Original Research Article

Hydatid cystic disease at unusual sites

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Abstract

Hydatid disease is a zoonotic infection caused by Echinococcus granulosus, which belongs to phylum Platyhelminthes. It is endemic in many parts of the world including India. Though Hydatid disease commonly occurs in liver and lung no organ or tissue is immune in the body. When it presents in unusual sites Hydatid disease often poses a diagnostic challenge. A high index of suspicion, comprehensive radio imaging investigations are often required to evaluate these cases. In the study we have conducted for 4 years, we reported a total no of 7 Hydatid cases that presented at unusual sites. Most common rare presentation in our study was renal hydatid disease, 3 cases; and one each in pancreas, Spleen, peritoneum and pelvis.

Key words

Echinococcus granulosus, Hydatid disease, Renal hydatid cyst, Unusual sites.

Introduction

Human Echinococcosis is caused due to infection by larval stages of Echinococcus granulosus. Humans act as an accidental intermediate host. It has a broad range of geographical distribution. Highest prevalence is in Eurasia, Africa, Australia and South America [1, 2]. Cystic echinococcosis commonly involves the organs liver (65%-75%) and lungs (15%-25%), although it can occur in any organ and tissue. Third commonest site is spleen (5%), followed by kidney (4%), bone (1%-4%) and pancreas (0.1%-2%) [3-5]. In both endemic and non-endemic areas Hydatid cystic disease at unusual sites frequently cause diagnostic problems giving rise to delay in diagnosis and potentially serious complications [5]. Morphologically hydatid cyst has 3 layers, outer fibrous layer formed by host

tissue, middle acellular layer that shows concentric laminated membrane on microscopy. Inner germinal layer is thin, translucent grossly, which is one cell thick with attached brood capsules and protoscolices. A clinical diagnosis of Cystic echinococcosis requires combination of physical examination, imaging techniques and serological test by ELISA. Though radiological signs are normally non-specific, Computed tomography, Magnetic resonance imaging and ultrasonography are useful for deep seated lesions in all organs [5]. The characteristic imaging findings described are calcification of the cyst wall, presence of daughter cysts and membrane detachment.

Objectives

Diagnosis of Hydatid cystic disease at unusual sites like pancreas, kidney, retroperitoneum, bone and brain often pose preoperative diagnostic difficulties, compared to those at common organs like liver and lung even in endemic areas. The present study is conducted to estimate the incidence of Hydatid cystic disease at rare sites, among all the Hydatid cystic disease cases; to analyze, compare and correlate histopathology diagnosis with that of the clinical mode of presentation and comprehensive radiological findings in these cases.

Materials and methods

The present retrospective study was conducted in the department of Pathology, Gandhi Hospital during a period of 4 years from January 2012 to December 2015. All the cases morphologically diagnosed as Hydatid cystic disease during this period are included in the study. A special emphasis is made in the study on cases which presented at unusual sites like kidney, pancreas, spleen, retroperitoneum and pelvic regions. Specimens are fixed in formalin; HP diagnosis is made on routine H&E sections. Relevant detailed clinical history and laboratory parameters are noted. Radiological findings and diagnosis for these 7 cases including Ultrasound and CECT findings are recorded from the department of Radio imaging, Gandhi Hospital.

Results

A total no of 34 cases which are diagnosed as Cystic echinococcosis on HPE during study period are included in the present study. Out of total no of 34 cases that were diagnosed as Hydatid cystic disease 7 cases are found to present in unusual sites (20.5%). Male and female ratio of incidence found to be 5:2. Youngest patient reported is 27 years. whereas the highest age is 65yrs. Out of 7 cases of cystic echinococcosis at unusual sites 6 are isolated cases and one secondary. Maximum no of cases i.e. 3 occurred in kidney (8.8%) as isolated cases. One case reported in pancreas (2.9%), spleen one (2.9%) and retroperitoneum one (2.9%). We have also reported one case of pelvic Hydatid cystic disease (2.9%) which is a secondary case. The commonest clinical presentation observed is abdominal pain, and in 3 cases o/e mass was palpated. Out of 7 unusual cases histologically diagnosed to be hydatid cystic disease, in 6 cases clinical diagnosis either established or suspected depending on clinical presentation and Radio imaging techniques (87.7%). In one case, pancreas with cystic mass, pre-operative diagnosis was missed (**Table – 1 to 4**).

Table - 1: Age incidence (unusual cases).

Age in years	No. of cases	Incidence %
20- 30	1	14.28
30- 40	1	14.28
40- 50	3	42.85
50--60	1	14.28
60- 70	1	14.28

Table - 2: Incidence of cases at unusual sites (Out of Total Cases).

