ACTA MEDICA SALINIANA

CASE REPORTS

PRIMARY HYDATID CYST IN THE SPLEEN — TWO CASE REPORTS TREATED SURGICALLY WITH REVIEW OF LITERATURE

Arif Hussain Sarmast, Hakim Irfan Showkat, Fazal Q. Parray, Mohammad Yousuf Kachroo, Suhail Farooq Mir, Yasir Ahmed Lone, Aamir Aalam Khan, Omar Masood Khan

ISSN 0350-364X

Arif Hussain Sarmast, Hakim Irfan Showkat, Fazal Q Parray, Mohammad Yousuf Kachroo, Suhail Farooq Mir, Yasir Ahmed Lone, Aamir Aalam Khan, Omar Masood Khan

DOI: 10.5457/266.11

Hydatid disease has been recognised since ancient times and has a worldwide distribution and is endemic in Kashmir. Berlot in 1790 is accredited with the first description of a splenic hydatid cyst as an autopsy finding. Echinococcus granulosus is the causative organism is classic hydatid cysts. Primary hydatid spleen is a rare entity. We present two cases of primary hydatid spleen picked up incidentally.

Key words: *Echinococcus granulosus*, Splenectomy, Splenic hydatid

Institution
Sher i Kashmir Institute of Medical Sciences
India

Received:
19.11.2011
Accepted:
19.05.2012

Correspondence to:
Dr Arif Hussain Sarmast
Dalipora Kawadara
Srinagar Kashmir
India
pin 190002
phone 0194 - 2425338

arifhsarmast@gmail.com

+387 61 272 990
almir.salkic@gmail.com

Competing interests
The authors declare no competing interests.

INTRODUCTION:

The larval form of the genus Echinococcus, of which Echinococcus granulosus is the most common,causes hydatid disease. The liver and lungs are the organs most commonly involved. Splenic involvement in hydatid disease is uncommon, representing less than 2% to 3.5% of all human infestations by Echinococcus 1,2. The infestation of the spleen usually takes place by arterial route after the parasite has passed through the two filters, hepatic and pulmonary. The retrograde venous route which avoids the liver and lung, is also considered 3. Cystic lesions of the spleen are generally rare, and a parasitic origin is relatively unlikely. Non-parasitic cysts account for less than one-third of all splenic cyst cases. Splenic hydatid cysts are rarely encountered, at least once in their career by most surgeons at first or second hand, especially in endemic areas. Hydatid spleen has no predilection for any age and sex. It may be detected incidentally or present with non specific complaints. However up to 30% are incidental findings in asymptomatic individuals. Splenic hydatid cysts may suppurate, fistulise to adjacent organs or rupture into the peritoneal cavity or may present with massive lower gastrointestinal bleeding. Rarely splenic hydatid cyst perforates into the stomach, colon, thorax, or fistulise to the skin. Serology, abdominal ultrasound and computed tomography make definitive diagnosis.Management varies from spleen preservation to Splenectomy.4

CASE 1:

A 45 year old female normotensive presented with vague abdominal discomfort for 6 months. Examination was normal except for palpable spleen .Her Hb was 10.7 with counts and blood chemistry normal .USG revealed enlarged spleen with a well defined rounded heterogenous echotextured lesion of 90 x 81 mm in its upper part. Lesion shows well defined wall with membrane like structures inside [fig 1]. CT scan showed a large cystic density measuring 81 x 85 mm with wall calcification with central walled cystic area with calcified nodule suggestive of calcified and degenerated hydatid spleen [fig 2]. ELISA IgG for echinococcus was suggestive of 1:160. Elective splenectomy was done with cut section showing hydatid cyst with membranes [fig 3].

