Abdominal hydatid disease. Complications and atypical locations

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Purpose

To review the complicated and atypical cases of abdominal hydatid disease (HD) at a second-level hospital in an endemic area, with emphasis on the radiological findings.
Methods and Materials

This is a retrospective descriptive observational study.

We retrospectively reviewed the clinical and imaging records of the patients with abdominal echinococcal disease coded as primary, secondary or tertiary diagnosis according to the International Classification of Diseases 10th Revision Procedure Classification System (ICD-10-PCS) at our institution from January 2003 to March 2011.

We selected those patients with pathologically or serologically proven HD. We defined complicated disease as that one inducing clinical signs or symptoms in an acute or subacute way. Any location different from the liver or the biliary tract was considered atypical. Exophytic growth from the liver was not considered either atypical location or complication by itself.

CT examinations were performed with single helical equipment prior to September 2008 and with a 40 row MDCT afterwards. MR studies were performed on a 0.5 T magnet prior to November 2009 and on a 1.5 magnet afterwards. The ultrasound equipment varied quite a lot throughout the study.
Results

Eighty-one patients (46 male, 35 female; 56.8 vs. 43.7%) were included. Their ages ranged from 17 to 87 years (M=54.7 ± 18.3). Among them, 53 (65.4%) had either complications and/or atypical locations of HD in the abdomen.

COMPLICATIONS

Forty complications were diagnosed in 36 (44.4%) patients: one complication in 33 cases, two in 2 and three in 1 case. 12/36 (33.3%) had undergone surgery for hepatic HD prior to the current episode. These complications are listed in descending order of frequency (fig. 1):

- Biliary frank fistulization (figs. 2, 3) of a hepatic cyst was the most frequent complication, seen in 14 cases (17.3%). There was a surgical antecedent for HD in 3/14 patients (21.4%).

- Acute or subacute rupture into the peritoneal space (fig. 4) was seen in 6 patients (7.4%). The median age of these patients was 38.3 years, significantly lower than that of the patients with complicated HD (55.8). The rupture was secondary to recent trauma in 3 cases, in 2 of them to a traffic accident. 2/6 patients had previously undergone surgery for HD.

- Pyogenic infection of a liver cyst (fig. 5) was demonstrated in 4 cases (4.9%). 1/4 had undergone surgery for HD.

- Transdiaphragmatic migration (fig. 6) (n=3; 3.7%): one case of liver cyst migrating across the bare area into the right thoracic base and two left subphrenic cysts eroding into the left pleural space. 1/3 had undergone surgery for HD.

- Biliary tract compression (fig. 7) (n=3; 3.7%) causing jaundice. 2/3 had undergone surgery for HD.

- Abdominal wall direct involvement (fig. 8) (n=2; 2.5%): one hepatic cyst invading the right upper quadrant abdominal wall and directly opening into skin and one left subphrenic cyst that invaded the posterior lumbar upper wall, with rib erosion and erector spinae muscle involvement in addition to transphrenic migration. 1/2 had undergone surgery for HD.

- Retroperitoneal invasion (fig. 9) (n =2; 2.5%) of right liver lobe cysts extending into the upper retroperitoneal right space across the bare area, with contiguous renal involvement in one of them. 2/2 had undergone surgery for HD.
- Acute rupture of the endocyst (fig. 10), rupture into the subcapsular liver space (fig. 11), spontaneous fistulization into the bowel (duodenum, fig. 12), urinary bladder invasion (fig. 13), seminal vesicle involvement (fig. 13) and portal vein luminal invasion (fig. 14) were each seen in one patient (1.2%).

**ATYPICAL LOCATIONS**

There were up to 38 atypical locations of HD in the abdomen or pelvis of 26 patients (32.1%): one single atypical location in 18, two in 4 and three in 4 different patients. 13/26 (50%) also had complicated disease. Liver HD coexisted in 19/26 patients (73.1%) (fig. 15). There was an antecedent of surgery for HD in 9/26 patients (34.6%) (fig. 16). This antecedent was more frequent (5/8: 62.5%) among those patients with more than one atypical location.