Site	No. of cases	Incidence %
Kidney- isolated	3	8.8
Pancreas- isolated	1	2.9
Spleen- isolated	1	2.9
Retroperitoneum- isolated	1	2.9
Pelvis- secondary	1	2.9

Table - 3: Clinical findings and lab parameters.

Case	Clinical Findings	Lab Findings
1. Male 39, kidney	Pain in right loin	CUE – 8 to 10 RBC
2. Female 47, kidney	Pain in right loin and lump abdomen	WNL
3. Male 49, kidney	Past history of Hydatid disease Passage of vesicles through urethra	Hydatidurea
4.Female 27, pancreas	Pain abdomen, lump abdomen 5x5 cm in epigastric region	WNL
5.Male 65 years, Spleen	Pain in umbilical region, extending to left hypochondriac region with irregular lump of 4x5 cm.	Hb-9.0gms%
6. Male 50 years, Retro-peritoneum	Pain right side of abdomen. Fever on off 2 months	WNL
7.Female 55 years Pelvis	Pain in lumbar region	WNL

Table - 4: Radiological findings and morphology.

Case	Radiology findings	Morphology
1.Male 39 years, kidney	CECT - Well defined cyst with daughter cysts in lower poll (**Figure - 8**) US - 8x4 cm cyst containing daughter cysts	Hydatid cyst membranes with brood capsules and protoscolices, focal compressed renal parenchyma (**Figure - 9, 10, 11**)
2.Female 47 years, kidney	CECT - Multiple non enhancing cysts with small peripheral calcifications in right kidney (**Figure - 3**) US - Multiple diffuse cysts	Fibro collagenous cyst wall with chronic inflammation and Laminated membranes of Hydatid cyst
3.Male 49 years, kidney	CECT - Hydatid cyst occupying entire Left kidney. No parenchyma seen (**Figure - 1 and 2**)	Nephrectomy specimen, entire kidney transformed into cystic mass. Consistent with Hydatid disease
4.Female 27 years, Pancreas	CECT - 3.5x3 cm mass, cystic and solid lesion in body and tail US - echoic thick walled lesion in body and tail. ?Pancreatic neoplasm ?Pseudo cyst of pancreas	Wipples resection specimen. Cystic lesions with laminated membranes, protoscolices s/o Hydatid disease of Pancreas (**Figure - 5, 6, 7**)
5. Male 65 years, Spleen	US - Well defined heterogeneous multiple lesions of size 18x9 cm. ?Hydatid cyst of spleen	Structure of spleen with Hydatid disease
6.Male 50 years, Retro peritoneum	CECT - ?Retroperitoneal Hydatid cyst (**Figure - 4**) US - S/o Hydatid cyst	Necrosis, Laminated membranes with daughter cysts of Hydatid disease.
7. Female 55 years, Pelvis	CECT and US - Multiple cystic lesions In both lobes of liver Well defined cystic lesions in pelvis S/o Peritoneal Hydatid disease	Sections from both liver and pelvic mass show cystic lesion Consistent with Hydatid disease.

Figure - 1 and 2: CECT abdomen show multiseptated Hydatid cyst left kidney with peripheral calcification.

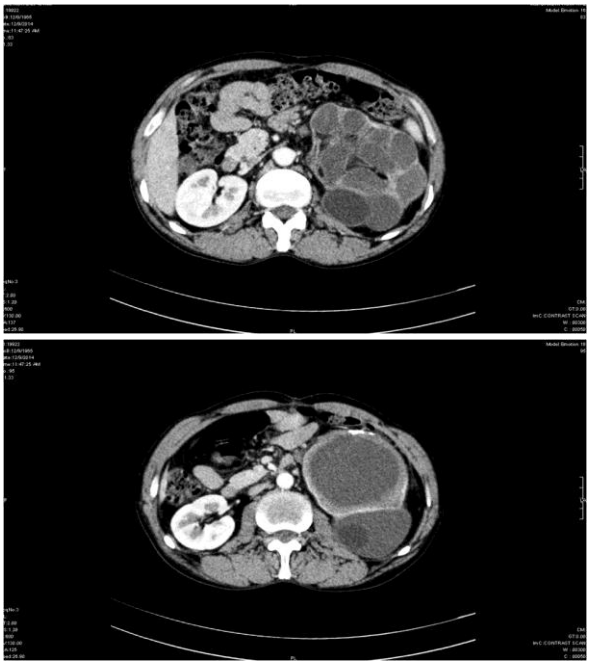


Figure - 3: CECT abdomen show large Hydatid cyst lower pole of right kidney.

