UNCOMMON SITES OF A COMMON DISEASE — HYDATID CYST

Shahjehan Alam[1], Ummara Siddique Umer[2], Seema Gul[3], Syed Ghaus[4], Bushra Farooq[5], Farah Gul[6]

[1-5] Radiology Department, Rehman Medical Institute, Peshawar - Pakistan.
[6] Scientific Officer, PCSIR Laboratories, Peshawar - Pakistan.
Address for correspondence:
Dr. Shahjehan Alam
Assistant Professor, Radiology Department, Rehman Medical Institute, Peshawar - Pakistan.
E-mail: shahjehan135@yahoo.com
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ABSTRACT

Objective: To review uncommon sites of hydatid cysts and to assess radiological features of hydatid disease in head, neck, spine and heart.

Methodology: A retrospective study of 50 cases of hydatid disease attended at Radiology department of Rehman Medical Institute, Peshawar between May 2012 and November 2013 was conducted to determine the incidence and imaging presentations of atypical localization of the disease. After taking permission from ethical committee, indoor and outdoor patients with hydatid cysts were selected for the study. All data was entered and analyzed using SPSS version 10.0.The data was assessed using Microsoft excel 2007.

Results: A total number of 50 patients had Hydatid cysts. Two patients had multiorgan involvement i.e., one had liver and lung involvement while other had liver and brain involvement. The cysts were present in brain (n=3, 6%), spine (n=2, 3%), neck soft tissues (n=1, 1%), heart (n=2, 3%), ovary (n=3, 6%), kidney (n=1, 1%), spleen (n=3, 6%), peritoneal cavity (n=2, 4%) and pancreas (n=1, 1%). Liver was involved in 20 (41%) cases while lung was involved in 14 (28%) cases.

Conclusion: Hydatid disease can involve unusual sites like heart, brain, neck, spine and pancreas. It may occur anywhere, from the big toe to the crown of the head and should be kept in consideration when a cystic lesion is encountered anywhere in the body especially in endemic areas.

Key Words: Hydatid cyst, Computed tomography, Magnetic Resonance Imaging, Ultrasound.

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INTRODUCTION

Hydatid disease is a zoonosis caused by the infestation of the oncosphere (embryo) of Echinococcus Granulosus. The disease is prevalent in most parts of the world, especially in sheep and cattle farming areas of Asia, North and East Africa, South America, Australia and the Middle East[1]. In Pakistan, many studies have been conducted describing regional incidence of disease. A study conducted by Gandahi et al of Sindh agricultural university revealed human hydatidosis incidence of 0.1 %[2].

The adult worm lives in the intestine of the definitive host, which includes dogs and other carnivorous animals. Sheep, cattle and humans act as the intermediate host. Man becomes infected through contact with a definitive host or by consuming contaminated water or vegetables[2].

The wall of hydatid cyst (HC) contains three layers. The outermost layer is the pericyst, the middle layer is the laminated membrane and the innermost layer is the germinal epithelium (endocyst) which represents the true wall of the cyst with the living parasite[3].

A pathology-based classification has recently been introduced by Lewall [4]. The hydatid cyst always starts as a fluid-filled, cyst-like structure (Type I) which may proceed to a Type II lesion if daughter cysts and/or matrix develop. In some instances the Type II lesion becomes hypermature and due to starvation dies to become a mummified, inert calcified Type III lesion. Type I and II lesions may undergo three types of rupture: contained, communicating and direct. Contained rupture is clinically silent, but communicating rupture may cause biliary obstruction and evacuation or infection of the cyst. Direct rupture has the greatest clinical consequences which include anaphylaxis, dissemination of hydatid

disease (secondary hydatosis) within the host, and bacterial infection of the pericyst cavity[5].

The most frequently involved organs are the liver (70%) and lung (20%)[6]. Beside systemic dissemination through the portal vein, the larvae also can disseminate through lymphatic channels to other organs. Other organs like muscle (5%), spleen (1%), bones (3%), kidneys (2%), heart (1%), pancreas (1%) and central nervous system (1%) are involved only rarely [2].

Most hydatid cysts are asymptomatic and are diagnosed incidentally. Ultrasonography (US) is particularly helpful in the detection of daughter cysts, internal membranes and hydatid sand. Computed tomography (CT) is important in evaluation of calcification and cyst infection. CT and magnetic resonance (MR) imaging may demonstrate cyst wall defects as well as the passage of contents through a defect[7]. MR imaging is especially helpful in detecting HCs of the central nervous system [8].

