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Case Report

Primary Hydatid Cysts at Unusual Sites and Role of Serology in Diagnosis: Case Series and Review of Literature

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Abstract

The Cestodes, Echinococcus granulosusand Echinococcus multilocularis are the causative agents of hydatid disease. Dog is the definitive host while humans are accidental intermediate hosts. The liver and lungs are commonly affected though other exotic sites can also be uncommonly affected. This is of great clinical relevance because of the risk of anaphylaxis during inadvertent surgical exploration or other invasive procedures. Imaging and hydatid serology (Indirect Hemagglutination, Immunoelectrophoresis, ELISA and Western Blot) are the key to pre-operative diagnosis. We present a series of hydatid cysts presenting at unusual sites including kidney, parotid, and ovary and limb muscle. Hydatid cyst should be suspected in the differential diagnosis of any cystic lesion particularly in endemic areas.

Keywords: Echinococcus, hydatid serology, ELISA, Western Blot.

INTRODUCTION

Hydatidosis or Cystic Echinococcosis (CE) is a cyclo-zoonoses caused by Echinococcus species[1]. CE is endemic in the Middle East, Africa and South America. Dog is the definitive host while humans are accidental intermediate hosts. It most frequently affects the liver (50-77%) and lungs (18-35%) followed by abdominal cavity and brain[2].Other organs are uncommonly affected posing diagnostic difficulties. CE at these exotic locations is usually secondary to dissemination from another organ and primary involvement is rare. CE is often associated with characteristic imaging findings and hydatid serology is usually considered as corroborative evidence. However, when imaging is non-diagnostic, serology can be a key pointer to the presence of CE. This is important because of the risk of anaphylaxis with invasive procedures including unprepared surgical exploration. We present a series of four patients with primary CE occurring at unusual sitesand review the role of hydatid serology in their prompt diagnosis and further management.

Case Series

We present 4 patients who presented with CE at exotic locations namely ovary, kidney, parotid gland and skeletal muscleto a Tertiary Hospital in Kolkata, India (Table 1 and Figures 1-4). Three patients (75%) presented with history of mass like swelling. Hemogram and routine blood biochemistry was normal except for mild eosinophilia in two patients. Imaging revealed simple cyst like lesion (consistent with WHO CE1 or Gharbi Type 1) in all the patients except the lady with ovarian CE. As such imaging was not diagnostic of CE in any of the patients. Surgical exploration with cyst excision was done in the patients in view of symptoms related to mass effect of the cyst. Histopathology confirmed the diagnosis of CE. Fortunately, none of the patients suffered from any perioperative complications including anaphylaxis. Hydatid serology was not done pre-operatively in our patients as it is not routinely available at our Institute and is not part of our usual protocol for evaluation of non-hepatic cystic lesions. Nonetheless, hydatid serology (IgG ELISA) was retrospectivelypositive in three patients (75%) in the immediate post-operative period. All patients were put on albendazole therapy after diagnosis and are doing well on follow-up.

