CASE REPORT

A rare large symptomatic simple hepatic cyst

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SUMMARY

A fit and well 4-year-old girl presented with a 1-year history of abdominal distension, pain and vomiting with signs of non-shifting dullness on the right side of the abdomen. An abdominal CT revealed a 19 cm epigastric cyst that did not appear to connect to any solid organ and was therefore thought to represent a mesenteric cyst. However, laparotomy revealed that the cyst was arising from the liver. The cyst was resected and subsequent histology revealed a simple hepatic cyst. The patient has been well post-operation. The case represents the largest described simple hepatic cyst in the 0–16 years paediatric age group in Europe to date. We outline the epidemiology, embryology, key investigations and management options for these lesions.

BACKGROUND

Hepatic simple cysts are uncommon lesions with a reported prevalence of 1% on autopsy-based studies although the estimated prevalence on imaging-based studies is higher, ranging from 2.5% to 18%. The cysts are mostly seen in the over-40 age group, although they can occur in children. They are mostly seen in the right lobe of the liver. In adults and children, simple hepatic cysts are usually small with symptoms occurring in less than 20% of patients. Asymptomatic cysts occur slightly more frequently in females (female to male ratio 1.5:1) while symptomatic cysts occur much more commonly in females (ratio 9:1). When they do occur, the symptoms include early satiety, bloating, pain, nausea, vomiting and—if very large—the presentation of a mass. Rare complications include

torsion, rupture, secondary infection and obstructive jaundice. This report highlights a rare case of a large symptomatic simple hepatic cyst in a 4-year-old girl.

CASE PRESENTATION

An otherwise well 4-year-old girl presented with a 1-year history of gradual abdominal distension and intermittent abdominal pain associated with non-bilious vomiting. At presentation, her height was 104 cm and her weight was 16.6 kg. There was a history of occasional dry cough and allergy to penicillin. There was no significant family history and liver function tests were normal.

INVESTIGATIONS

Following an initial abdominal ultrasound, a CT scan of her abdomen revealed a 18×14×19 cm, thin-walled epigastric cyst compressing the pancreas (figures 1 and 2). The cyst was not obviously connected to any solid organs and an initial diagnosis of mesenteric cyst was made.

DIFFERENTIAL DIAGNOSIS

The initial differential diagnosis included duplication cyst.

TREATMENT

In contrast to the impression on imaging, laparotomy revealed a large cyst arising from the inferior surface of the liver. The cyst contained 2 L of fluid and was separated from the gallbladder. The cyst fluid was drained and the cyst completely excised except 4×5 cm of cyst wall on the liver surface with the cyst lining excised. The histology revealed

Figure 1 CT of the abdomen/pelvis (sagittal section).

Figure 2 CT of the abdomen/pelvis (coronal section).

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a cyst wall lined by flattened and cuboidal cells and containing nerves, vessels and bile duct-like structures with occasional foci of hepatocytes. The diagnosis was of a benign simple hepatic cyst.

OUTCOME AND FOLLOW-UP

The postoperative outcome was uneventful as the patient made a full recovery and is well at 1-year follow-up.

DISCUSSION

Simple hepatic cysts are believed to be congenital in nature: an anterior foregut diverticulum appears at 3 weeks gestational age and differentiates into both the fetal liver and the extrahepatic biliary tract. By 6 weeks, bile ductuli cells appear within primitive hepatocyte sheets. Over the next 7 weeks these bile ductuli cells form first a single layer and then a double layer of epithelial cells surrounding the primitive portal tract. This double cylindrical structure is known as the ductal plate and after 13 weeks it forms a network of tubules essentially representing primitive bile ducts. It is thought that liver cysts arise from an excess of these tubules which fail to involute and fail to communicate with the main biliary tract. Accumulation of the fluid secreted by the epithelial lining leads to progressive dilation of the tubule and eventually formation of the cyst. This is supported by the presence—seen histologically—of attenuated bile duct epithelium lining the cyst and of ductular structures within the fibrous wall of the cyst.

The main investigations are ultrasound, abdominal CT and sometimes an MRI. Our case is unusual in that the hepatic nature of the cyst was not obvious on ultrasound and CT and therefore, other abdominal cysts such as mesenteric cyst and duplication cyst were on the differential. In the majority of cases where it is clear that the lesion is hepatic, a simple hepatic cyst needs to be differentiated from abscess, hydatid cyst, vascular lesions, cystic mesenchymal hamartoma and biliary cystic neoplasms. The radiological features with appropriate clinical correlation are usually diagnostic. On ultrasound, a simple hepatic cyst is well circumscribed, non-echoic and with a thin indiscernible wall. CT reveals a well-circumscribed, thin-walled, non-septated lesion showing water attenuation and no evidence of enhancement following administration of contrast. On MRI, simple hepatic cysts show a thin indiscernible wall, a low signal on T1-weighted images and a high signal on T2-weighted images. However, haemorrhage into the cyst can confound the appearances with high attenuation signal, thickened wall and occasionally the development of septa. This can lead potentially to confusion with other cystic lesions.¹

In contrast to a simple hepatic cyst, a hepatic abscess will be associated with clinical and laboratory evidence of infection. A hydatid cyst can usually be diagnosed on serology. Furthermore, they display peripheral enhancement of the wall and formation of daughter cysts. On ultrasound, adult haemangioma tends to show hyperechogenicity while infantile haemangioendothelioma may be hypoechoic or of mixed echogenicity. Following contrast administration, CT and MRI scans of adult and paediatric vascular lesions show nodular enhancement of the periphery on arterial phase images while in the delayed phase there is centripetal filling-in.²