METHODOLOGY

This was a retrospective study of 50 patients conducted at Radiology department of Rehman Medical Institute, Peshawar. Duration of study was 18 months i.e., from May 2012 to November 2013. Approval from RMI medical research committee of ethics and research was taken. All the OPD and indoor patients referred to Radiology department with referrals from both surgeons and medical specialists and meeting the inclusion and exclusion criteria were approached. Mostly the patients had complaints due to pressure or compression effects of the hydatid cyst.

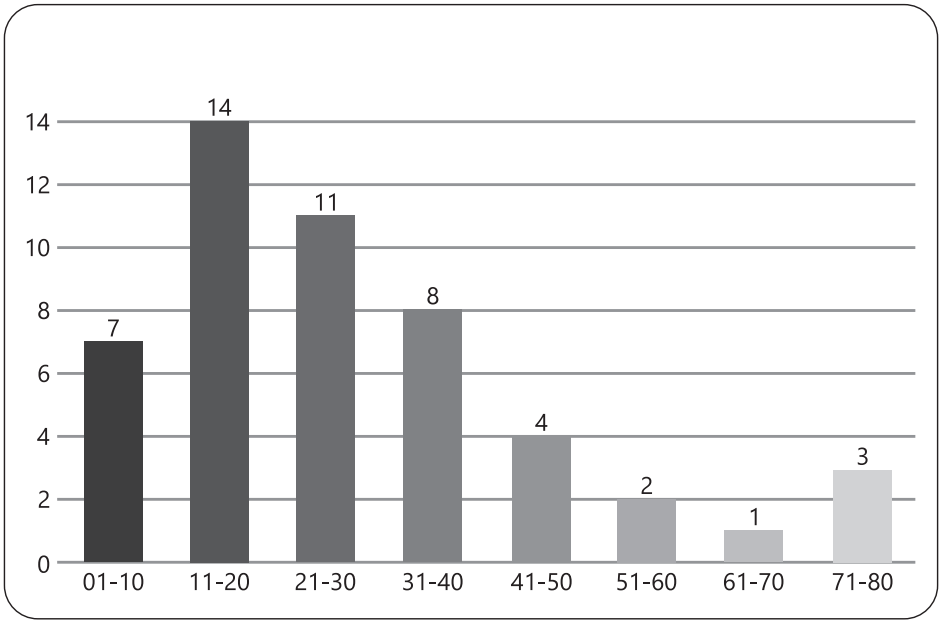
Multidetector Computed Tomography (MDCT) was performed on a 128 slice Toshiba scanner for chest, abdomen and pelvis disease whereas MRI was done on Airiselite Hitachi 0.3 tesla for nervous system involvement (brain and spine). Complimentary ultrasound was done when required. All 50 patients which were included in the study were proven cases of hydatid disease on the basis of Biopsy or classical imaging features pathognomonic of hydatid cysts and positive haemagglutination hydatid tests.

All data was entered and analyzed using SPSS version 10.0 and processed using Microsoft excel 2007. Frequencies (%) were calculated for age, gender and site of the lesion. Mean and standard deviation was calculated for age.

RESULTS

A total number of 50 patients had Hydatid cysts. Two patients had multiorgan involvement i.e., one had liver and lung involvement while other had liver and brain involvement. The mean age was 27 years with range of 8–80 years. Maximum numbers of patients were in the age group of 11-20 years followed by age groups of 21-30 and 31-40 years (Figure 1). 54% of the patients were female and 46% were male.

Figure 1: Age distribution of patients



The cysts were present in brain (n=3, 6%), spine (n=2, 3%), neck soft tissues (n=1, 1%), heart (n=2, 3%), ovary (n=3, 6%), kidney (n=1, 1%), spleen (n=3, 6%), peritoneal cavity (n=2, 4%) and pancreas (n=1, 1%). Liver was involved in 41% (20) cases while lung was involved in 28% (14) cases (Figure 2).

