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Hepatic epidermoid cyst

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ABSTRACT

We describe the extraordinary finding of a hepatic epidermoid cyst in a 5-year-old patient, treated successfully with laparoscopic deroofing and mucosal stripping. The pathologic examination revealed a cyst with the same features of a true epithelial splenic cyst whose origin is still controversial, even though a coelomic derivation is the most accredited hypothesis.

A cyst in this anatomic district with such characteristics has never been described in existent literature.

1. Introduction

We describe the extraordinary case of a 5-year-old female patient brought to our attention for a symptomatic cyst attached to the liver (Vth segment) and the biliary tree.

After an extensive pre-operative work-up including both radiologic and laboratory investigations, considering all possible differential diagnosis, we decided to deroof the outer wall of the cyst and to strip the remaining mucosa with a laparoscopic approach. The definitive histologic and immunohistochemical examination identified a cyst lined with a stratified epithelium, of the same characteristics of splenic cysts whose origin is still controversial. We agree with the most accredited hypothesis according to which these cysts have a coelomic origin, through a metaplastic process of the mesothelium.

A cyst in this anatomic district with such features has never been described in existent literature.

2. Description of the case

A 5-year-old female patient was brought to our attention with acute abdominal pain in the upper left quadrant. She did not present any relevant medical history, although routine prenatal screening was missing. No traumatic episodes were reported.

Lab investigations were normal (no signs of cholestasis, no hepatic cytolysis, no pancreatic enzyme anomalies). Ultrasounds showed the presence of a 5 cm cyst attached to gallbladder and the liver (Vth segment).

MRI showed the presence of a roundish formation in the hepatic hiulum with net margins of about 47 × 47 mm, with an evident fluid level in the context, with signal intensity compatible with biliary content; according to our radiologists this finding appeared compatible as first hypothesis to a diverticular extroflexion of the common hepatic duct (Todani's type II of choledochal cyst) (Fig. 1).

Anyway, differential diagnosis included gallbladder duplication, intestinal duplication, choledochal cyst, pyogenic liver abscess, simple hepatic cyst.

After routine pre-operative investigations and serological tests for cystic echinococcosis (which resulted negative), we decided a laparoscopy approach. Attachment to the medial part of the gallbladder and the liver were confirmed; relation to the gastro-intestinal system were not seen (Fig. 2a). From the cyst we first drained 65 ml of serous yellowish material followed by purulent material.

Communication of the cyst with the biliary tree was excluded by intraoperative laparoscopic cholangiography. Furthermore, in this study, the biliary tree appeared to be regular (no dilatation seen) (Fig. 2b).

The free wall of the cyst was then excised (deroofed). After wide deroofing (Fig. 3a), frozen pathological examination of the cystic wall for extemporary analysis was also performed to rule out intraoperatively neoplastic cyst. The extemporaneous biopsy showed a fibrous wall with no muscular layer and the presence of epithelial lining on the inner surface of the cyst. The absence of muscular layer excluded an intestinal duplication or a biliary diverticulum.

The part of the cyst tenaciously attached to the liver and the biliary structure was not removed; we preferred to perform a mucosal stripping of the remaining cyst (Fig. 3b).

We decided not to use an argon beam coagulator, as we usually do for simple hepatic cysts, because of the proximity and the adhesion to the biliary tree.