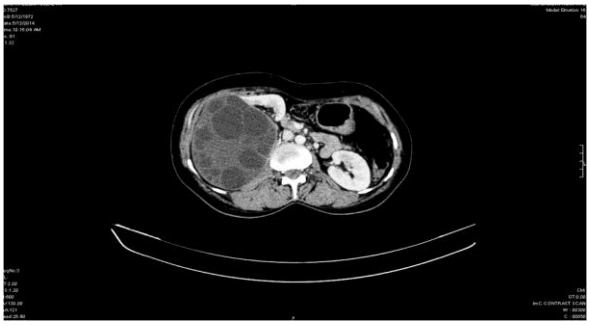
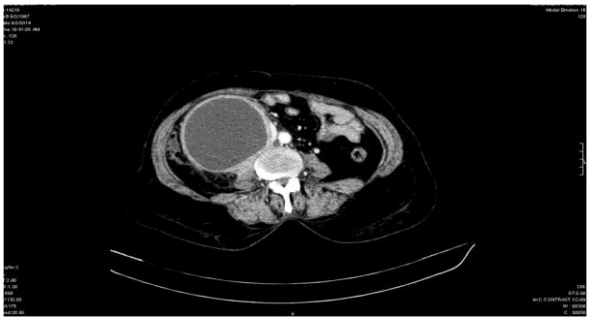


Figure - 4: CECT abdomen show large retroperitoneal Hydatid cyst.



Discussion

Hydatid cystic disease is endemic in several countries and constitutes a major public health problem, in several other countries, it is an emerging and reemerging chronic disease [3]. Primary echinococcosis is caused by ingestion of eggs, which hatch into larvae in the intestine. Embryo enters the circulation by penetrating the mucosa of intestine. In most of the cases final destination is through portal circulation into liver. If embryo continues through pulmonary capillary bed and enters systemic circulation Hydatid cyst can lodge and develop at any organ or site in the body [4]. Organs also may be reached through lymphatics resulting in primary Hydatid cyst. Secondary echinococcosis follows spillage of small daughter cysts or protoscolices due to trauma or surgery.

Figure - 5: Gross specimen showing Hydatid cyst of pancreas along with part of small intestine.

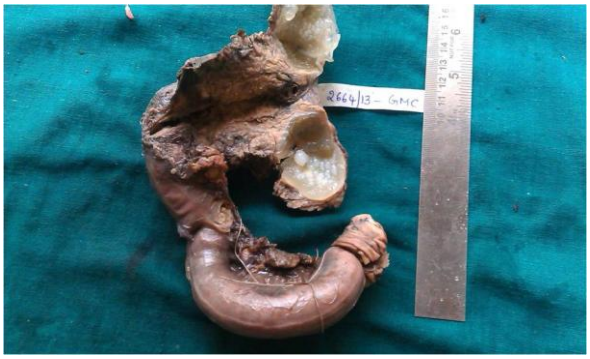
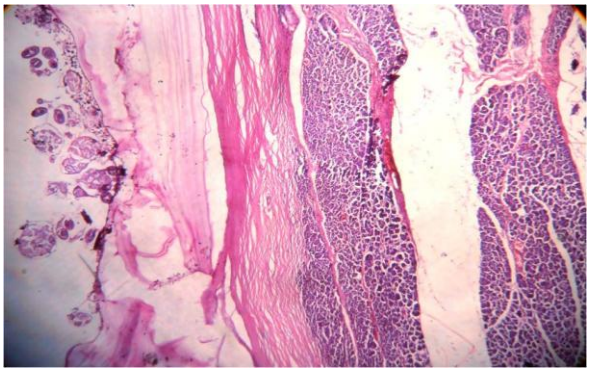


Figure - 6: H & E section (10X) showing Hydatid cyst with laminated membranes along with daughter cysts within pancreas.



Out of 7 cases at unusual cases in our study 3 cases are isolated renal hydatid cysts, one reemerged primary case. The incidence we found is 8.8% which is little high compared to other studies (2-5%) for reasons unknown [6, 7]. Patient with isolated reemerged case, presented

with passing of vesicles through urethra 4 yrs after laparotomy for Hydatid cystic disease of the same left kidney. If all three layers of hydatid cyst rupture, communicates with calyces and pelvis, it is called communicating cyst. This results in hydatiduria, passing of daughter cysts and protoscolices through urethra, seen in 10%-20% of renal Hydatid cystic disease [6]. All the three cases have shown cystic lesions with daughter cysts inside on CECT. (**Figures - 1, 2, 3 and 8**)

Figure - 7: H & E section (40X) showing Hydatid cyst with laminated membranes along with daughter cysts within pancreas.

