# Hydatid Disease at Atypical Sites: A 5 Year Experience in a Tertiary Care Centre

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**ABSTRACT**

**Introduction:** Hydatid disease is a zoonosis caused by the larval stage of Echinococcus granulosus. It is prevalent in areas where livestock is raised in association with dogs. Humans are the accidental intermediate host. This study was done to study the clinicopathological pattern of hydatid diseases presenting at unusual locations

**Material and methods**: One hundred and forty- four patients with hydatid cysts managed surgically in Department of Surgery, Government Medical College, Srinagar during our study period of five years were evaluated retrospectively. Only those patients with hydatid disease at atypical locations (n=8) were included in the study, including the five patients with concomitant disease of the liver.

**Results:** Liver was involved in 106 patients, of which 87 (81.30%) had isolated liver disease, 15 (14.02%) had associated lung cysts and another 5 (4.67%) patients had co-existant hydatid disease at atypical locations.3 patients had primary isolated hydatid cysts involving atypical locations.

**Conclusion:** Since hydatid cyst can be found in all the body sites, it should be taken into account in the differential diagnosis of all cystic lesions especially in endemic regions.

**Keywords:** Hydatid Cyst, Echinococcus Granulosus, Atypical, Zoonotic, Livestock

## INTRODUCTION

Hydatid disease caused by the larval stage of Echinococcus granulosus (EG), is considered one of the most important zoonotic helminthic diseases throughout the world. It is common in sheep raising areas like Australia, New Zealand, China, South America, Middle East, African countries around the Mediterranean and in India.[1–7]

It is commonly found in rural areas where domestic livestock-raising is common causing serious health problems. Dogs or other carnivores are considered as definitive hosts, while sheep or other ruminants, are intermediate hosts. Humans are accidental hosts, usually infected by ingestion of food or water contaminated with tapeworm eggs or by direct contact with host.[8] The eggs hatch after digestion of the outer capsule in the intestinal mucosa and the larvae penetrate the mucosa, reaching the liver through the portal vein. Most of these embryos become lodged in the hepatic sinusoids, where they either die or develop into one or several hydatid cysts.[9,10] A systemic spread will occur if the larva passes through the capillary sieve. The most frequent locations of the disease are the liver (50–77%) and lungs (18–35%), followed by the abdominal cavity and brain.[10] In abdominal cavity, splenic hydatosis is primarily seen in 1.5–3.5% cases.[10] Similarly in brain, orbital involvement is seen in < 1% of cases affected by this larval tapeworm.[11] Hydatid cyst at unusual sites have been reported around the world including the spleen, kidney, heart, bones, muscles and cranium, but soft tissue hydatid disease represents less than 3% of all hydatid disease.[12]

Majority of rural population in Kashmir Valley, Jammu and Kashmir State in North India is dependent on agriculture and livestock grazing (sheep and cattle) as their primary source of income. Besides this there is a huge population of stray dogs in Kashmir valley. The slaughtering of livestock without veterinary control, the widespread rural practice of feeding dogs with the viscera of sheep slaughtered at home is a common practice. All these factors are highly favorable for transmission of echinococcosis in this valley.

## MATERIAL AND METHODS

The present hospital based descriptive study was done within a period of five years (Feb 2015- Jan 2020) in the Department of Pathology, Government Medical College, Srinagar. One hundred and forty- four patients with hydatid cysts managed surgically in Department of Surgery, Government Medical College, Srinagar during this study period were evaluated retrospectively. Of these patients 80 (55.55%) were males and 64 (44.4%) females. Age of the patients ranged from 15 to 65 years. Liver was involved in 107 patients, of which 87 (81.30%) had isolated liver disease, 15 (14.02%) had associated lung cysts and another 5 (4.67%) patients had coexistant hydatid disease at atypical locations. Three patients had primary isolated hydatid cysts involving atypical locations. Only those patients with hydatid disease at atypical locations (n=8) were included in the study, including the five patients with concomitant disease of the liver. Patient's clinical data, age, sex, residence, location of the lesion, complete blood counts, liver and kidney function tests, serological tests (enzyme linked immunosorbent assay and indirect hemagglutination test),and details of imaging

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techniques [X-ray chest, ultrasound and magnetic resonance imaging (MRI) or contrast enhanced computerized tomography (CECT)] were obtained in all cases. The treatment for all the patients was surgical. All the patients received preoperative and postoperative albendazole chemotherapy.