An abdominal drain was left in place and removed on the 2nd post-

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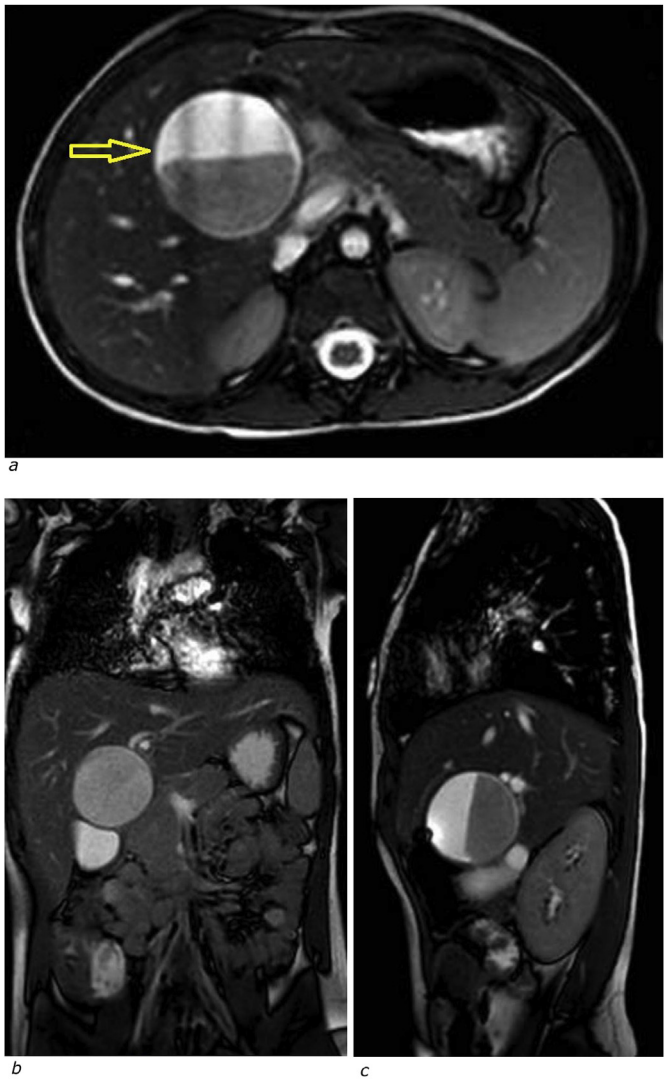

Fig. 1. a) MRI (axial section); b) MRI (frontal section); c) MRI (sagittal section). MRI showed the presence of a roundish formation in the hepatic hilum with net margins of about 47 × 47 mm, with an evident fluid level in the context, with signal intensity compatible with biliary content.

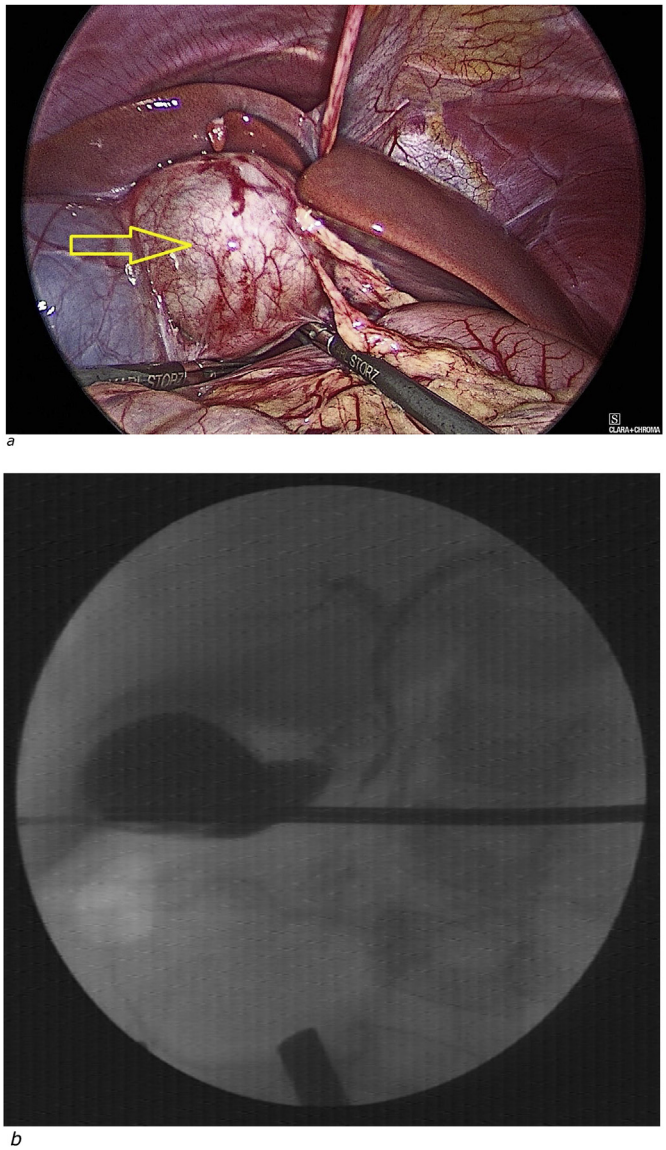

Fig. 2. a) At laparoscopic exploration, attachment to the medial part of the gallbladder and the liver were confirmed; relation to the gastro-intestinal system were not seen; b) Communication of the cyst with the biliary tree was excluded by intraoperative laparoscopic cholangiography. Furthermore, in this study, the biliary tree appeared to be regular (no dilatation seen).

operative day (no significant output registered). The post-operative course was uneventful (no complications reported). The patient was discharged on the 4th operative day.