- The most common atypical location was the peritoneum (figs. 17, 18) (n=13; 16%). These 13 patients included 6 with acute or subacute rupture of a liver cyst into the peritoneal cavity. 11/13 patients also had hepatic HD and 6/13 had undergone surgery for HD.

- Spleen (fig. 19) (n=6; 7.4%). 4/6 patients also had hepatic HD and 2/6 had undergone surgery for HD.

- Diaphragm (fig. 20) (n=3; 3.7%). 3/3 patients also had hepatic HD and 2/3 had undergone surgery for HD.

- Abdominal wall (fig. 21) (n=3; 3.7%): 2 cases with direct extension of intrabdominal cyst (also considered complicated disease) and 1 with isolated muscular involvement. 3/3 patients also had hepatic HD and 1/3 had undergone surgery for HD.

- Retroperitoneum (fig. 9) (n=2; 2.5%). 2/2 patients also had hepatic HD and 2/2 had undergone surgery for HD.

- Kidney (fig. 22) (n=2; 2.5%). Only hematogenously spread disease was considered here. One case of renal involvement of a hepatic cyst extending to the retroperitoneum is only included as retroperitoneal location. 1/2 patients also had hepatic HD and 1/2 had undergone surgery for HD.

- Gastrointestinal tract (n=2; 2.5%): gastric wall cyst (fig. 23) in one case and duodenal fistulisation of a liver cyst in other (fig. 12). 1/2 patients also had hepatic HD and 0/2 had undergone surgery for HD.

- Portal vein (fig. 14), adrenal (fig. 24), pancreas (fig. 25), psoas muscle (fig. 26), urinary bladder (fig. 27), seminal vesicle (fig. 27) and a diaphragmatic hernia (fig. 28) were each involved by HD in one case (1.2%).
Fig. 1: Graph showing the distribution of complications of HD, and their relationship with previous surgery.

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Fig. 2: Biliary fistulization. Contrast enhanced CT and MRI show a hepatic hydatid cyst (*) communicating (arrow) with a dilated bile duct. There is also hyperenhancement of choledocal mucosa (arrowhead) as a manifestation of hydatid cholangitis and regional differences in liver parenchymal enhancement with a straight border, in relation to inflammatory phenomena.

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Fig. 3: Biliary fistulization in three different cases. There is hydatid material ("sand" and small vesicles) in the common bile and intrahepatic ducts in cases 1 and 2 (arrowheads), and also in the gallbladder in case 2 (*), finding that confirms the diagnosis in an appropriate setting. In case 3 MRI-cholangiography shows a liver hydatid cyst with a lobulation towards a dilated bile duct. Operative cholangiogram, following cholecystectomy, choledocotomy, clearance of hydatid material and biliary irrigation, confirms the existence of a communication between the cyst (+) and the biliary tree.
**Fig. 4:** Acute peritoneal rupture. Emergent US performed to a 28 years old man that suffered a traffic accident, with intense abdominal pain. A peripheral, exophytically growing hepatic hydatid cyst is seen, with detachment of the endocystic layer (arrows) and surrounded by subhepatic ascitis (*). Peritoneal fluid, which proved to be hydatid clear liquid after urgent surgery, was also seen in peritoneal pelvic recesses. Vejiga: urinary bladder.

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**Fig. 5:** Pyogenic infection in three different patients. 1. There is gas within a hydatid liver cyst with peripheral calcification (*), which is diagnostic of pyogenic infecion in the absence of an antecedent of intervention. 2. There is loss of the sharp delineation of the margins and of the spherical morphology of a cystic liver lesion (*). This appearance is indistinguishable from hepatic abscess if no other typical findings are present. Pleural effusion is also seen (+). 3. Fever and jaundice. A liver hydatid cyst is present (*), with a focal peripheral calcification and no other features suggestive of pyogenic infection, that was nevertheless demonstrated after surgical treatment of this lesion that also showed biliary fistulization. Arrow points to a dilated bile duct.

