Galbladder-associated ectopic liver: A rare finding during a laparoscopic cholecystectomy

Carlos Augusto Real Martínez a,*, Hermínio Cabral de Resende Júnior b, Murilo Rocha Rodrigues c, Daniela Tiemi Sato c, Cynthia Viegas Brunialti c, Rogério Tadeu Palma b

a Department of Surgery of São Francisco Medical University Hospital, Av. São Francisco de Assis, 218, CEP: 12916-900, Bрагança Paulista, São Paulo, Brazil
b Discipline of Gastrointestinal Surgery of Faculty of Medicine of ABC, Av. Príncipe de Gales, 821, CEP: 09060-650, Santo André, São Paulo, Brazil
c São Francisco University Medical School, Av. São Francisco de Assis, 218, CEP: 12916-900, Bрагança Paulista, São Paulo, Brazil

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Abstract

INTRODUCTION: Ectopic hepatic tissue is due to an uncommon failure of embryological liver development that is rarely described in the world medical literature. The incidence of ectopic liver (EL) has been reported to be anywhere from 0.24% to 0.47% as diagnosed at laparotomy or laparoscopy. We describe a case of EL adherent to the gallbladder, removed at laparoscopic cholecystectomy.

PRESENTATION OF CASE: A 37-year-old female was admitted for elective cholecystectomy having had an episode of acute cholecystitis provoked by gallstones. During the procedure, a 30 mm × 10 mm × 5 mm section of EL tissue attached to the anterior wall of the gallbladder was identified and removed by en-bloc excision during laparoscopic cholecystectomy. Histological examination confirmed the absence of malignant degeneration of the hepatic tissue. The patient recovered well postoperatively and was discharged the day after the operation. She was well when seen six months later.

DISCUSSION: EL has been reported in several sites, such as the gallbladder, gastrohepatic ligament, adrenal glands, esophagus, and thoracic cavity. EL is often clinically silent and discovered incidentally during abdominal surgical procedures or autopsies. Because patients with ectopic liver may suffer complications such as torsion, peritoneal bleeding, fatty change, and evolution to cirrhosis or malignant degeneration to hepatocellular carcinoma, any ectopic liver tissue needs to be correctly identified and removed.

CONCLUSION: Despite the rare occurrence of EL, it should be recognized and removed by the surgeon to prevent a higher risk of complications and malignant transformation.

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1. Introduction

Congenital abnormalities of embryologic liver positioning are rare. The first case was published in 1922,1 and a recent review of the literature revealed fewer than 80 published cases.2 The abnormalities are classified as accessory liver when the hepatic tissue is attached to the native liver and ectopic liver (EL) when the ectopic hepatic tissue is not directly connected to the liver.3-5 EL has been found both above and below the diaphragm, and the principal sites in the abdominal cavity are the gallbladder, gastrohepatic and umbilical ligaments, omentum, and stomach.6 EL can also develop above the diaphragm, and has been reported in the pleural cavity, mediastinum, lungs and heart.5-6 In the abdominal cavity, the gallbladder seems to be the anatomic structure most frequently affected.5-6 A review of the literature reveals at least 61 cases of well-documented EL attached to the gallbladder. Because of a lack of symptoms, its small dimensions and the difficulty in recognizing this unusual condition, a diagnosis of EL attached to the gallbladder is uncommon before surgery, and it is usually discovered during a surgical procedure within the abdominal cavity, or at autopsy.6 The possibility of malignant transformation into hepatocellular carcinoma (HCC) and the possible differential diagnoses of gallbladder wall masses make EL challenging for surgeons.3 We report a case of EL attached to the gallbladder wall that was discovered during an elective laparoscopic cholecystectomy for gallstones.