The definitive histologic report of the stripped mucosa described the coexistence of both stratified squamous and transitional epithelium and the presence of muciparous cells. (Fig. 4).

Immunohistochemical analysis showed the cyst to be negative for calretinin (mesothelial marker) and GATA 3 e CK 20 (markers for urothelial lining); it resulted positive for CK 14 e CK 19 (full-thickness positive), p63 only on the basal layer (indicating squamous differentiation) and for BerEp 4 and CK 7, only on the inner surface and not on the basal layer (Fig. 5).

In conclusion, the cystic wall was lined by both stratified squamous and transitional epithelium, lying directly on fibrous tissue.

On the ultrasonographic follow-up at 1, 6 and 12 months after surgery, no signs of recurrence have been identified.

3. Discussion

This cyst had the same features of epithelial splenic cysts.

To date the main subject of controversy is whether epithelial splenic cysts arise from the peritoneal mesothelium or result from the developmental displacement of epithelial tissue.

In favour of the hypothesis, at present the most accredited, Morgenstern reported that mesothelial, transitional, and “epidermoid” epithelium can be found in different portions of single splenic cysts, suggesting that transitional and “epidermoid” epithelium are derived from a more or less complete metaplasia of mesothelium. Stratified squamous epithelium of epidermoid cysts would derive from metaplasia of mesothelium, perhaps because of pressure and stretching forces during cystic expansion.

It could well be that metaplastic mesothelium is the “motherboard” source for all sorts of cells lining congenital cysts.

Mesothelial cysts originate from invagination of capsular peritoneal mesothelium or collection of peritoneal mesothelial cells trapped in splenic sulci. As a matter of fact, in our case, something similar had probably happened but unprecedently in another anatomic region.

From other studies instead, it emerges that epithelial splenic cysts are either of teratomatous derivation or originate from inclusion of fetal

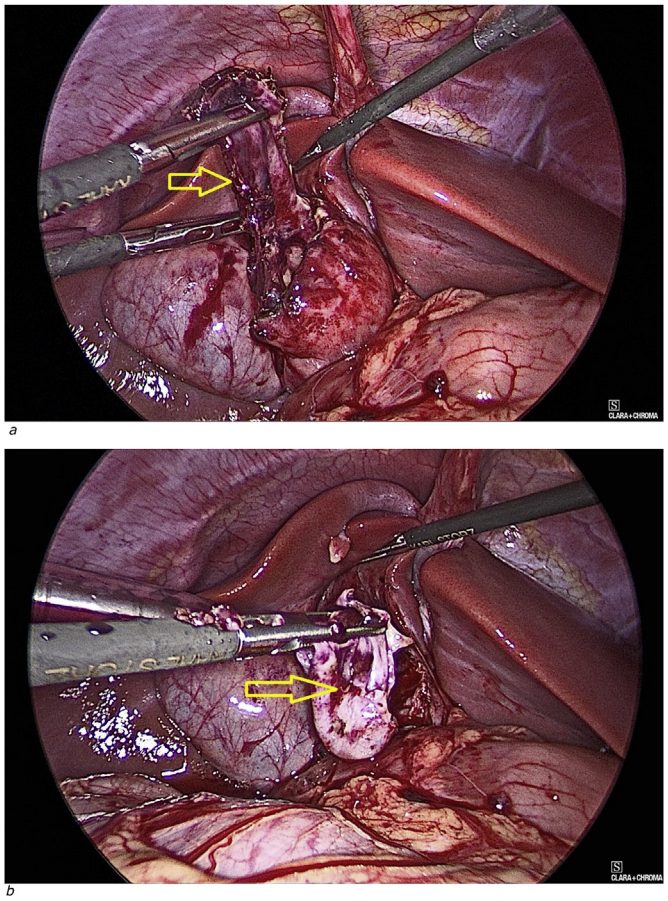

Fig. 3. a) excision (deroofing) of the free wall of the cyst was then excised; mucosal stripping of the remaining cyst.

squamous epithelium [1,2].