Figure 2: Frequency of visceral involvement

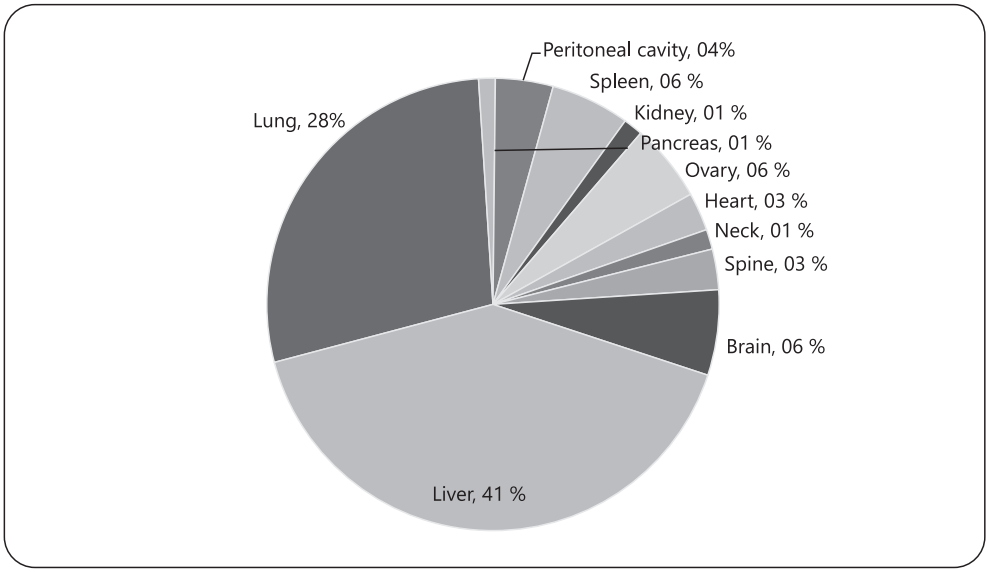
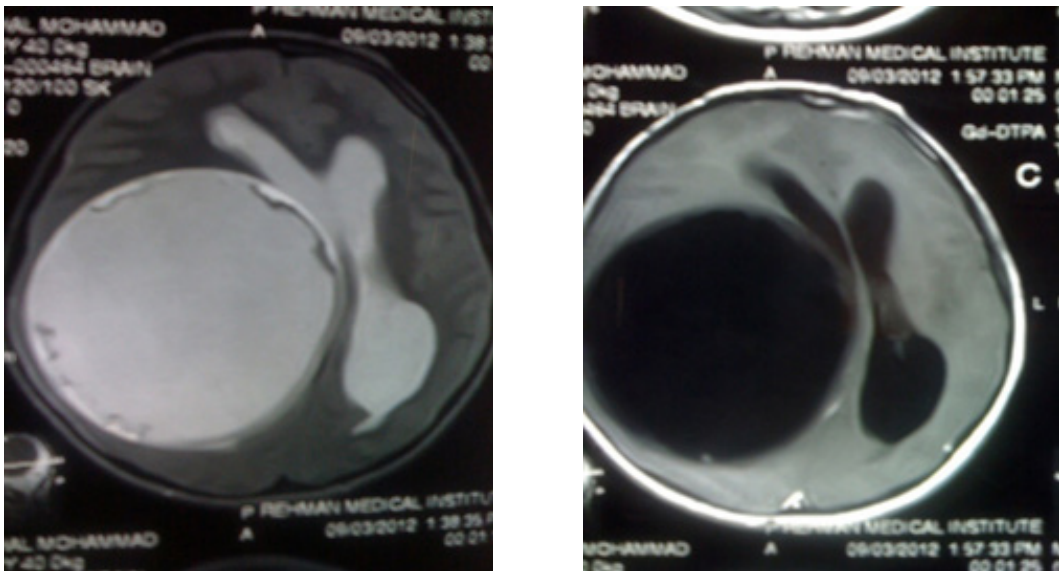
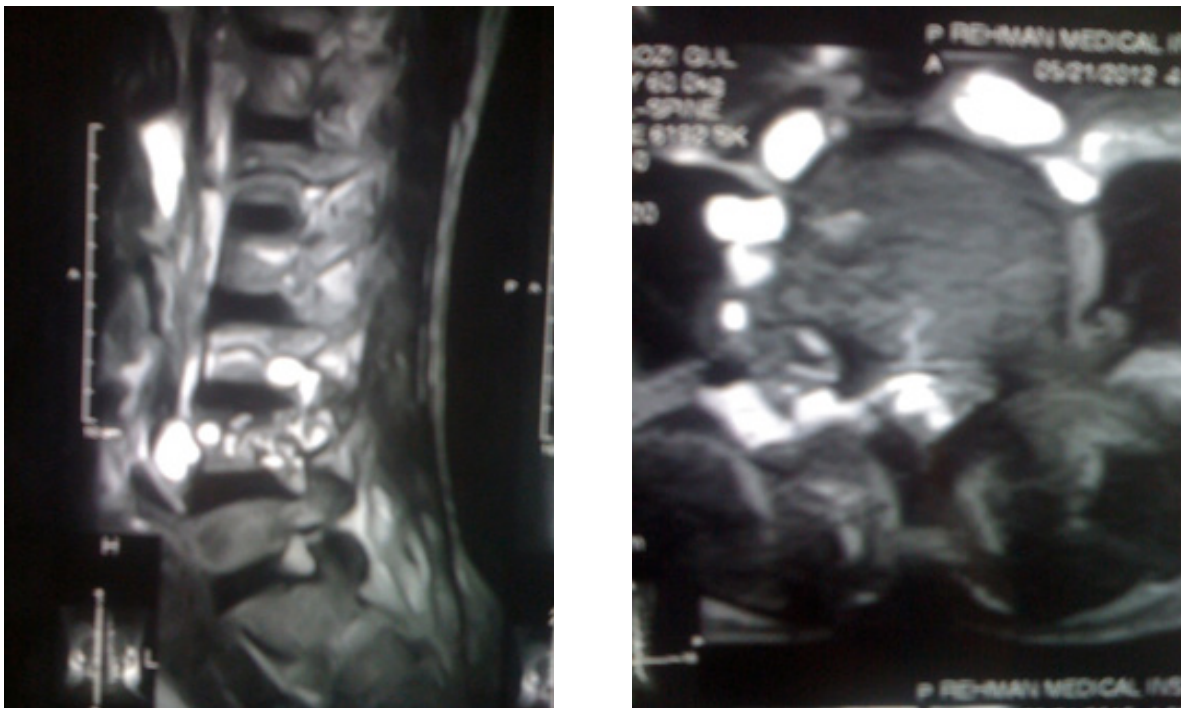