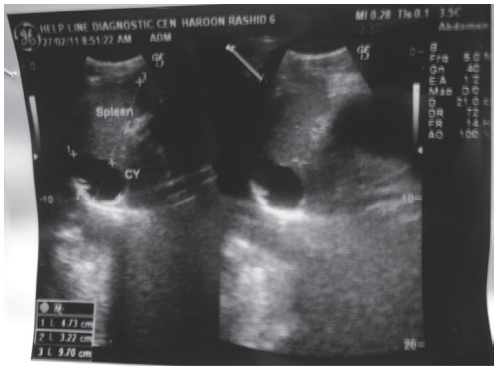

Fig 1 showing usg with splenic hydatid

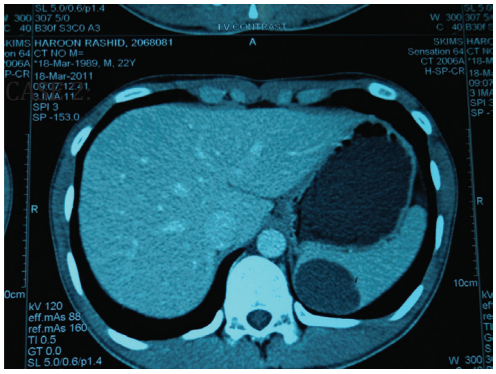

Fig 2 CECT of the same patient showin cytic lesion in spleen

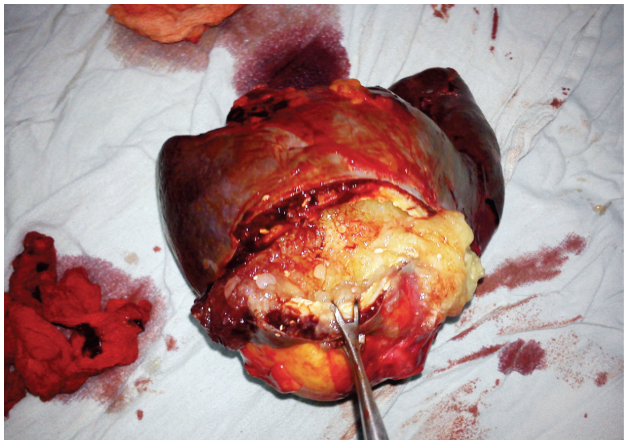

Fig 3 post operative specimen showing membranes suggestive of hydatid cyst

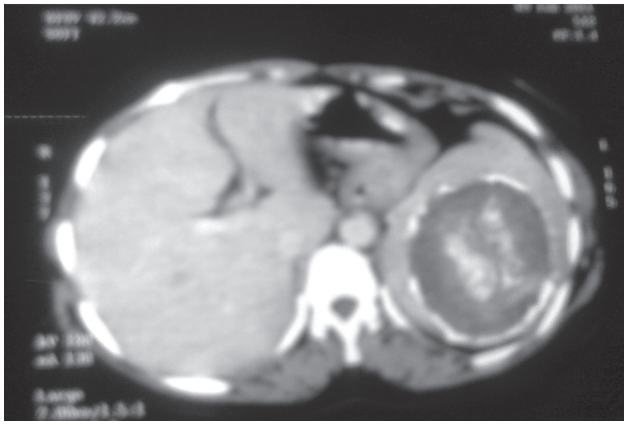

Fig 4 CECT of patient showing cyst with a calcified wall

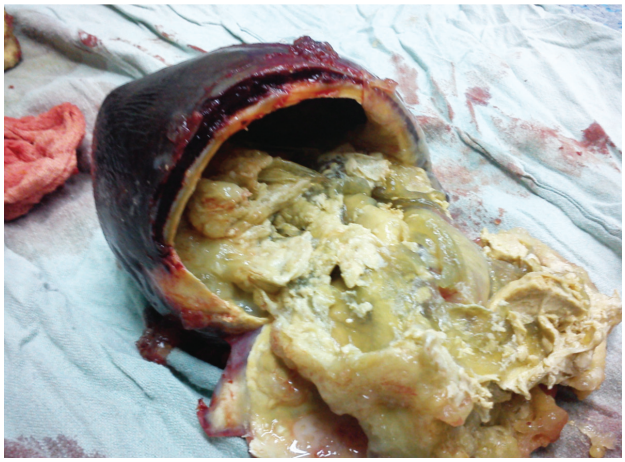

Fig 5 post operative picture of specimen of same patient(as in fig 4) showin hydatid spleen