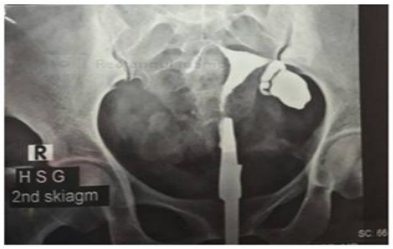

Fig-1A-Bilateral Tubal Block with Left Hydrosalpinx

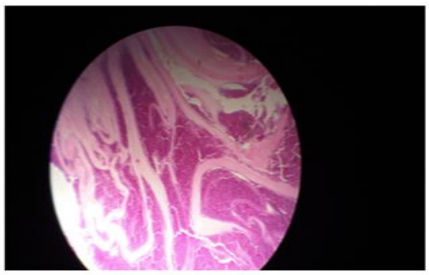

Fig-1: B-Ovarian Hydatid Cyst with Germinal Layer and Ectocyst

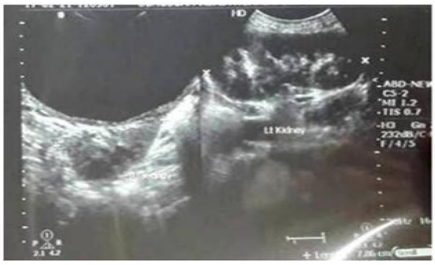

Fig-2: A-USG –Left Renal Cyst

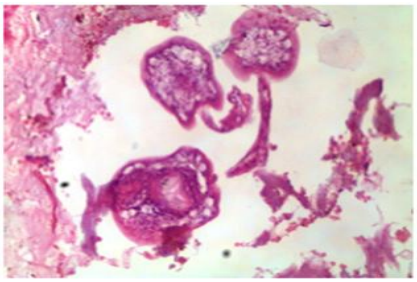

Fig-2: B-Microscopy of Hydatid Scolices of Left Kidney

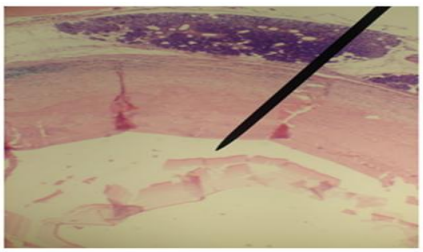

Fig-3: A- Gross Photograph of Cystic Lesion of Parotid Gland

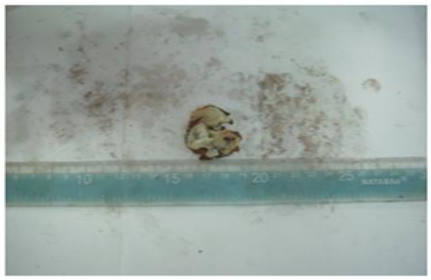

Fig. 3B-Microspic Picture of Parotid Hydatid Cyst

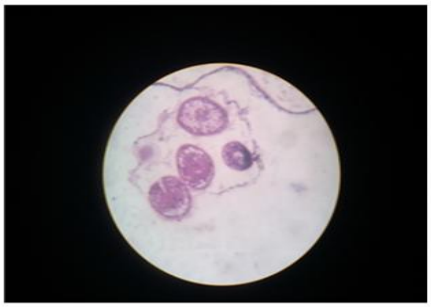

Fig-4: Microscopy of Muscle Hydatid Cyst

Table: Detail of Cases of Primary Hydatid Cyst at Unusual Sites:

Details	Case 1	Case 2	Case 3	Case 4
Age(years)	30	60	5	35
Sex	Female	Male	Female	Male
Clinical Feature	• Mild tenderness left iliac fossa • No definite palpable mass	• Low Back Pain • No hematuria • No lower urinary tract infection • Tender left lumbar mass	Parotid swelling Soft cystic mobile non-tender mass	Gradually enlarging, painless right thigh swelling for eight months. No history of fever No weight loss No trauma **Clinical examination** A soft,non-tender deep-seated mass over right thigh measuring 4.5×4 cm^2
Imaging	Hysteroslpingography Bilateral tubal block with left hydrosalpinx CT SCAN Multiple nodules left cornu encroaching ovarian surface	Contrast-enhanced CT scan – Left renal cyst (5.5×4.5×1.5 cm^3) Thin smooth wall compressing renal pelvis Hydronephrosis No enhancement of cyst contents	Ultrasound examination - A simple cyst of 4.5 cm diameter. Fine-needle aspiration cytology showed scattered, benign epithelial cells in a clear background	X-ray : A large soft tissue mass over right thigh with intact underlying bone
Other Relevant Investigation	• Routine Blood Examination • Tumour markers • All within normal limit	• Routine Examination-Within normal limit • Slight Eosinophilia	Fine-needle aspiration cytology - scattered, benign epithelial cells in a clear background Routine Examination-Within Normal Limit	Routine laboratory investigation revealed eosinophilia (AEC 610/mm^3)
Operative Finding and Gross Pathology	**Laparotomy**- • 3×2 cm^2 calcified mass near cornu with left hydrosalpinx • 5-6 nodules on lateral wall of uterus.	**Laparotomy** Cyst extending from renal hilum towards lower pole resulting hydronephrosis. **Gross Pathology**: Encapsulating membrane	**Gross Pathology** A white translucent, unilocular, fluid filled cyst measuring 4.5×4 cm^2	**Laparotomy** A 5×4 cm^2 cystic lesion with clear content and few scolices

	Gross Pathology: Multiple globular tissue pieces with intact outer capsule **Cut Section**: Extrusion of whitish acellular material on cut-section	along with resection of neighbouring tissue		
Histopathology Examination	Ectocyst,Endocyst with scolices pf Hydatid cyst	Histopathology confirmed the diagnosis of Hydatidosis.	Histopathology lamellated ectocyst with few scolices and compressed salivary acini	Histopathology revealed Echinococcosis
Hydatid Serology	Retrospectively- Hydatid serology in patient serum by ELISA (IgG4) was positive.	Immediate postoperative : Hydatid serology by ELISA(IgG4) positive	Immediate post excision : Hydatid serology in serumELISA (IgG4) - positive	Hydatid serology by ELISA from patient serum was negative in immediate postoperative period.

DISCUSSION

Hydatid disease or CE is a zoonotic disease caused by the larval stage of Echinococcus species. It is a systemic zoonosis with high prevalence in the Mediterranean region and Middle-East countries. In India, the annual incidence is estimated to vary from 1-200 per 100,000 population[3]. Liver and lungs are the commonest sites of involvement. Presentation of hydatid disease as cystic swelling of ovary, kidney, parotid and muscle is rare even in endemic areas with a reported incidence of 2.5%,3%,1.5% and 0.5%, respectively[4-7]. Occurrence of disease at such exotic locations is usually secondary to dissemination from other primary sites. We present a series of four hydatid cyst cases with primary involvement of such unusual sites diagnosed on histology.