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Fig. 6: Transphrenic migration. Left subdiaphragmatic hydatid peritoneal cysts (+) with invasion across the left hemidiaphragm into the left pleural space (arrows) in a patient that had previously undergone surgery for liver and splenic HD, with splenectomy. There is another peritoneal cyst in the gastrocolic ligament (*).

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Fig. 7: Biliary compression. CECT and MRI in a patient with progressive jaundice. There is a liver hydatid cyst with a fine capsular calcification in segment IVa (*) compressing the left biliary duct and inducing biliary dilatation circumscribed to the left lobe. There is another more densely calcified hydatid cyst in hepatic segment VII.

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Fig. 8: Abdominal wall invasion. A liver hydatid cyst in the anterior surface of segment II (*) is extending through a small defect into the perihepatic space and the anterior abdominal wall, reaching the skin surface (open arrows). Hydatid material flowed through a gap.
Fig. 9: Retroperitoneal invasion. Hepatic hydatid cysts in segment VII (*) directly extending into the upper right retroperitoneum (+) through the bare area of the liver, involving the upper pole of the right kidney (RK).
Fig. 10: Endocystic detachment. An undulating membrane representing the endocystic layer (arrow) is seen separating from the outer limit, which represents the pericystic layer (hollow arrow) of this liver hydatid cyst. This is a manifestation of contained rupture.

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Fig. 11: Subcapsular rupture. Small peripheral echogenic hydatid cyst in hepatic segment VII (*) with direct rupture, forming a subcapsular complex collection (+) with liver surface scalloping. There are fatty foci within the subcapsular cyst (arrows). The presence of fat in a hydatid cyst has been related to passage of bile into the cyst through a fistula, but it was not surgically demonstrated in this case.

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Fig. 12: Bowel fistulization. CT clearly depicts the communication (arrows) between this liver hydatid cyst (*) containing a small amount of gas and the duodenal bulb (+). This communication was previously suspected on US and later demonstrated on barium meal examination, with passage of ingested barium from the duodenum into the cyst.

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Fig. 13: Urinary bladder and seminal gland invasion. 59-year-old male patient with hydatiduria and hydatidospermia. There is a direct communication (arrows) between the anterior wall of a hydatid cyst in the retrovesical pouch of the peritoneal sac (+) and the posterior wall of the urinary bladder, allowing passage of hydatid material. The cyst also invaded the right seminal gland (*).

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**Fig. 14:** Portal vein invasion. Both US and CT show hydatid material (hollow arrows) including small vesicles along the main portal vein and its intrahepatic branches, with some peripheral calcifications and development of cavernomatosis. This is an exceptional form of presentation of HD, only previously demonstrated in five cases up to our knowledge. Portal vein thrombosis or occlusion secondary to compression is not so infrequent.

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Fig. 15: Graph showing the distribution of atypical abdominal locations of HD, and their relationship with the presence of liver involvement.

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**Fig. 16:** Graph showing the distribution of atypical abdominal locations of HD, and their relationship with previous surgery.

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Fig. 17: Peritoneum. US and MRI depict a large lobulated exophytically growing hepatic hydatid cyst (*) with a neighboring peritoneal lobulated cyst (+) secondary to chronic spillage of hydatid material from the liver cyst. There was no prior surgery.
Fig. 18: Peritoneum. Massive peritoneal hydatidosis (+) associated to liver disease (*) in a 90-year-old woman that had undergone surgery for hepatic HD more than 30 years previously.

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Fig. 19: Spleen. A. Multilocular and irregular peritoneal hydatid cyst invading the spleen (+) without associated hepatic HD. B. Unilocular splenic hydatid cyst (+) with floating membranes depicted on US, associated to recurrent liver HD (*) after previous surgery.

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Fig. 20: Diaphragm. Hydatid cyst in hepatic segment VI (*) with posterior growth across the bare area into the diaphragmatic right crura (+). There was also acute peritoneal rupture, with ascitis (a).