A 22 year old male of average built presented to our OPD with recurrent complaints of dyspepsia. He was managed with PPIs for significant time. Clinical examination revealed nothing significant. His counts and blood chemistry was normal . USG abdomen revealed a cystic lesion measuring 42 x 47 mm seen in posterior aspect of upper pole of spleen close to diaphragm. CECT showed a well defined cystic lesion 45 x 40 mm with internal septae at upper pole of spleen with no solid component and internal calcification was seen [fig 4]. Elective splenectomy was done revealing hydatid spleen on gross examination [fig 5].

DISCUSSION:

Hydatid disease caused by the genus Echinococcus is endemic in Eastern Europe, Middle East, South America, North Africa, Indian subcontinent, Australia and New Zealand. 5 Echinococcus granulosus is the commonest organism involved, with dogs usually as the definitive host and human beings acting as an accidental intermediate host.

Enzymatic digestion of the shells of the eggs (shed in the faeces of the dogs) in the duodenum of the intermediate host releases the embryonic forms. These migrate through the intestinal wall to enter the portal circulation, from where they make their way towards the liver or lung, developing a triple cystic covering which grows grows at a rate of about 1 centimeter a year.6

Biochemical tests employed in diagnosis are complement-fixation test, enzyme-linked immunosorbent assay, indirect hemagglutination test, serum immunoelectrophoresis and western blot test.7 Plain x-ray film can show cyst wall calcification, however, there are no definite signs of hydatid cyst on ultrasonogram or computed tomography which could differentiate it from non-parasitic cysts, abscesses, or cystic neoplasms. 8

World over as well in India the incidence of splenic hydatid cysts reported by different workers from different cities is as shown in Table 1, 2.

Table 1. Country incidence of splenic hydatid cysts

Country	Incidence (%)
Australia 9	2.10
Argentina10	2.14
Iceland11	0.78
Iran12	4.00

Table 2. Incidence of splenic hydatid cysts in Indian Cities

Cities	Incidence (%)
Ahmedabad14	2.00
Delhi15	4.20
Indore16	3.30
Nagpur17(Central India)	6.30
Pondicherry18	2.70
Srinagar19	4.10

Splenic hydatid disease is usually an accidentally discovered mass in the abdomen mostly in left hypochondrium and less frequently in the epigastrium. Pain usually a dull dragging ache, is often the first clinical sign. Abdominal pain, tenderness and fever are the most common presenting symptoms. Dyspepsia, constipation due to pressure on colon, hypertension due to left renal artery compression, and dyspnoea due to pushing up of the left diaphragm may also occur. Other forms of clinical presentation include the rupture of splenic echinococcal cyst with anaphylactic shock, traumatic or spontaneous, acute abdomen, rupture of longstanding splenic hydatid cyst into bronchial tree or even fistulizing to the colon.13 Most echinococcal cysts

are asymptomatic on presentation, but complications such as pulmonary infection, cholangitis, rupture, and anaphylaxis give good reason to consider treatment for all. Medical, surgical, and percutaneous approaches may be part of the treatment armamentarium.20 Medical treatment of hydatid cyst, involves oral intake of albendazole,mebendazole,or praziquental.21 However Albendazole therapy is the mainstay of treatment in the majority of patients with hydatid disease. It is given alone and for prolonged periods of time in patients who are poor candidates for cyst-directed intervention.22. These medications are also used as a complement to surgical treatment to avoid recurrence. These are orally ingested preoperatively to prevent the consequences of possible rupture of the cysts during surgery, and postoperatively as adjuvant therapy of the cysts that may have ruptured during the operation 21 The usual dosage of orally administered albendazole is 10–15 mg/kg per day in 2 divided doses, or as a fixed dosage of 400 mg twice per day. If mebendazole is administered, the daily dosage is 40–50 mg/kg in 3 divided doses.23

