

Case study

Multiple Schwannomas masquerading as a Disseminated Hydatid disease in a Female- A Diagnostic Dilemma- A case report

ABSTRACT:

Hydatid disease or Cystic echinococcosis (CE), is caused by infestation with larva of *Echinococcus granulosus*, a ~2–7 mm long tapeworm found in dogs (definitive host) cattles and pigs (intermediate host) [1]. Disseminated abdominal hydatidosis, is a rare presentation of hydatid disease [3]. Secondary peritoneal disease is a consequence of either traumatic or iatrogenic rupture, as in case, of a primary hepatic or splenic cyst [2].

Schwannomas are neurogenic tumours originating from many different organs throughout the body, but rarely in the gastrointestinal tract [8]. Schwannomas are usually benign, only occasionally malignant [11]. Unlike most of the tumours, making pathologic differential diagnosis of schwannoma from other benign conditions is an exceedingly difficult endeavour preoperatively. Confirmation of diagnosis of schwannoma is by pathological and immunohistochemical examination of resected surgical specimens.

Case presentation:

We report a 40-year-old woman who was initially diagnosed clinically and radiologically as case of multiple hydatid cysts with cholelithiasis which after surgery was found to be cholelithiasis with multiple schwannomas treated successfully with complete excision. Individually both are themselves an uncommon entity separately. To the best of our knowledge Multiple intra-abdominal Schwannomas mimicking as disseminated hydatid disease has never been reported in literature. We herein present such a case.

Conclusion: Hydatid disease needs be kept in the differential diagnosis of all cystic masses in all anatomic locations, more so in areas where the disease is considered endemic. The ideal treatment is the complete excision of the cyst without any spillage undercover of anti-helminthic. But concomitantly high index of suspicion must also be kept for other cystic or solid lesions in cases of unusual presentations or atypical findings on imaging studies and

histopathological studies must be undertaken to confirm the diagnosis, as in this case which was found to be multiple Schwannomas.

Key words: - Hydatid, cyst, schwannoma, neurilemmoma

BACKGROUND:

Hydatid disease (Greek hudatis =watery vesicle) is a chronic helminthic disease caused by metacestode Tape worm *Echinococcus*. Three observed forms of human echinococcosis: a) Cystic hydatidosis caused by *E. granulosus*, (most common) b) Alveolar hydatidosis caused by *E. multilocularis* (invasive), c) Polycystic echinococcosis by *E. vogeli* [1]. *E. granulosus* infections usually present as solitary cysts and single organ involvement with frequency of Liver (70%), Lungs (20%), uncommonly spleen (6%), heart (2%), kidney (2%) and brain (<2%). 10-15% of cases can have two organ involvements depending upon specific geographic region and parasite strain. Disseminated abdominal hydatid disease is therefore a rarity [3].

Schwannomas are solitary, soft, lobulated, well encapsulated tumours attached to or surrounded by nerve, but nerve palsy is uncommon [8]. They are ectodermal in origin and multiple lesions are rare. Vagus nerve is one of the most common peripheral site, whereas vestibular nerve is the overall commonest site [9]. Diagnosis needs imaging modalities like tomographic (CT) scan or MRI to differentiate it from other tumours, but the final diagnosis is confirmed by pathological, immunohistochemical examination of resected specimens. Being encapsulated, enucleation is possible without sacrificing the nerve.

CASE PRESENTATION

A 40-year-old village female presented in surgery OPD with complaints of chronic, diffuse, mild to moderate, dull aching type, non-radiating pain abdomen, which increased with activity & relieved with self-medications and rest. A sudden onset hematuria of 3 days prior to admission compelled patient to come immediately.

Barring a hysterectomy done ~2.5yrs back at a government setup with consequent menopause, patient has had no previous surgical history. She had no history of any chronic illnesses in the past.

Clinical examination revealed pallor. Per abdominal examination revealed well healed old infraumbilical surgical scar mark in supra pubic area. Diffuse tenderness in supra pubic area with clinically palpable tender lumps of size ~1x1.5cm & 1.5x 1.5cm in parietes were noticed.

ULTRASOUND ABDOMEN (USG) reported cholelithiasis, hemangiomas of liver, disseminated hydatid disease. Also note of clot in the urinary bladder.