2. Presentation of case

A 37-year-old woman with a 7-month history of biliary colic and an episode of acute cholecystitis was referred for surgery. A previous abdominal ultrasound scan showed multiples gallstones without evidence of acute cholecystitis and a 3 cm localized thickening of the anterior portion of the gallbladder wall. The patient underwent elective laparoscopic cholecystectomy. During the operation, a smooth fragment of reddish-brown tissue was seen attached to the anterior surface of the gallbladder, which was not obviously inflamed (Fig. 1A). The mass was removed en-bloc with...
the gallbladder inside a plastic bag through the umbilical port-site. On macroscopic examination there was ectopic liver tissue measuring 30 mm × 10 mm × 5 mm attached to the visceral peritoneum of the gallbladder (Fig. 1B). The EL did not invade the full thickness of the gallbladder wall. On being opened, the gallbladder was seen to contain numerous brown-yellowish gallstones, 5–10 mm in diameter, attached to the luminal surface. The wall of the gallbladder was of normal thickness, and microscopic examination confirmed the presence of chronic inflammation with EL tissue adherent to the gallbladder serosa; there was sinusoidal congestion, mild steatosis and focal deposits of hemosiderin without malignant degeneration (Fig. 2A). Perl’s staining (hemosiderin deposits) confirmed the presence of focal iron deposits and staining with Masson trichrome showed a small amount of collagen in the EL tissue (Fig. 2B). The patient had an uneventful postoperative course and was discharged on the first postoperative day. She was well when seen for follow-up six months later.

3. Discussion

EL is a rare condition. The first published case dates back to 1922.1,5 The diagnosis is uncommon, and there are fewer than 80 documented cases of EL.2 The real incidence of EL attached to the gallbladder wall is difficult to assess but is reportedly 0.24–0.47% of the population though as with other sites, most of the cases of EL attached to the gallbladder are diagnosed at laparotomy, laparoscopy or during an autopsy. By 2007, 61 cases had been reported.4,6,8–14 Even though the gallbladder is the main site for the development of EL, this finding is still exceptional. In 1940 in a review of 5500 autopsies, only 0.05% had EL and in only three cases was this attached to the wall of the gallbladder. More recently, a review of 1060 laparoscopic procedures found EL attached to the gallbladder wall in three patients (0.28%).10 EL is most frequently described in Asia and to the best of our knowledge, our case is only the second case of EL attached to the gallbladder described in South America.10

Collan et al.3 divided EL tissue into four distinct categories: (a) EL that is not connected to the main liver and is usually attached to the gallbladder or intra-abdominal ligaments; (b) microscopic ectopic liver found occasionally in the gallbladder wall (as in the patient discussed here); (c) a large accessory liver attached to the “mother” liver by a stalk; and (d) a small accessory liver lobe attached to the main liver.

Several theories have been proposed to explain the presence of EL at different locations.9 Most researchers believe that the cause of EL is an aberrant migration during the embryologic development of the liver.4 During the fourth week in utero, the liver and biliary system originate from the hepatic diverticulum in the direction of the septum transversum. Abnormalities of this migration or a displacement of a portion of the cranial part of the hepatic diverticulum of the liver bud to other sites may be the main cause of EL.1,2 The close relationship of the developing hepatic parenchyma cell cords to the pars cystica may explain why ectopic hepatic tissue could be found in the wall of the gallbladder.1,2,9 Although the EL is usually

![Fig. 1.](image1) (A) Laparoscopic view of ectopic liver attached to the anterior surface of the gallbladder wall. (B) Gallbladder removed by laparoscopic approach containing ectopic liver with reddish brown coloration measuring 30 mm × 10 mm × 5 mm firmly attached to the peritoneal surface of the gallbladder wall with an increased vascular supply (white arrow).

![Fig. 2.](image2) (A) Photomicrograph cross-section of the gallbladder showing normal columnar epithelium without signs of malignity with a group of rounded cells containing cytoplasm filled with fat; the ectopic liver is attached to the serosa of the gallbladder (H&E × 40). (B) Photomicrograph cross-section of the gallbladder showing the presence of ectopic liver superficially adhered to the gallbladder wall, with the architecture of the liver tissue preserved, middle steatosis of the hepatocytes, scattered points of hemosiderosis and formation of collagen strips (trichrome of Masson staining × 100).
attached to the serosa of the gallbladder or lies within its wall, it can also occur in the gallbladder lumen. EL is sometimes associated with other congenital anomalies, such as biliary atresia, agenesis of the caudate lobe, omphalocele, bile duct cysts or cardiac anomalies; however, these abnormalities are not present when the heterotopic tissue is attached in the surface of gallbladder wall. EL attached to the gallbladder usually remains asymptomatic and is occasionally discovered during laparoscopy, as was the case with the patient in the present report. When symptomatic, the principal complaint is usually upper abdominal pain due to complications such as torsion, hemorrhagic necrosis, rupture or some form of compression by the mass due to malignant transformation to HCC. The differential diagnosis of lesions attached to the gallbladder includes other diseases that lead to a mass effect, such as carcinoma of the gallbladder, polyps, accessory liver, adenomyomatosis, hyperplastic lymph nodes and metastatic disease.