Surgical dictum has stated that percutaneous puncture of a hydatid cyst is a dangerous and contraindicated activity. It was believed that the risk of anaphylaxis, communication with the biliary tree, and spillage outweighed any potential advantages with this nonoperative approach. In 1983, Fornage24 challenged this axiom and reported an accidental puncture of a hydatid cyst by US that had no clinical consequences. In patients with anatomically appropriate lesions PAIR (percutaneous aspiration, injection and re-aspiration) is the preferred initial treatment. The most frequently utilized protoscolecidal agents used for percutaneous treatment are 15–20% saline, 95% ethanol, a combination of 30% saline and 95% ethanol, and mebendazole solution. The PAIR technique has also been combined with albendazole therapy with 70% success rates and a low rate of recurrence.22 Gabal AM 25 and colleagues modified the standard PAIR technique and gave a version to be used in high risk hydatid cysts . Despite these reports, percutaneous treatment is not benign. Spillage, anaphylaxis, and recurrence can be life-threatening. Complete aspiration of all cyst contents, especially multivesicular disease, is difficult, and complete sterilization with protoscolecidal agents is uncertain. If the protoscolecidal agent enters the biliary tree, serious damage also can occur within the liver. Exogenous vesiculation may also go undiscovered. Long-term results are unknown at this time.26 Surgery has stood the time as the gold standard treatment for most if not all types of hydatid cysts . A number of operations have been used, but in general, the abdomen is completely explored, the liver{other involved organ mobilized}, and the cyst exposed. Packing off of the abdomen is important because rupture can result in anaphylaxis and diffuse seeding. Usually, the cyst is then aspirated through a closed-suction system and flushed with a scolicidal agent .. The cyst is then unroofed, which can then be followed by a number of possibilities, including excision (or pericystectomy), marsupialization procedures, leaving the cyst open, drainage of the cyst or omentoplasty to encompass the cyst. Total pericystectomy or formal partial hepatectomy can also be performed without entering the cyst 27 . Pericystectomy involves complete resection of the cyst wall without entering the cyst cavity. This procedure is done through a plane outside of the pericyst or along the cyst wall itself.Pericystectomy decreases the risk of spillage of cyst contents into the peritoneal cavity and also lowers the risk of recurrence. The disadvantage to this approach is the potential for bleeding or damage to bile ducts in proximity to the cyst wall.33 Radical (resection) and conservative (drainage and evacuation) surgical approaches appear to be equally effective at controlling disease, although a prospective comparison has never been done. Recurrence rates after surgical treatment range from 1% to 20% but are generally 5% or less in experienced centers.28

Peripherally located echinococcal hepatic cysts may be safely managed by laparoscopic cyst evacuation. The lesions best suited for this approach are situated anteriorly and do not have thick calcified walls. A right lateral approach also works for cysts in segments VI and VII .The cavity may be plugged with omentum or closed over a closed suction drain.29 Multiple cysts within proximity to a major blood supply or to each other, or a cyst in a relatively safe location (i.e., segments II/III) are candidates for resection provided a complete resection can be achieved. 20