Contrast enhanced scan (CECT) ABDOMEN reported multiple round to oval enhancing lesions retroperitoneal area with multiple internal septations at right paramedian L1 vertebral level ,4.5x4x3.6 cm causing mass effect & lateral displacement of vena cava, renal vein & abdominal aorta in posteromedial aspect. (Fig.no.:1)

Another round to oval enhancing lesions at left paramedian 1st lumbar vertebral level 4.9x4.2x3.9 cm left suprarenal area displacing left adrenal laterally abutting left renal artery kidney & pancreatic tail.

Another round to oval enhancing lesions lesion 3.9x2.2x1.8cm seen in pelvic cavity, anterior to urinary bladder abutting the anterior urinary bladder wall. Left anterior abdominal wall in subcutaneous plane of size 2.3x1.1cm has round to oval enhancing lesions.

Visualized basal segments of lung show few nodules in tree in bud pattern (likely infective).

Liver showed 6x5mm hypodense lesion (benign) in VI segment. Cholelithiasis with Common Bile Duct 7.2mm size.

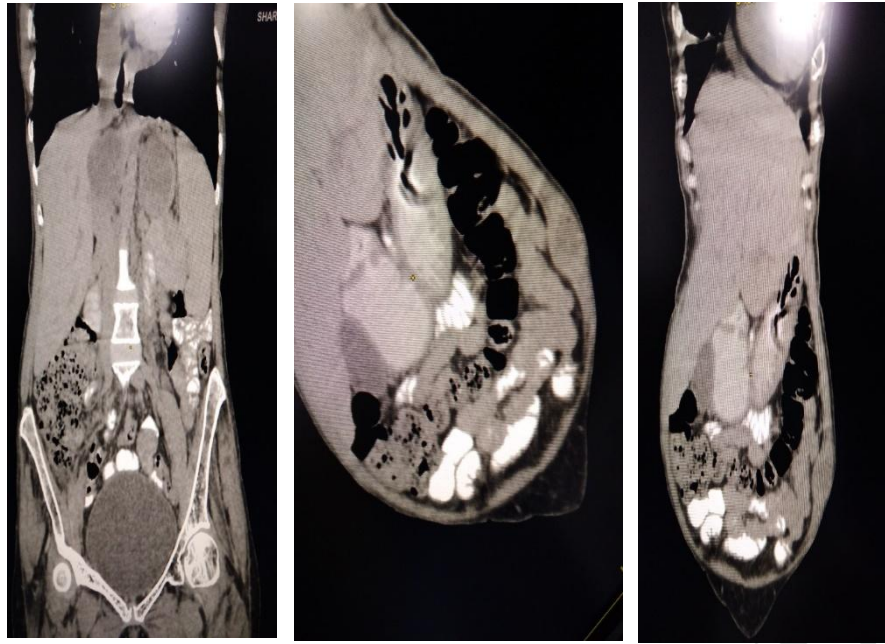


Figure 1: Pre-operative CECT image showing cysts: b) 2 around IVC, (c) & (d) abdominal wall

LAB. REPORTS have Hemoglobin-10.2gm%, Total Count-5120 /dl, Differential Count -Polymorphs 66, Lymphocytes 20, Eosinophils 4, Monocytes 10, Basophils-nil, ESR- 40mm in 1st hour, Absolute Eosinophil Count-204/cumm, HbA1C- 5%, urine- full field Red Blood Cells , rest within normal limits.

USG & CECT abdomen were suggestive of disseminated hydatid disease.

She was started on Albendazole dose of 10 mg/kg/day. Hematuria responded to conservative treatment fully with routine examination after 4 days.

Patient was discharged with Albendazole and other medications with advice to review after 3 weeks for definitive surgical treatment.

RESULT: -

PRE-OP:

After 3 weeks of Albendazole medication, of suspected disseminated hydatidosis, patient was readmitted for revaluations.

USG ABDOMEN showed cholelithiasis, liver hemangiomas, disseminated hydatid disease.

Since the repeat USG did not reveal resolution or regression of existing lesions, the patient after pre anesthetic work up was taken up for surgery.