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Fig. 21: Abdominal wall. Left subphrenic hydatid cyst with transphrenic migration (*), also invading the posterior lumbar upper wall, with rib erosion (arrow) and erector spinae muscle involvement (+).

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Fig. 22: Kidney. 1. Left cystic lesion (hollow arrows) in the upper pole of the left kidney (LK). It had a single septation in 2007, progressively becoming more solid in appearance thereafter, still preserving some vesicles within. In 2010 a contrast enhanced sonographic examination was performed, showing only minimal enhancement in the wall of the largest of these vesicles (*). A liver hydatid cyst had been surgically removed 15 years earlier. 2. Unilocular cystic lesion in the lower pole of the left kidney (LK) with peripheral calcifications (arrows). There was no previous or simultaneous liver HD.

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Fig. 23: Gastric wall. Large complex cystic mass (+) with echogenic foci abutting the anterior wall of the gastric antrum (arrows). There was no previous or simultaneous liver HD. The patient underwent surgery without a definitive diagnosis. A cystic mass growing from the gastric wall was resected that proved to contain scolices and hydatid membranes at pathological examination.

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Fig. 24: Adrenal gland. Complex cystic mass in the right adrenal gland, with a spoke-wheel appearance (*). There was no previous or simultaneous liver HD.

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Fig. 25: Pancreas. Lobulated cystic mass (+) with internal echoes and peripheral calcification (arrow) in the pancreatic body. Hepatic HD was also present (*).

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**Fig. 26:** Psoas. Complex cystic mass (+) in the left psoas muscle (pm), which proved to be a hydatid cyst, with no coexisting liver HD.

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Fig. 27: Urinary bladder and seminal gland. Same patient as in figure 13. Peritoneal HD in the retrovesical sac (*) compressing the posterior wall of the urinary bladder (arrows) and eroding into its lumen, with spillage of hydatid material depicted on the excretory phase of CECT (hollow arrows). Invasion of the right seminal gland is also seen (+).

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Fig. 28: Diaphragmatic hernia. Large hernial sac through an anterior left diaphragmatic defect (arrows), containing a colonic segment (c) and a large and lobulated peritoneal hydatid cyst (+) with floating membranes and daughter vesicles. This 67-year-old woman had been earlier operated for hepatic and pleural HD.

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Conclusion

HD is a zoonosis caused by the larval stage of Echinococcus granulosus. The human can incorporate to the parasitic life cycle as the intermediate host, role that is generally played by sheep. The disease usually involves the liver (75%) and lung (15%, usually by means of hematogenous spread) as slowly growing cysts that contain daughter vesicles in which larvae develop. They tend to invade neighbor structures, particularly small bile radicals, which are incorporated into the pericystic layer that surround the cyst as a reaction of the hepatic tissue of the host.

HD is endemic in the great grazing regions of the world, which includes the Mediterranean area. It is not a notifiable disease in this moment in Spain, but an increase of its incidence is being appreciated in the last two decades, not only among immigrant population.

Complications of abdominal HD were quite frequent in our series (44.4%). This rate was not substantially different from previous studies. There is clearly a selection bias in our study and in others, based on the fact that only proven cases are included, so excluding many minor cases, in which the percentage of complicated and atypical disease is obviously lesser.

Agreeing with previous series, the most usual complication in ours was biliary frank fistulization (n = 14; 17.3%; 5-15% in the literature), a type of communicating rupture that implies the spillage of hydatid material into the bile duct tree, usually leading to jaundice. The clue to radiological demonstration of this complication is the depiction of hydatid vesicles or sand in a dilated bile duct system. This is better accomplished by US or MR than by CT. It is more difficult to directly demonstrate the fistula, although it is relatively frequent to see a lobulation of the cyst towards the communication. An older age, multiple, lobulated and large cysts have been cited as factors predisposing to this complication.