Surgical resection is curative in up to one third of cases of E. multilocularis infection. In most cases the disease is advanced when the diagnosis is made. In such cases, palliative drainage procedures or long-term treatment with albendazole or other benzimidazole carbamates may prolong survival. 30,31 Splenectomy is the treatment of choice because there is no effective medical treatment for splenic hydatid cyst.32 Since 1980 there has been a trend towards splenic conservation to avoid overwhelming post splenectomy infection (OPSI). For conservation of the spleen, enucleation of a unilocular cyst can be done. Secondary hydatid disease of spleen may be caused by rupture of abdominal cysts (e.g. in the liver) with dissemination to the spleen.33 Once ruptured, cysts metastases occurs to the peritoneal surface and despite its benign, infectious aetiology, hydatid cysts exhibit malignant behaviour and the whole of the abdomen can get studded with multiple hydatid cysts of different size. Recently Ballaux KE performed a hand assisted laparoscopic splenectomy for a large splenic hydatid cyst.34 Children are more prone to OPSI. The clinical syndrome of OPSI comprises fulminant bacteraemia, disseminated intravascular coagulation, multiple organ failure, severe hypoglycaemia and often rapid death. 35 Its reported incidence after splenectomy varies from 0.9 to 60% with mortality exceeding 50%. 36.Preservation of spleen should always be tried especially in children to prevent OPSI.37

REFERENCES:

1. Franquet T, Montes M, Lecumberri FJ, Esparza J, Bescos JM. Hydatid disease of the spleen : imaging findings in nine patients. Am J Roentgenol 1990;154:525-8.

2. Uriarte C, Pomares N, Martin M, Conde A, Alonso N, BuenoA. Splenic hydatidosis. Am J Trop Med Hyg 1991;44:420-3.

3. Ionescu A, Jakab A, Jutis T, Forai F, Ota A. Splenic hydatid cyst (Article in Romanian). Rev Med Chir Soc Med Nat Iasi. 1990;94:525-8.

4 .Imtiaz Wani et al ,Internet Journal of Medical Update 2010 July;5(2):68-70

5. Ruiz Castro M, Vidal Marcos A, de Frutos S, et al. Shock anafiláctico por rotura traumática de quiste hidatídico hepático desconocido. Rev Esp Anestesiol Reanim 1997;44:321-3.

6. Bertó JM, Martínez F, Ojeda JA. Shock anafiláctico como manifestación de hidatidosis hepática. Rev Clin Esp 1991;188: 60-1.

7.Kune GA, Morris Di, Schwartz & Ellis. 9th edition Appleton and Lange 1989: 1225 – 40.

8.Kalouidouris A, Pissilosis C, Pontifix GCT charecterization of multi vesicular hydatid cysts. J Comput Assist Tomograp. 1984: 8; 839 – 45.

9.Braqaehaye J (1892) Cited by Fowler RH. Hydatid cysts of spleen, collective review International abstract of surgery. 1953; 96 : 105-116.

10.Greenway DJ, J Am M Ass 1922; 79 : 2018. Cited by Fowler RH, Hydatid cysts of the spleen, collective review International abstract of surgery. 1953; 96 : 105-116.

11.Finsen 1867. Cited by Fowler RH, Hydatid cysts of spleen collective review International abstract of surgery. 1953; 96 : 106-116.

12.Torbati M. Hydatid diseases of the spleen, Report of an Unusual case. Brit J Surg 1972; 59 : 489-91.

13. 3. Caballero P, Ocon E, Robledo AG, et al. Splenic hydatid cysts opening to the colon. Am J.Roentgenol1986;147:859-60.

14.Shah SH, Kanavinde MS, Shukla BK, Mehta KN. Hydatid disease. A comparative review of its incidence in Ahmedabad with that in other parts of India. Ind J Med Sci 1969; 23 : 380-86.

15.Nair SK, Singhal RN, Prasad P. Hydatid cyst of the Breast. Ind J Surg 1982; 441 : 130-33.

16.Gupta JC, Nagrath CL, Salgia KM. Hydatid disease in man. J Ind Med Assoc 1966; 46 : 649-51.

17.Bhobhate SK, Kedar GP, Kher AV. Incidence of hydatid disease in Vidarbha division. Ind J Surg 1984; 46: 162-64.

18.Shrivastava BK, Mehdiratta KS, Satya Prakash. Hydatid disease is Pondicherry. Ind J Surg 1982; 44 : 88-94. 19.Wani NA. Hydatid Disease of the spleen. Ind J Surg 1993; 55 (3) : 155-60.