INTRA OP

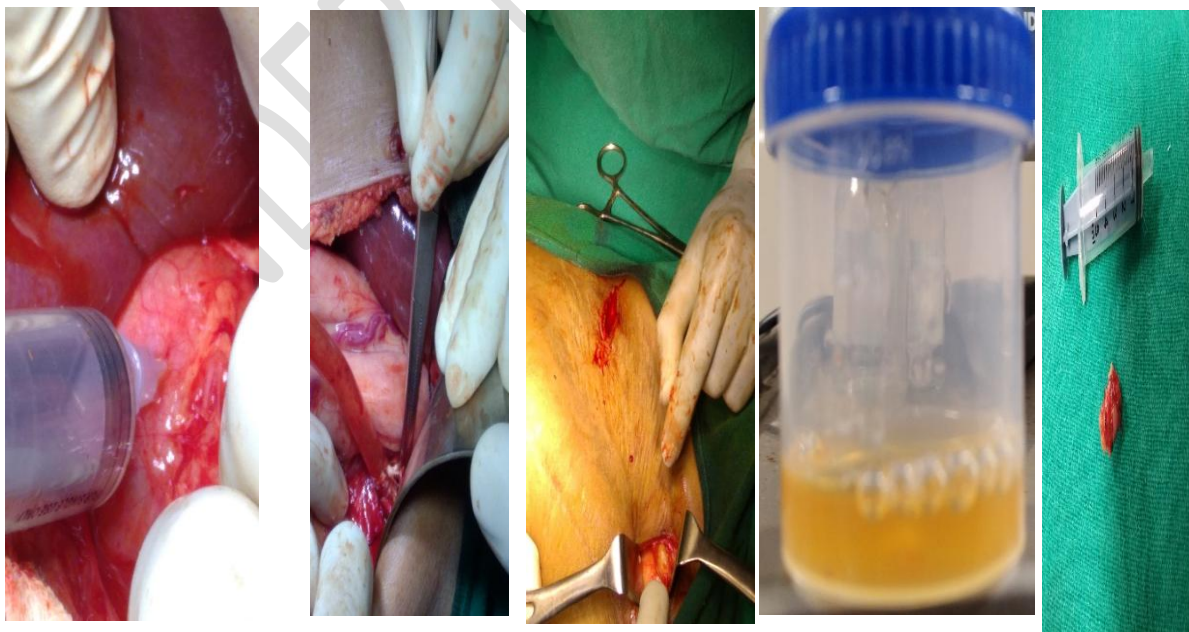
Under general anaesthesia, a diagnostic laparoscopy followed by an uneventful laparoscopic cholecystectomy was completed. However, abnormal thick fixed appearance of pericaval lesion the procedure was converted to a formal open laparotomy. The lesion in the pericaval (left) region superior to pancreas although pliable but was not cystic. After placing isolating packs soaked with 3% hypertonic saline (NS) an aspiration of suspected hydatid cyst was attempted, it drew ~15ml of straw-coloured fluid (send for lipase estimation) and reinjected with 3% NS . (Fig.2 a, b, d) The cyst wall was opened and fleshy to putty like material taken out with ovum forceps. A well-formed cyst wall was excised in toto..

Lesion to the left of the first one was firm, and aspiration did not reveal any fluid and it was excised in toto .

Lesion in the left lower parietal wall above the anterior superior iliac spine & the peri vesical area were found to be firm, solid well encapsulated masses in the wall of internal oblique & the rectus abdominis, respectively, were excised . (Fig.2c)

Wound closed after lavage & haemostasis. Diagnostic cystoscopy showed normal bladder.

Macroscopically, the lesions were lobulated and pale yellow in colour. (Fig. 2 e, f, g, h, i, j)