Figure 3: T2WI (a) and T1 axial post contrast (b) MRI images show large cystic lesion in right cerebral hemisphere compressing right lateral ventricle and midline shift towards left. Internal membranes are more evident peripherally on T2WI. There is no peri-lesional edema or post contrast enhancement



(a) **(b)**

Figure 4: Sagittal (a) and axial (b) T2WI of lumbar spine of a 35 year old female patient revealed a multiloculated cystic lesion in spinal canal at L3-4 level extending through right neural foramen into adjacent pre and paravertebral space



(a) **(b)**

Figure 5: Ultrasound neck of a 10 year old boy revealed a cystic lesion containing multiple daughter cysts

Figure 6a: CT chest with contrast of a 29 year old male revealed a large cystic lesion arising from interventricular septum bulging into both ventricular cavities.

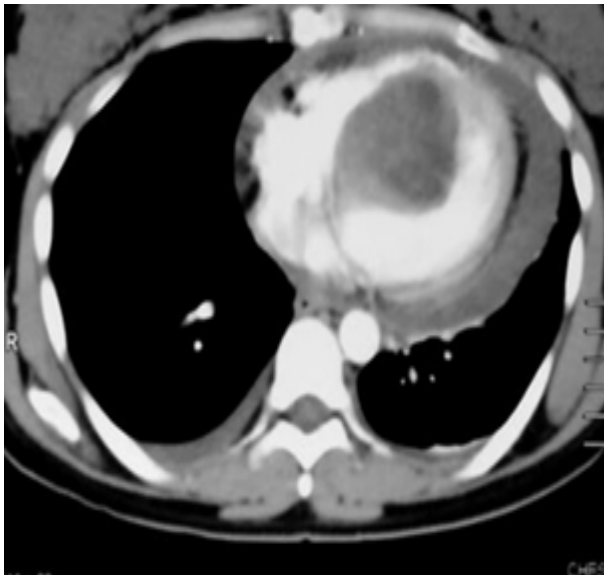
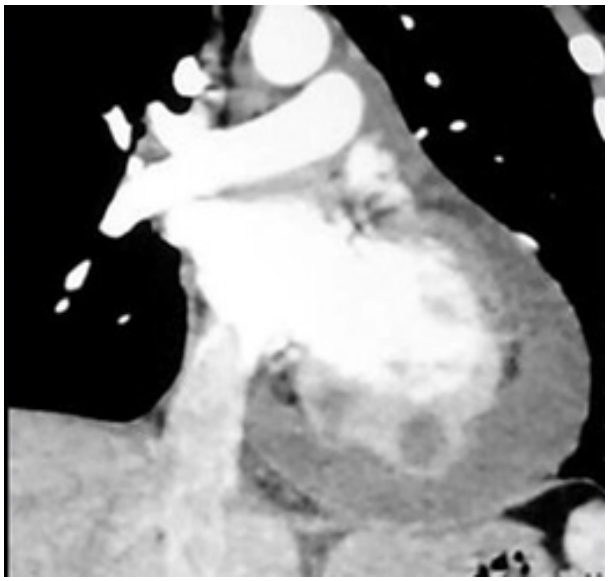


Figure 6b: Coronal view of same patient revealed cysts in left ventricular wall. There is moderate pericardial effusion suggesting rupture into pericardial cavity.