20. Goldblatt M, Pitt H. Hepatic echinococcosis. In: Cameron J (ed). Current Surgical Therapy, 8th ed. Philadelphia, PA: Elsevier Mosby; 2004:306–311

21. Keshmiri M, Baharvahdat H, Fattahi SH, et al. A placebo controlled study of albendazole in the treatment of pulmonary echinococcosis. Eur Respir J. 1999;14:503–507. [PubMed Citation]

22. Khuroo MS, Wani NA, Javid G, et al: Percutaneous drainage compared with surgery for hepatic hydatid cysts. N Engl J Med 337:881, 1997. [PMID: 9302302]

23. Smith AL, Rego LP, Williams R. . Albendazole monitoring. Am J Health Syst Pharm 1997;54:319-20

24. Fornage B. Fortuitous diagnosis by fine needle puncture under real-time ultrasound control of an atypical hydatid cyst of the liver. J Radiologie 1983;64:643–645 [PubMed: 6663561]

25. Gabal AM,Khawaja FI,Mohammad GA. Modified PAIR technique for percutaneous treatment of hih-risk hydatid cysts.Cardiovasc Intervent Radiol.2005;28:200-208.[pubMed]

26. Milicevic MN. Hydatid disease. In: Blumgart LH, Fong Y (eds). Surgery of the Liver and Biliary Tract, 3rd ed. London, England: WB Saunders; 2001:1167–1204

27. McManus DP, Zhang W, Li J, et al: Echinococcosis. Lancet 2003; 362:1295-1304.

28. Dziri C, Haouet K, Fingerhut A: Treatment of hydatid cyst of the liver: Where is the evidence?. World J Surg 2004; 28:731-736

29. Ertem M, Karahasanoglu T, Yavuz N et al. Laparoscopically treated liver hydatid cysts. Arch Surg 2002;137:1170 [PubMed: 12361429]

30. Akinoglu A, Demiryurek H, Guzel C: Alveolar hydatid disease of the liver: A report on thirty-nine surgical cases in eastern Anatolia, Turkey. Am J Trop Med Hyg 1991; 45:182.

31. Craig P: Echinococcus multilocularis. Curr Opin Infect Dis 2003; 16:437.

32. Seymour I, Schwartz, Harold Ellis, Maingot's Abdominal Operations. Tenth edition Vol II, 1534-45.

33. Von-Sinner WN, Stridebeck H. Hydatid disease of the spleen. Ultrasonography, CT and MR imaging. Acta Radiol 1992; 33 (5) : 459-61.

34. Ballaux KE, Himpens JM, Leman G, Van den Bossche MR. Hand assisted laparoscopic splenectomy for hydatid cyst. Surg Endosc 1997; 11 (9) : 942-43.

35. Singer DB. Postsplenectomy sepsis. In : Rosenberg HS, Bolande RP, eds. Perspectives in paediatric pathology, Chicago :Yearbook Medical publishers. 1973; 285-311.

36. Holdsworth RJ, Irving AD, Cushieri A. Postsplenectomy sepsis and its mortality rate : actual versus perceived risks. Br J Surg 1991; 78 : 1031-8.

37.Dickerman JD. Traumatic asplenia in adults. A defined hazard? Arch Surg 1981; 116 : 361-63.

Consent

A detailed informed consent was taken from the two patients regarding the publication of their presentation as a case report .

Authors' contributions

Author 1 was involved in conception and design, or acquisition of data, or analysis and interpretation of data; &has been involved in drafting the manuscript.

Author 2 was also involved in conception and design, or acquisition of data.

Author 3 gave his final approval of the version to be published and was involved in conception and design, or acquisition of data, or analysis and interpretation of data;

Author 4 was involved in revising it critically for important intellectual content

Author 5 was involved in type setting and collecting references.

Author 6 was also involved in conception and design