Primary hydatid disease of ovary is extremely rare.In most of the cases, ovarian involvement is secondary to dissemination from another primary site. Diagnostic difficulty is common because of non-specific symptoms and inconclusive imaging findings leading to suspicion of polycystic disease or cystic neoplasm. While Cattorini *et al*. reported secondary cases of pelvic hydatid disease, Ray *et al*. documented a case of huge primary hydatid cyst in pouch of Douglas[8,9]. Our case was challenging because of its unusual presentation,no definite palpable mass and lack of involvement of other common sites. Anthelminticsare useful for asymptomatic cysts but for large, symptomatic cysts, complete surgical resection followed by confirmatory histopathology examination is the only definite management to avoid complications[10].

Mongha *et al*. reported primary hydatid cyst of kidney and ureter with gross hydaturia[11].The route of entry of primary hydatid disease to the kidney is debated but it seems that it passes through the liver and retro-peritoneal lymph nodes[11]. Abdominal X-rays provide limited information.Ultrasonography is more supportive and may demonstrate hydatid daughter cysts and hydatid sand.In general, surgery is the treatment of choice in renal hydatid cyst. Laparoscopic surgery should be avoided because of chance of rupture and spillage. Our case presented with diagnostic difficulties because of the vague clinical presentation and imaging suggestive of simple cyst. Cystectomy was mandated in view of hydronephrosis secondary to compression by the cyst. Subsequent histopathology examination was confirmatory.

Primary parotid gland hydatid cyst may have myriad presentations and should be considered in the differential diagnosis of any head and neck cystic swelling[6]. Presentation of a cystic swelling in pre-auricular region of a 5-year old girl with inconclusive imaging posed a diagnostic conundrum. FNAC was suggestive of inflammatory or developmental cystic lesion. Subsequent resection as well as histopathology examination led to definitive diagnosis of hydatid cyst.

Muscular involvement is another rare site of primary involvement, but it may also occur secondarily when cysts spread from other areas spontaneously or by iatrogenic implantation. Preoperative diagnosis can be made by radiological demonstration of daughter cyst[12]. Though the definitive management is surgical resection, addition of anthelminticsis preferred to cover risk of dissemination.

Serology generally has an adjunctive role in the diagnosis of CE which is usually made on imaging. Several characteristic imaging findings of CE have been described on ultrasonography including double line sign, honey-combing, detached membranes, daughter cysts and calcification. However, when imaging findings are non-classical or when cysts occur at unusual sites, positive hydatid serology can be an important clue to the presence of CE.Varios techniques including Indirect Hemagglutination, Immunoelectrophoresis, ELISA and Western Blot have been described for the serologic diagnosis of CE. Advanced serological techniques like ELISA and Western blot permit the pre-operative diagnosis of Hydatid disease with a sensitivity and specificity of 86% and 97% respectively[13]. This may help reduce the risk of anaphylaxis due to inadvertent spillage during surgical exploration. However, hydatid serology may be false negative in 15-50% of patients with extra-hepatic CE. Performance of hydatid serology also depends on the stage of the disease (as defined by WHO). Nonetheless, it does not have adequate sensitivity or specificity to reliably differentiate

between active, transitional and inactive CE. Another vexing problem is the possibility of positive serological results despite adequate treatment thereby limiting its utility in the follow-up of CE patients. Testing for specific IgG subclasses may give superior yields. Evidence suggests that IgG2 and IgG4 may the best markers for diagnosis and IgG4 may also be useful in follow-up.

Antigen 5 and Antigen B are the most commonly studies targets for diagnostic purposes. Antibody detection is usually preferred to the detection of antigens as the sensitivity of the latter is unsatisfactory (35-80%) and is influenced by the location and activity of the cyst. Antigenic detection may have some relevance in the post-treatment follow-up of patients because of the tendency of antibodies to persist despite treatment. However, even in this aspect the usefulness of antigen detection is limited by its low sensitivity.

Hydatid serology is not routinely available at our institute and was not performed till the diagnosis of CE was suggested on histology. Fortunately, none of our patients suffered from any peri or post-operative complications. Inadvertent spillage of hydatid fluid during invasive procedures or surgery can lead to life threatening anaphylaxis. Surgery is generally advocated in presence of active or transitional cysts that are not amenable to locoregional therapy like PAIR or large inactive cysts causinmg compressive symptoms. Pre-operative knowledge of the diagnosis allows the surgeon to take extra care while draining the cyst fluid. Infusion of scolicidal agent and its aspiration can then be carried out before cystectomy is completed. Moreover, patients can be put on pre-operative Albendazole when CE is strongly suspected.IgG hydatid serology using ELISA was positive in three of our patients (75%), retrospectively. Thus, routine hydatid serology for cystic lesions irrespective of their location or imaging findingsmay help in the diagnosis of CE, particularly in endemic areas. Prior knowledge of CE helps optimise management when surgery is required and may be vital for avoiding therapeutic misadventures.

CONCLUSION

Unusual locations of hydatid cyst can pose a diagnostic conundrum. Knowledge of these myriad presentations is required to avoid inadvertent spillage of contents during invasive procedures with potentially disastrous consequences. Hydatid serology, as corroborative evidence, may aid in clinical decision making and providing appropriate treatment procedure.

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