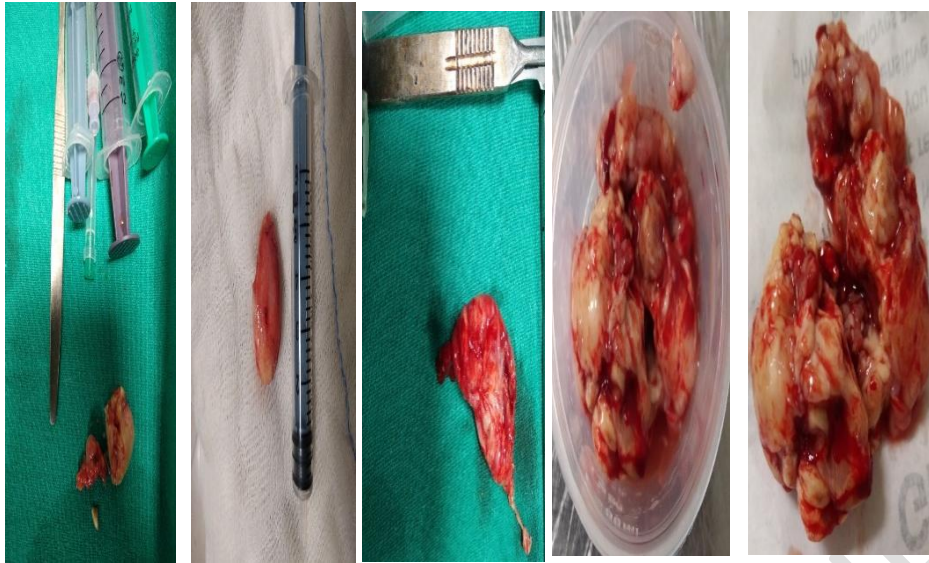


Figure 2: Intraoperative image (a) aspirating cyst wall (b) post aspiration. (c) pelvic area cyst (d) cyst fluid (e) Left suprarenal cyst (f) Pelvic cavity cyst (g) Abdominal wall cyst (h) Excised cyst wall (i) & (j) multiple cysts

Post-Op:

Post-operative period was uneventful. Patient was allowed liquids orally, the next day evening following surgery. Drain was removed on 3rd post-operative day. Patient was discharged with sutures in situ and with antibiotics advised.

On Histopathological examination: All the masses showed well encapsulated spindle cell tumor with cells arranged in fascicles. (See Fig. 3 d, e) There were hyper & hypocellular areas with occasional Verocay body formation, hyalinized thick-walled blood vessels & areas of secondary degenerative and Xanthogranulomatous changes, without any evidence of atypia, mitosis, or malignancy thus suggestive of, peripheral nerve sheath tumors (Schwannomas) (See Fig. a, b, e f). Chronic calculous cholecystitis was also reported. No evidence of granuloma or malignancy noted. The fluid from the cyst has normal lipase levels.

