient underwent splenectomy, and the lesion was proven to be a hydatid cyst pathologically [14]. Cases of hydatid cysts in the spleen, lung, brain, and kidney have been reported in the literature [Table/Fig-14] [1,3-11,13,14]. A case series discussing valuable insights into the clinical presentation, management strategies, and outcomes associated with these manifestations would contribute to the existing medical literature and increase our knowledge of the disease. One potential topic of discussion is the various uncommon sites of presentation of hydatid cysts, exploring the clinical signs, symptoms, and challenges associated with hydatid cysts in such uncommon sites would help healthcare professionals recognise and manage such cases.

Name of the author and year of the study	Place of case reported	Site
Keser SH et al., 2017, [1]	Istanbul, Turkey	Spleen, kidney
Al Taei TH et al., 2022, [3]	Busaiteen, Bahrain	Kidney
Senapati SB et al., 2015, [4]	Cuttack, Odisha, India	Brain
Merad Y et al., 2021, [5]	Sidi-Bel Abbes, Algeria	Spleen
Ghallab NH et al., 2008, [6]	Sana'a, Yemen	Lung
Paramythiotis D et al., 2016, [7]	Thessaloniki, Greece	Left kidney
Choi H et al., 2014, [8]	Seoul, Korea	Kidney
Dudha M et al., 2018, [9]	Surat, Gujarat, India	Lung
Kumar L et al., 2021, [10]	Chennai, Tamil Nadu, India	Spleen
Misra A et al., 2021, [11]	Bhubaneswar, Odisha, India	Kidney
Garg M et al., 2015, [13]	Haryana, India	Spleen
Dilli A et al., 2011, [14]	Ankara, Turkey	Spleen

[Table/Fig-14]: Published cases of hydatid cysts of last 15 years [1,3-11,13,14].

CONCLUSION(S)

This case series highlights the occurrence of hydatid cysts in unusual sites, which can pose diagnostic challenges and require a multidisciplinary approach for effective management. The presented cases demonstrate that hydatid disease is not limited to the liver and lungs but can involve various organs throughout the body. Radiological imaging techniques such as ultrasound, CT and MRI play a crucial role in detection. The differential diagnosis was ruled out by histopathological examination. In conclusion, present case series highlights the significance of considering hydatid cysts in unusual sites, as well as the need for a comprehensive approach involving clinical suspicion, radiological imaging, and surgical expertise for effective management.

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