In patients with simple liver cysts, it is generally agreed that laparoscopic unroofing offers the best balance between efficacy and safety. We applied the same concept for our case, even though the cyst was way different from a simple liver cyst. The latter are lined by biliary-type epithelium and perhaps result from progressive dilatation of biliary microhamartomas. Because these cysts seldom contain bile, the current hypothesis is that the microhamartomas fail to develop normal connections with the biliary tree [3].

The principal laparoscopic technique currently in use is deroofing. In addition to wide deroofing of the cyst wall, destruction of the epithelium lining in the residual cystic cavity using an argon beam coagulator is the most important factor in avoiding cyst recurrence and perhaps malignant transformation [4]. We preferred not to use an argon beam coagulator, as we usually do for simple hepatic cysts, because of the proximity and adhesion to the biliary tree. Ablation of the cyst lining in the residual fenestrated cystic cavity was achieved by mucosal stripping in order to eliminate epithelial secreting cells and avoid cyst recurrence.

4. Conclusion

We describe the first case of hepatic epidermoid cyst of coelomic origin. The cyst shared the same features of a true epithelial splenic cyst. Laparoscopic deroofing of the cyst and mucosal stripping of the remaining cystic cavity resulted in successful treatment.

A cyst in this anatomic district with such features has never been described in existent literature.