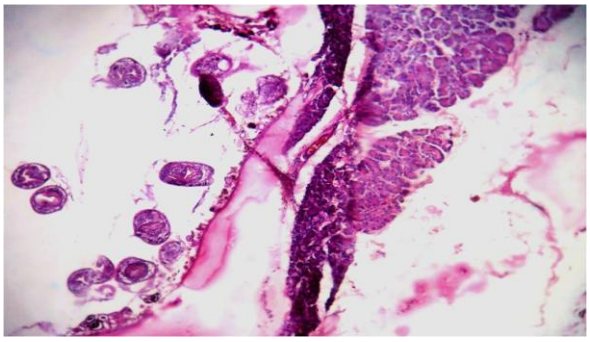


Figure - 8: CECT abdomen show Hydatid cyst in lower pole of left kidney.

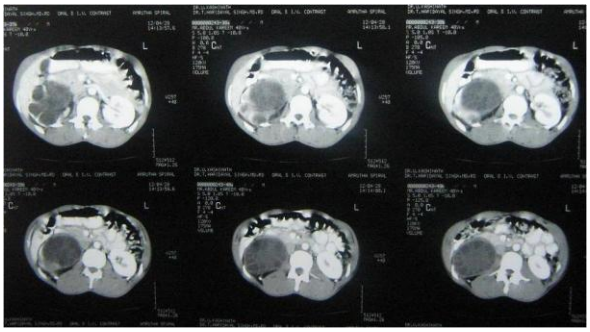


Figure - 9: Gross specimen showing Hydatid cyst along with membranes & daughter cysts.

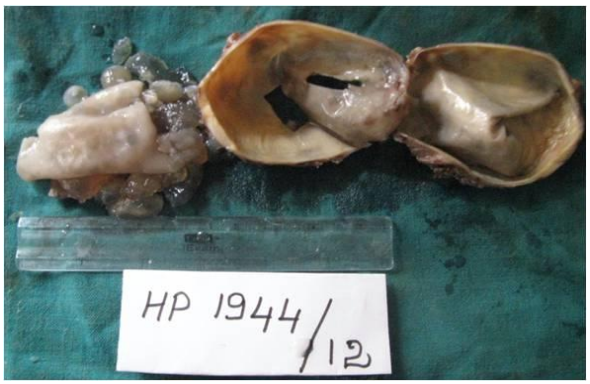
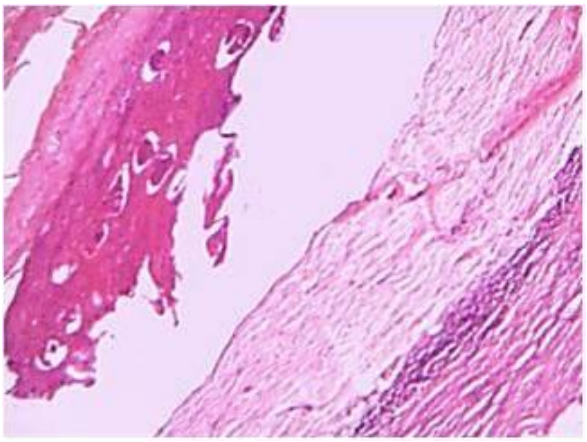


Figure - 10: Wet mount of Hydatid cyst fluid showing protoscolices.

Figure - 11: H & E section (10X) showing Hydatid cyst laminated membrane along with compressed renal parenchyma.



Incidence of primary Hydatid cystic disease of pancreas is also rare, varies from 0.1% to 2% [4]. We reported one case of isolated cystic echinococcosis (2.9%) in a 27 years female patient. Although cystic lesions of pancreas are easily identified by ultrasound, CT scan and MRI have limited sensitivity [4, 8]. CECT of this patient revealed hypodense lesion of 3x3.5 cm with both solid and cystic lesions in body and tail. A clinical diagnosis of a neoplasm, pseudocyst of pancreas was done, for which Wipples resection was done and sent for HPE. Pancreas showed grossly and microscopically well-defined hydatid cysts with daughter cyst within body and tail. Spleen is the third commonest site for Echinococcosis in endemic areas after liver & lung. Worldwide incidence is 0.5%-4% [9]. It usually presents as a simple cyst of spleen and does not have classic imaging features. But Hydatid cyst of spleen should be

considered in all endemic areas with cystic lesion of spleen, until proved otherwise [9]. Among 7 cases at unusual sites there is one isolated hydatid cystic case of spleen in our study (2.9%). Patient is 65yrs male who presented with pain in the left hypochondrium. US revealed a well-defined heterogeneous multicystic lesion of 1.8x9 cm. Preoperatively possibility of Hydatid cyst is suspected, and per operation findings are consistent with that of Hydatid cystic disease of spleen, for which splenectomy was done.