Detection of EL tissue before surgical intervention or autopsies by means of imaging studies is rare.1,10 This may be due to the small size of most EL, the lack of awareness of this unusual condition among radiologists, difficulty interpreting the imaging and the frequent lack of symptoms.2 The diagnosis of EL should be considered when radiologists identify a soft tissue mass on the gallbladder wall on imaging (whether ultrasound, CT scan, or MRI, or as an incidental finding during laparoscopy). Percutaneous biopsies should be avoided because of the risk of bleeding and the possibility of malignant degeneration to HCC. To the best of our knowledge, a preoperative diagnosis of gallbladder-associated EL was made in only two reported cases.2,4 In the patient described in the present report, the ultrasonographic examination before surgery showed only a slight thickening of the anterior wall of the gallbladder, insufficient to suggest EL.

The histological findings of EL tissue are similar to those of the liver proper, including regular lobules, a central vein, and normal portal spaces. In some cases, it is possible to observe an increase in the number of blood vessels in the outer surface of the gallbladder attached to the ectopic liver tissue similar to what was observed in our patient. As in the liver itself, EL attached to the gallbladder can show fatty infiltration, cholestasis, hepatitis, hemosiderosis, cirrhosis, or malignant degeneration to HCC. In our patient there was mild steatosis and posterior focal areas of hemosiderosis confirmed by specific staining for iron pigments.

There has been evidence to suggest that ectopic liver is at increased risk of developing HCC.9,15 Arakawa et al.17 found 21 cases in the literature, mostly from Japan, of HCC arising extrahepatically. These authors suggest that ectopic tissue is more susceptible to the development of malignancy because it does not have a complete vasculature or ductal system like a normal liver, and is perhaps functionally impaired. This altered hepatic function may lead to chronic inflammation or cirrhosis, which increases the possibility of developing HCC. Yamashita et al.18 reviewed 70 cases of ectopic liver reported in the literature before 1986, including nine cases of HCC originating in EL tissue. Of 48 cases (excluding those localized to the gallbladder), 22 developed HCC.17 Contrasting with only one of 42 cases of EL attached to the gallbladder, a possible explanation for this difference is that EL attached to the gallbladder is an anomaly occurring later during the development of the biliary bud and is therefore well differentiated.9 Despite the presence of steatosis and areas of hemosiderosis, we could not find any evidence of malignant degeneration in the patient in this report. However, due to the perceived risk of malignant degeneration in EL generally, resection of the EL en bloc with the gallbladder is strongly recommended.2

With the increase in laparoscopic cholecystectomy, more cases of EL attached to the gallbladder have been identified in recent years and are easily removed using this approach, preferably inside a bag, and then carefully examined by a pathologist. If the histopathological examination confirms the presence of an invasive HCC, a further surgical intervention should be considered, to extend the hepatic margins of resection and perform a regional lymphadenectomy.

4. Conclusion
EL attached to the gallbladder wall is due to an uncommon failure of embryological development of the liver that is usually asymptomatic. EL carries an increased risk of malignant degeneration to HCC and therefore EL should be recognized (especially at laparoscopic cholecystectomy), removed and examined histologically.

Conflict of interest
All authors of this article declare no conflict of interest.

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Ethical approval
Written informed consent was obtained from the patient for publication of this case report and accompanying images. This article was approved by Ethical Committee of Sao Francisco University (process No. 126.340).

Author contributions
Carlos Augusto Real Martinez contributed toward study design, writing and final revision. Herrminio Cabral de Resende Júnior contributed toward data analysis andd osurgical team. Murilo Rocha Rodrigues contributed toward data collections, review of the literature. Daniela Tiemi Sato contributed toward data collections, surgical team. Cynthia Viegas Brunialti contributed toward histopathological analysis and histochemical staining interpretation. Rogerio Tadeu Palma contributed toward surgical team, writing, data analysis.

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