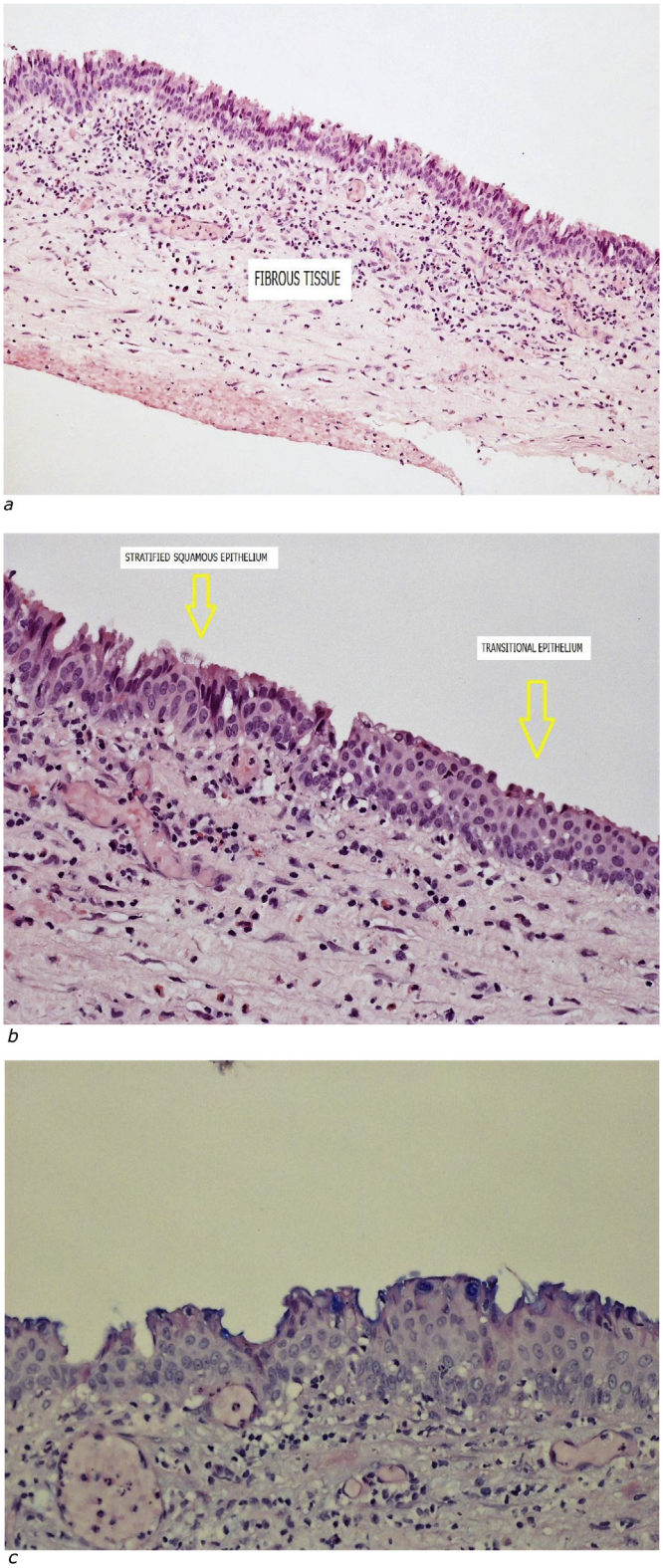

Fig. 4. Definitive pathological examination. 4a) hematoxylin-eosin staining 10x, full-thickness section: stratified epithelium lying directly on fibrous tissue; 4b) hematoxylin-eosin staining 20x: coexistence of both stratified squamous (on the left) and transitional epithelium (on the right); 4c) Alcian Blu staining revealing the presence of muciparous cells.

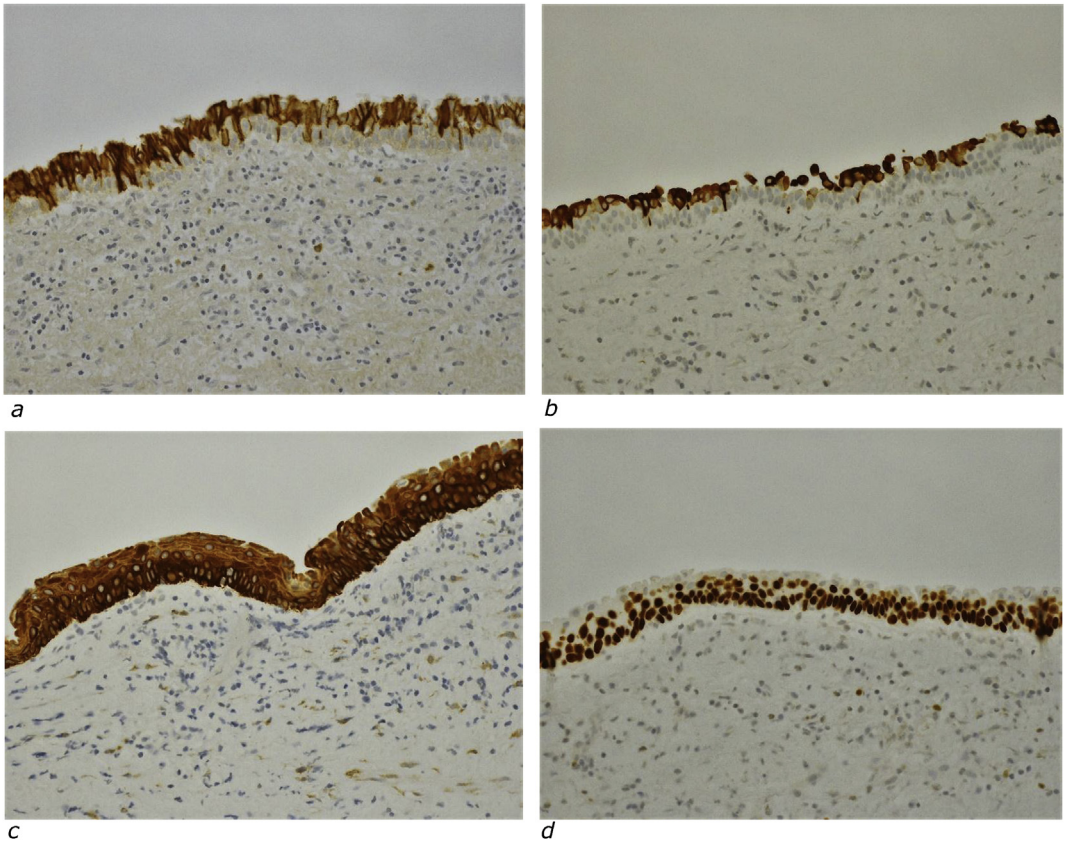

Fig. 5. Immunohistochemical analysis. 5a-b) BerEp 4 and CK 7 positive only on the inner surface and not on the basal layer. 5c) CK14 positive, full-thickness. 5d) p63 only on the basal layer indicating squamous differentiation.

Declarations of interest

None.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Conflict of interest

The following authors have no financial disclosures: DSN, LM, GT, SN, RG, LM.

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