## RESULTS

Eight patients were diagnosed and confirmed to have hydatid disease at rare sites. The mean age of the patients was 34 (range: 15 - 55) years. Of these 5 were males and 3 were females giving a male to female ratio of 1.67:1. The mean duration of symptoms was 15 months (range: 6 months – 3.5years). Among this study group, primary isolated hydatid cysts involving atypical locations were present in 3 [37.5%] patients and another 5 [62.5%] patients had coexistant hydatid disease in liver. Atypical locations included were ovary 4 (50%), one case each in spleen, kidney, breast and retroperitoneum [**Table 1**].

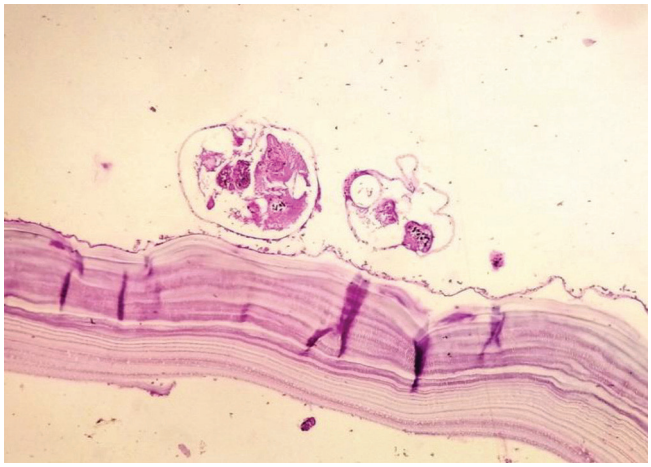

**Figure-1:** 20X showing inner germinal layer and outer laminated cyst wall

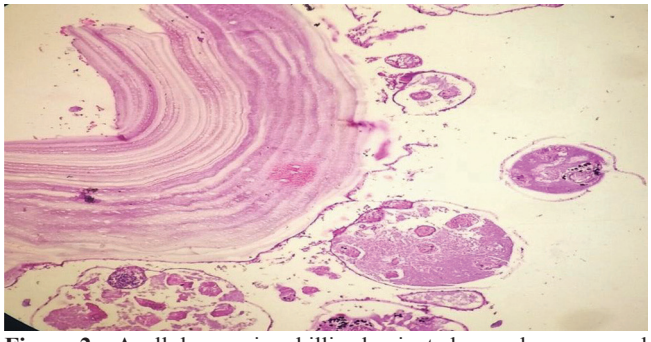

**Figure-2:** Acellular eosinophillic laminated membranous and scoleces [H&E 20X]

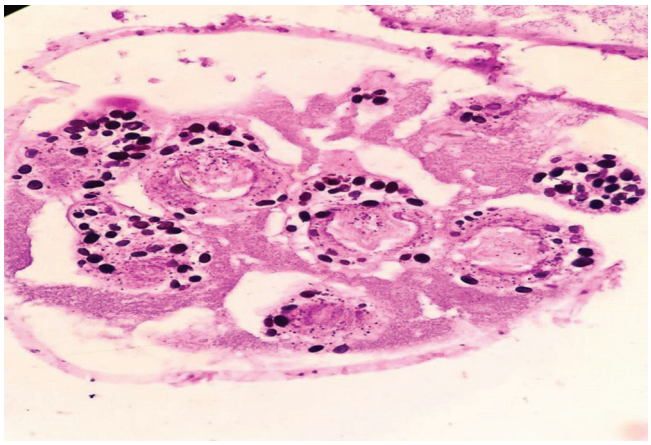

**Figure-3:** Brood capsule [H&E 40X]

Nonspecific abdominal pain and a non tender palpable abdominal lump were the most predominant symptoms; other symptoms varied according to the location of the cyst. Surgical treatments include complete cyst excision (cystopericystectomy) in most of the patients. Partial nephrectomy, ovarian cystectomy, lumpectomy and splenectomy, were performed whenever cysts invaded these organs. Eosinophilia was observed in 6 out of 8 patients. Diagnosis was confirmed by hydatid serology, ultrasonography, MRI/CECT, and histopathological examination of the specimen. In the histopathological examination of all cases, we have searched structures in conjunction with HC to be able to come up with a definite diagnosis.