The second form of complication was acute or subacute rupture to the peritoneal cavity (n=6; 7.4%), which is a form of direct rupture that manifests as acute abdominal pain and occasionally systemic anaphylaxis. We did not consider peritoneal chronic seeding as complicated, but as atypical HD. In 3/6 cases peritoneal rupture of a liver cyst was induced by abdominal trauma, as have also been previously reported. This might explain the younger age of these patients, which was also seen in our study. Other predisposing described factors, that were also present in our series, are peripheral location and greater size of liver hydatid cysts.
Pyogenic infection (4; 4.9% in our series; 5-8% in the literature) requires rupture of both the pericyst and endocyst layers for the passage of bacteria into the cyst. It can clinically manifest similar to an abscess. The demonstration of gas in a lesion suggestive or known to be an echinococcal cyst is virtually diagnostic of its infection. Poorly defined margins, loss of the regular shape of the cyst and inflammatory perfusion phenomena in the surrounding liver parenchyma are other signs that suggest bacterial infection.

Transdiaphragmatic migration (n=3; 3.7% in our series; 0.6-16% in the literature) has been more frequently reported from the posterior segments of the right hepatic lobe across the bare area. We saw this type of extension in one case, but migration through the left hemidiaphragm in two.

The abdominal wall (n=2; 2.5% in our series) is rarely invaded, usually by hepatic cysts adopting an hourglass configuration through a small defect in the liver surface, as occurred in one of our patients, who presented with exudation of pus and hydatid contents through a skin orifice. In the other case the posterior left lumbar wall was invaded by a left subphrenic hydatid cyst.

We have also seen some other exceptional forms of complicated HD in isolated cases: rupture into the subcapsular liver space, spontaneous fistulization into the duodenum (reported in less than 0.5 % in the literature), urinary bladder invasion by peritoneal "cul-de-sac" HD in one patient with hydatiduria, seminal gland involvement in the same patient and portal vein luminal invasion (only radiologically demonstrated in five previous cases up to our knowledge).

The rate of extrahepatic HD (n = 26; 32.1%) was quite high in our series. Hepatic HD coexisted in a majority of cases (73.1%), and complications were also present in half of these patients. An antecedent of surgery for HD was more frequent in cases with more than one extrahepatic involvement (62.5%) than in cases with just one extrahepatic HD (22.2%).

The most usual non-liver abdominal disease in our study, as in others, was the peritoneum (n = 13; 16%; 13 % in the literature). It is usually the result of the seeding of a liver ruptured cyst, often after surgery (6/13 in our study), with the development of similarly behaving cysts anywhere in the peritoneal sac. We also saw two cases of primary peritoneal HD, without coexisting hepatic cysts.

The spleen (n=6; 7.4% in our series; 0.9-8% in the literature) was the second most common atypical location, coexisting with liver HD in 4/6. At least in one case splenic involvement was thought to be secondary to invasion from peritoneal disease. Splenic primary HD is infrequent, accounting for less than 2%.
The kidneys (n=2; 2.5% in our series; 3% in the literature) can be involved by hematogenous spread. None of our cases showed rupture of the cyst into the collecting system, which is present in up to 18% of renal HD.

Among other atypical extrahepatic abdominal HD, apart from those discussed as complicated, we present also some exceptional isolated cases: one hepatic cyst in the gastric wall without coexisting liver HD, one pancreatic hydatid cyst with associated liver cyst (0.25% in the literature, requiring differential diagnosis with pancreatic pseudocyst and cystic pancreatic neoplasm), one psoas muscle hydatid cyst without liver disease, one adrenal gland cyst (0.06-0.18% in autopsy series in the literature, radiologically leading to differential diagnosis with pheocromocitoma or other adrenal tumours with cystic or necrotic degeneration, among other conditions) and one large hydatid peritoneal cyst growing into a left diaphragmatic hernia that also included a segment of colon.

In conclusion, there is a relatively high rate of abdominal complications of HD, the most frequent being communicating fistulization into the biliary tree, followed by acute peritoneal rupture. The extrahepatic abdominal structure most usually involved in our series was the peritoneum, followed by the spleen. There seems to be an association between previous surgery for HD and extrahepatic hydatid infection. We also report some exceptional cases of abdominal and pelvic HD.
References