Isolated retroperitoneal hydatid infection is extremely rare (0.8%), refers to the presence of Hydatid cysts in retroperitoneal region without involvement of intraperitoneal (mainly liver) organs [10, 11]. Excision of cysts may not be possible because of dense adhesions [10]. We have reported one case (2.9%) of primary retroperitoneal hydatid cyst in a 50 years male patient. Both US and CECT of this patient revealed a lesion 10x 10 cm with internal septations and peripheral calcifications within the lumbar region. At the time of surgery a huge retroperitoneal mass in the lumbar region with hydatid cysts and daughter cysts observed, Cysts are extracted from adjacent structures and daughter cysts are evacuated and sent for HPE.

Primary pelvic Hydatid cystic disease is a very rare entity and almost always secondary rupture of hepatic cysts into abdominal cavity [12]. A 55 years female patient presented with pelvic hydatid mass in our study. CECT of this patient revealed well defined heterogeneous cysts with multiple daughter cysts in both right and left liver lobes. In addition a well-defined hypoechoic multiple cystic structures noted in both adnexae of pelvis. A secondary bilateral pelvic Hydatid cystic disease on radio imaging findings was made. The above both cases are confirmed by HPE subsequently.

Conclusion

Hydatid disease is a complex chronic infection, when it occurs in rare sites preoperative diagnosis poses a challenge because of atypical clinical presentation. Hydatid cystic disease should always be suspected in all cystic lesions of radio imaging investigations particularly in endemic areas to prevent life threatening complications and avoid unnecessary radical surgeries.

References

1. Imod S. Dander, B.S. Anand. Hydatid cysts. Medspace, updated. Aug 2014.
2. Francesca Rinaldi, Enrico Brunetti, Andreas Neumeier, Marcello Maester. Cystic echinococcosis of the liver: A primer for hepatologists. World j. Hepatol., 2014; 6(5): 293-305.
3. Pedro moro, Peter N. Schantz. Echinococcosis; A review. International journal of infectious disease, 2009; 13(2): 125-33.
4. J.E.J. Krige, K. Mirza, P.C. Bornman, S.J. Beningfield. Primary Hydatid cysts of the pancreas. SAJS, 2005; 43(2): 37-40.
5. Marvet Yuksel, Demirpolat, Ahmet sever, Sevgi Bakaris. Hydatid disease involving some rare locations in the body. Korean j. Radiology, 2007; 8(6): 531-540.
6. Mongha R, Srinivasan Narayan, Anup K. Kundra. Primary Hydatid cyst of kidney and ureter with Gross hydatiduria: A case report and evaluation of radiological features. Indian J. of Urology, 2008; 24(1): 116-117.
7. R. Younz Akmen, Abantlzzet Bayasal Bolu, Semih Y. Sargin, Ahmet H. Yazicioglu. Hydatid disease of the kidney, A retrospective review of 13 cases; Infect Urol., 2000; 13(5).
8. Amir haushang Mohammed Alizadeh. Primary Hydatid cyst of pancreas; A review. J. oF Pancreas, 2016; 17(3): 250-256.
9. Khaleed Rasheed, Showkat Ali, Ajaz Ahmed Telwani. Hydatid cyst of spleen, a diagnostic challenge. N. American J. Med Sciences, 2013; 5(1): 10-20.

10. Sekar N.N, Madhavan K.K, Yadav RV, Katariya RN. Primary retroperitoneal Hydatid cyst; A report of 3 cases and review of literature. JPGM, 1982; 28(2): 112-114.
11. Yashdeep Sharma, Raghunath Prabhu, Vijay Koduru, Amardeep Nandra, Sakshi Sadhu. Primary Retroperitoneal Hydatid cyst; A rare presentation of Echinococcosis. J. Surg Arts, 2015; 1: 26-28.
12. V. Seenu, M.C. Misra, S.C. Tiwari, C. Chandrasekhar. Primary pelvic Hydatid cyst presenting with Obstructive uropathy and renal failure. Post graduate medical journal, 1994; 70(830): 930-932.