## DISCUSSION

Hydatid disease may affect humans, especially in endemic areas, where livestock raising is a common practice such as Mediterranean countries, the Middle East, South America, New Zealand, Australia, and Southeast Asia and many parts of China.[13,14,15] The disease is attributed to occupational exposure during farming practices (sheep rearing), particularly in rural areas; consumption of egg-contaminated water and vegetable, and traditional consumption of beef and mutton. Because the practice of close contact of humans with domestic animals such as dogs is not much prevalent in Kashmir, being a Muslim dominated place, the sheep-dog association apparently seems to be most commonly implicated in the life cycle of the parasite in our valley.

Hydatid cyst at atypical sites have been reported worldwide which include the spleen, kidney, heart, bones, muscles and

| Cyst location | No. of patients | %age | Symptoms | Surgical treatment |
|---|---|---|---|---|
| Ovary | 4 | 50 | Mass abdomen, pain lower abdomen | Ovarian cystectomy |
| Spleen | 1 | 12.5% | Left upper quadrant pain,abdominal lump | Splenectomy |
| Kidney | 1 | 12.5% | Abdominal lump | Partial nephrectomy |
| Breast | 1 | 12.5% | Breast lump, pain | Lumpectomy |
| Retroperitoneum | 1 | 12.5% | Retroperitoneal mass, pain | Cystopericystectomy |

**Table-1:** Distribution of cases based on atypical sites of involvement

cranium, but soft tissue hydatid disease represents less than 3% of all hydatid disease.[12] Although portal blood stream remains the main pathway of parasite spread, systemic dissemination through lymphatic route, and retrograde migration of parasite from vena cava to subclavian vein have also been documented and explain some of these unusual presentation sites. In our study, we presented atypical localization of hydatid cysts from a single tertiary care institute.

Localization of hydatid disease at ununsual sites usually poses a serious diagnostic challenge. Hydatid disease in extra-hepatic locations usually follows a silent clinical course unless it grows and produces pressure symptoms or develops complications which may include local pressure, rupture, secondary infection, and an allergic reaction.[16] Clinical signs and symptoms are nonspecific and never pathognomic of hydatid cyst.[17] In our study percentage of cases located at atypical sites amounts to 5.55%. In the literature, the rate of organ involvement other than lung or liver was reported between 5.7-13.9%[18,19] and our result was similar (5.55%). Almost all of these cases are associated with concomitant existence of cysts in primary organs like lung and liver, but in our series all except three patients had isolated primary location of hydatid disease at an atypical site.

The ovary is a rare primary target organ for hydatid disease. Owing to its multilocular cystic appearance, a hydatid cyst may not be differentiated from ovarian lesions with septal structures such as cystadenoma or ovarian cystic teratoma (with intracystic floating globules).[20] In our study we encountered four cases of ovarian hydatidosis. Among them, majority [3 cases] had a preoperative diagnosis of ovarian cystic neoplasm for which cystectomy was done.

Routine laboratory tests can only reveal eosinophilia. Diagnosis is established by imaging, serological tests and histopathological examination. USG is a cost effective imaging technique but when available, CT scan is superior owing to its higher sensitivity (21). Serologic tests are also very useful and usually involve a screening test such as ELISA or Indirect hemagglutination assay. Sensitivity of serology is high (80-100%) for liver cysts, but low for lung (50-56%) and other organs (25-56%).[22] The diagnosis of hydatid cyst is confirmed by histology.[23] Histopathological examination reveals three layers of hydatid cyst-pericyst, ectocyst and endocyst/germinal layer **[Fig1 and 2**]. The inner most germinal layer, which is thin and translucent on gross examination, consists of daughter cysts and brood capsules with scolices which develop from an outpouching of this layer **[Fig3].** The cyst fluid is crystal clear, as it is transudate of serum containing proteins and is therefore antigenic. The middle ectocyst comprises of a hyaline and acellular laminated white membrane, 2mm thick and is easily ruptured. It is selectively permeable to nutrients but not to bacteria. The outer layer or pericyst is fibrous, a few millimeters thick, representing response of the host to the parasite.[24]

Medical treatment alone may be effective in 30–40% of cases. Gold standard treatment is the total excision of the cyst, avoiding its rupture and spillage to prevent any spread of hydatid with subsequent secondary echinococcosis; anaphylactic shock is also a major risk connected.

## CONCLUSION

From our study we tried to establish that hydatid cyst can be found in all parts of the body and this should always be considerd in the differential diagnosis of cystic lesions or unidentified tumor formations, especially, in patients from endemic areas. Postoperative long term follow-up is also essential to detect any late complications such as local recurrence of the disease and development of hydatid cyst at the usual primary sites.

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