In children, cystic mesenchymal hamartoma is an important differential as although it is considered a benign lesion, it can, rarely, show malignant transformation to an undifferentiated embryonal sarcoma.³ On ultrasound of cystic mesenchymal hamartoma, there is evidence of echogenic variably thick septa and also echogenicity of any solid areas of the lesion. The CT

appearance is of a complex cystic mass with contrast induced enhancement of the septa and any solid areas. On MRI, the T1-weighted images show a variable signal of cystic areas while the T2-weighted images show a fluid signal. The solid areas of the lesion are generally hypointense in comparison to background liver and the contrast-induced enhancement of septal and solid areas is generally mild in degree.⁴ These features are reflected in the histology of mesenchymal hamartoma which shows stromal tissue rich in fibroblast-like cells and containing bile duct structures, hepatocytes and multiple cysts. The latter are the result of dilated bile ducts or lymphatics or from cystic degeneration of the stroma. In undifferentiated embryonal sarcoma, the solid areas are obvious on ultrasound and will enhance on the delayed phase of CT and MRI scans following contrast administration.⁵

Biliary cystic neoplasms are classically seen in middle-aged patients and can usually be differentiated from a simple hepatic cyst by the presence of septation, multiloculation and irregularity of the wall on ultrasound and CT. There is contrast-induced enhancement of the wall and septa. Furthermore, in contrast to simple hepatic cyst, these lesions can be associated with upstream bile duct dilation and arterial phase high attenuation of perilesional hepatic tissue.⁶ Serum or cyst fluid CA 19-9 levels may be raised though this is not entirely a specific feature. Within the umbrella category of biliary cystic neoplasm, the differentiation of biliary cystadenoma from biliary cystadenocarcinoma can be challenging. However, the presence of coarse calcifications, mural nodules and luminal papillary projections are associated with biliary cystadenocarcinoma. Furthermore, the radiological differentiation is not usually vital to the patient's management as the surgical management of biliary cystadenoma and biliary cystadenocarcinoma is similar (given that biliary cystadenoma is a premalignant lesion).

Examination of aspirated contents of simple hepatic cyst is generally not needed for diagnosis. However, if such an examination is performed, the fluid should be sterile and free of malignant cells.

Active treatment of simple hepatic cyst is only required in the event of symptoms. Aspiration only as a treatment modality is not an effective intervention: in a study of 15 sole aspiration procedures on 13 patients, there was a 100% recurrence rate to their original size.⁷ However, aspiration together with the use of a sclerotherapy agent such as alcohol or minocycline hydrochloride is more effective; a review of 91 cases of simple cysts treated with alcohol sclerotherapy (involving multiple investigators) recorded recurrence in only three cases.⁸ However, in many cases treatment resulted in only partial volume reduction of cysts and one cyst failed completely to respond to treatment. Sclerotherapy techniques have therefore yet to gain widespread acceptance and the management of choice of a simple hepatic cyst is considered to be surgical. The surgical options include laparoscopic deroofing of the cyst, open deroofing of the cyst and open resection. In deroofing procedures, the presence of residual cyst lining is considered not to be of concern as the production of further cyst fluid from the residual epithelium can drain into the peritoneal cavity. While there is considerable variation in the literature in the reported morbidity and recurrence rates of the various surgical approaches, the general consensus is that laparoscopic deroofing is the most appropriate initial surgical management for most patients. In one Argentinian series, 66 patients who underwent laparoscopic deroofing had morbidity and recurrence rates of 2% and 2%, respectively. This compares very favourably with 37 patients in the same series who underwent open deroofing of simple hepatic cyst and had

morbidity and recurrence rates of 29% and 8%, respectively.⁹ In a separate Scottish series, 48 patients who underwent laparoscopic deroofing had morbidity and recurrence symptoms of 15% and 29%, respectively compared to morbidity and recurrence symptoms of 30% and 36%, respectively in the 11 patients who underwent an open deroofing procedure and morbidity and recurrence symptoms of 42% and 25%, respectively in the 12 patients who underwent open resection.¹⁰ In a large German and Greek series of 59 patients of whom 51 had simple hepatic cysts, the recurrence rate for the 42 patients who underwent laparoscopic intervention (of whom 41 underwent cyst reroofing) was 5% compared to a recurrence rate of 18% for the 17 patients who had open surgical intervention (of whom 14 underwent open cyst deroofing).¹¹ In a smaller series of 12 patients of whom 9 had simple hepatic cysts, the morbidity rate for laparoscopic deroofing was reported as 25% and the recurrence rate as 0%.¹² The weight of evidence therefore favours laparoscopic deroofing as the treatment of choice in the management of simple hepatic cyst. There may, however, be a role for open resection as a second-line procedure where initial laparoscopic deroofing has been unsuccessful, where there are perioperative/postoperative complications following laparoscopic intervention requiring open surgery or potentially in the small cohort of patients in whom the multitude or location of simple cysts renders laparoscopic deroofing unsuitable.^{4 5}

In summary, the case described above represents a rare example of large symptomatic simple hepatic cyst in a 4-year-old girl. To our knowledge the case represents the largest hepatic simple cyst ever described in the 0–16 years paediatric age group in Europe and the second largest in the world. Diagnosis of these lesions is mainly based on imaging studies and the management of choice is laparoscopic deroofing of the cyst.

Learning points

- ▶ Simple hepatic cysts are rare in children and tend to be asymptomatic.
- ▶ They form part of the differential diagnosis for upper abdominal cyst especially in the right upper quadrant.
- ▶ Imaging studies are the main modality of investigation.
- ▶ Aspiration alone is not advised.
- ▶ Laparoscopic deroofing is the management of choice.

Contributors KL identified the case and managed the patient. The article was primarily written by RB but various drafts were reviewed by KL who suggested ways it could be improved.

Competing interests None.

Patient consent Obtained.

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