DISCUSSION

Brain

Only 1-2% of the hydatid cysts reach the brain after passing through the liver and the lungs [9, 10]. It accounts for only 2% of all intracranial masses, even in countries where this disease is endemic[3]. HCs are mostly located in the territories of the middle cerebral artery (MCA) but can be seen anywhere within the brain. Most cysts are supratentorial. The parietal lobe is most frequently involved. Cerebral HC is more common in children than in adults[11,12] . Multiple cerebral cysts can sometime occur as a result of rupture of primary cerebral cyst or embolization from a ruptured peripheral cyst [13,14,15].

CT and MR imaging demonstrate a well-defined oval or cystic mass with an attenuation or signal intensity similar to that of cerebrospinal fluid. There is no associated edema as is typically seen in abscesses and cystic tumors (Figure 3). The lesion does not enhance after intravenous administration of contrast material, and calcification is extremely rare[1]. However, when HCs are infected, the lesions show enhancement and differential diagnosis is sometimes difficult.

We had three patients with cerebral hydatid cysts. All of them were solitary with no perilesional edema or contrast enhancement. One was type 1 and two were type II lesions. All of them were located in the territory of right MCA. All of these patients underwent surgery and excisional histopathology confirmed hydatid disease.

Spinal Canal

Spinal HC accounts for less than 1% of all HC [3].The thoracic spine is most frequently involved (50% of cases), followed by the lumbar (20%), sacral (20%), and cervical (10%) spine [3,16].Spinal HC is classified into five groups: intramedullary, intradural extramedullary, extradural intraspinal, vertebral, and paravertebral. Imaging features are similar to as elsewhere in the body. Although CT efficiently demonstrates bony erosion and the extent of the lesion, MR can demonstrate any cord compression throughout the length of the spinal cord and is the investigation of choice [17]. Differential diagnosis may include TB, pyogenic infections, metastasis & benign bone tumors (fibrous dysplasia, enchondroma etc) [17].

We had two patients of biopsy proven spinal hydatid cyst, both in lumbar spine. They were extramedullary intradural with extradural extension through neural foramina (Figure 4). MRI had shown pure cystic signals i.e., hypointense on T1 and hyperintense on T2. Normal intervening discs and no post contrast enhancement were the features against TB spine.

Neck Soft tissues

Hydatid Cyst is seen considerably rarely in the region of the neck. Ozekinci et al [18]stated that only one hydatid cyst case out of 234 cases was diagnosed as located in neck region in Diyarbakir between 2002 and 2007. In the absence of typical imaging features (i.e daughter cysts, floating membranes), differentials include cystic

hygroma, cystic lymphangioma, cold abscess, chronic haematoma, dermoid / epidermal cyst etc.

In our study, only one case was seen with hydatid involving neck soft tissues. This was a 10 year old boy presented with lump in neck on left side. On palpation, it was smooth and non-tender. Ultrasound revealed a cystic lesion deep to left sternocleidomastoid muscle (Figure 5). It was partly involving the muscle with extension into anterior triangle of neck. It contained multiple daughter cysts (Type II). There was no flow on color doppler. FNAC revealed findings suggesting hydatid cyst which was later confirmed by excisional histopathology.

Heart

Echinococcus enters the heart via the coronary circulation, either via a patent foramen ovale or the pulmonary circulation[19]. Most common location is the free wall of left ventricle (50-77%) followed by interventricular septal wall and atria[20, 21, 22]. A variety of tumors in the heart and congenital pericardial cyst must be considered in the differential diagnosis; however, multivesicular nature of the cystic mass and membrane detachment indicate the true diagnosis. Transthoracic echocardiography is the most efficient investigation of choice.

We had two patients in which hydatid disease was involving only heart. They were initially diagnosed with echocardiography followed by CT chest with contrast. They revealed multivesicular cysts pathognomonic of hydatid associated with pericardial effusion (Figure 6). They were confirmed by hydatid haemagglutination test and FNAC.

CONCLUSION

Hydatid disease may occur anywhere from the big toe to the crown of the head wherever the bloodstream reaches. Imaging features such as floating membranes and daughter cysts are highly suggestive for hydatid disease. Despite the characteristic imaging findings, hydatid disease in unusual anatomic locations like heart, neck, spine and brain may make differential diagnosis difficult. Hydatid cyst should be kept in mind when a cystic lesion is encountered anywhere in the body especially in patients from endemic regions.

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CONTRIBUTORS

SA planned the study, did data analysis and wrote the manuscript. US, SG, SG, BF and FG helped in data collection and manuscript writing. All authors contributed significantly to the final manuscript.