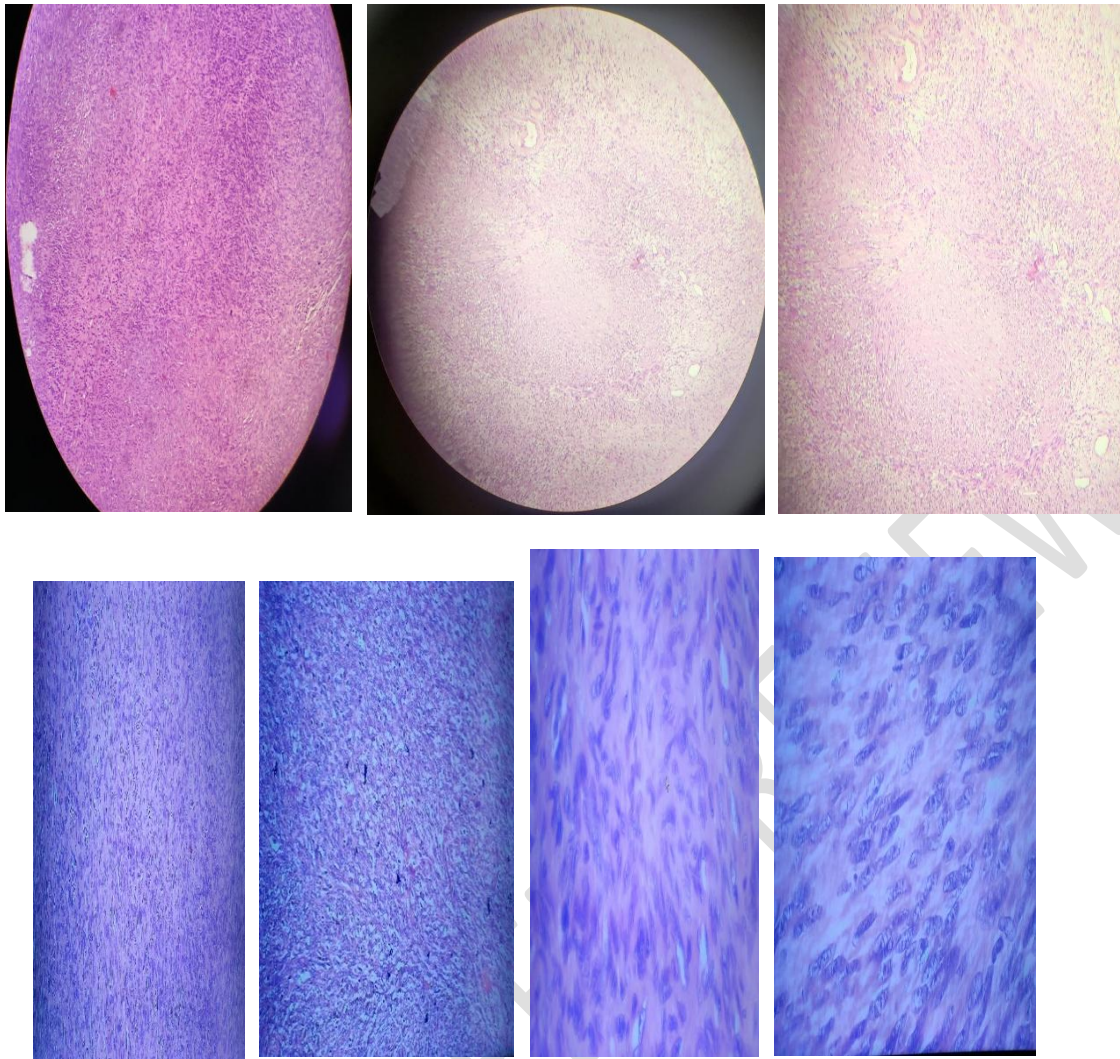


Figure 3: (a) (b) (c) High power view showing Verocay bodies. (d) (e) showing spindle cell tumor with cells arranged in fascicles, occasional Verocay bodies. (f) (g) showing spindle cells

Follow up:

Follow-up ultrasound at 2 weeks showed complete resolution of the mass. Patient remained asymptomatic in follow up.

DISCUSSION: -

Hydatid disease usually manifests as a slow growing cystic mass, either liver or lungs commonly. The cysts may be single or multiple; uni- or multiloculated,

and thin or thick walled. Specific signs include visualization or calcification of the cyst wall, presence of daughter cysts and membrane detachment; however, hydatid cysts with unusual localizations make diagnosis difficult [1, 4].

Echinococcal involvement of other than these two organs are extremely rare. Possible sources include hematogenous dissemination, local spread via the pancreatobiliary ducts and peripancreatic invasion [2, 5]. The variable clinical picture is caused by compression of nearby structures.

Schwannomas or neurilemmoma, occur as solitary tumours often in adult females [9]. Up to 20% of cases are usually associated with neurofibromatosis type 1 [10]. They are usually seen over head, neck, and flexor surface of extremities. Uncommonly sporadic reports of them arising in the porta hepatis [11], retroperitoneum, pelvis, adrenals, kidneys, and vagina [12] has been noted. Neurilemmoma arising in the intercostal nerves can give impression of a liver tumour [13] or a malignant tumour [14].

In the skin, schwannomas generally do not interfere with nerve conduction, but when they become large, they can compress the nerve of origin causing allodynia, paraesthesia, or dysesthesia [15].

Surgical excision is the treatment of choice. Recurrence of schwannomas has been reported in the literature probably suggesting that malignant lesions may arise *ab initio* [9]. It is also likely that recurrence after excision may be due to incomplete resection [13].

CONCLUSION: -

Hydatid disease should be considered in the differential diagnosis of all cystic masses in all anatomic locations, especially in endemic region. The ideal treatment is the complete excision of the cyst without any spillage.

Imaging studies therefore were suggestive of disseminated hydatid disease.

Tissue diagnosis post surgically however revealed the true nature of swellings to be schwannomas.

Intrabdominal schwannomas are rare tumours which most commonly exhibit gastrointestinal involvement. Since these tumours are mostly benign, the long-term prognosis of patients is good. Schwannoma should be kept in mind in the differential diagnosis of intrabdominal masses. Radical resections with high morbidity and mortality can be avoided if preoperative diagnosis is made or suspected.

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