



Gallbladder-associated symptomatic hepatic choristoma: Should you resect?

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ABSTRACT

INTRODUCTION: Hepatic choristomas or ectopic livers are uncommon, and occur due to a failure of embryological liver development. They pose a risk of carcinogenesis, with transformation to hepatocellular carcinoma (HCC) being described in the literature (Arakawa et al., 1999). It is often a silent clinical finding that can occur anywhere in the body and is usually diagnosed incidentally during abdominal surgical procedures or autopsies (Eiserth et al., 1940). We present the case of a patient with a symptomatic ectopic liver that was detected preoperatively, and removed laparoscopically with the gallbladder.

PRESENTATION OF CASE: A 73-year-old lady was referred to our unit for a gallbladder tumor on ultrasound which was done for biliary colic. Tumor markers were normal. Computed tomography (CT) scan showed an enhanced soft tissue lesion measuring about 3 × 1.5 cm interposed between the gallbladder and liver. Laparoscopic exploration revealed a bean-shaped hepatic choristoma attached to the liver on the medial wall of the gallbladder. The lesion was removed by en-bloc resection during laparoscopic cholecystectomy and extracted carefully in an endobag. Histopathological examination confirmed the absence of carcinogenesis.

DISCUSSION AND CONCLUSION: Hepatic choristomas (HC) are a rare entity, usually identified during abdominal surgeries. It had been reported in several studies with different presentations. Awareness of this unexpected finding and familiarity of its potential complications and carcinogenesis will improve care delivery when encountered. Surgical treatment should be considered when the choristoma is not attached to the liver, in light of its potential transformation into HCC.

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1. Introduction

Hepatic choristomas or ectopic livers, also known as hepar succenturiatum, are an extremely rare developmental anomaly that can be found anywhere in the body, and is usually located in the abdomen, retroperitoneum and the chest. The incidence was reported to be 0.05%, with 3 cases in a series of 5500 autopsies in 1940, but more modern studies document a higher incidence during laparoscopic surgery, varying between 0.47% and 0.7% [2,3]. The natural course of hepatic choristomas is unpredictable. Ectopic livers can undergo fatty change, haemosiderosis, cholestasis or cirrhosis. They are also at increased risk of carcinogenesis [4]. A review of the literature reveals at least 65 cases of well-documented gallbladder associated hepatic choristomas, with few documented cases diagnosed preoperatively [5–7]. We describe a case of suspected symptomatic ectopic liver tissue, which was

removed laparoscopically to prevent any potential risk of malignant transformation.

2. Presentation of case

In our manuscript, we present a 73-year-old lady with a background history of diabetes mellitus and hypertension. She was referred to our hepatobiliary unit for a gallbladder tumor that was diagnosed on ultrasound, which was done for right upper quadrant postprandial abdominal pain (Fig. 1). The patient was investigated thoroughly, with liver function tests and tumor markers found to be within normal values. A triphasic computed tomography scan (CT) noted a normal liver, with absence of abnormal gallbladder findings. It also showed an enhanced soft tissue lesion with the same density as the liver, measuring about 3 cm × 1.5 cm, interposed between the gall bladder and the liver (Fig. 2). A Gallbladder polyp, tumor as well as lymph node were ruled unlikely, with the most suspect diagnosis being that of a hepatic choristoma. Although the risk of malignant transformation of a gallbladder associated hepatic choristoma into hepatocellular carcinoma (HCC) is low, a decision to proceed for surgical excision was made due

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Fig. 1. Ultrasound liver demonstrating a lesion inside the gallbladder.

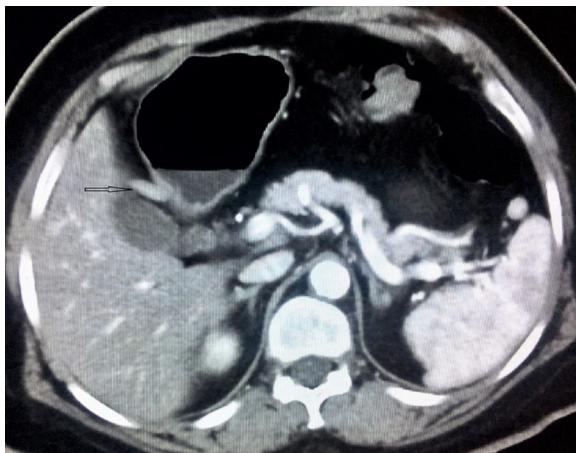


Fig. 2. CT scan showing an enhanced soft tissue lesion with the same density as the liver, measuring about 3×1.5 cm, interposed between the gall bladder and the liver.

to the patient's symptoms. We decided to perform a diagnostic laparoscopy, where surgical exploration revealed a bean shaped liver choristoma lying on the medial wall of the gallbladder with an attachment to the liver (Fig. 3). The lesion was removed by en-bloc resection along with cholecystectomy with extraction of the specimen in an endobag. Histopathological examination confirmed the suspected finding of liver tissue, with fatty infiltration and the absence of carcinogenesis or any gallbladder pathology. The patient had an unremarkable post-operative course and was discharged the following day in good condition. On follow-up her right upper quadrant was completely disappeared.

3. Discussion and conclusion

Hepatic choristomas have been previously classified into three entities which are separated into accessory liver lobes, ectopic nodules, or aberrant microscopic liver tissue. Liver tissue which is attached to the gallbladder, and is separate from the main liver, has been described by various names. Yet the correct term for this entity is a choristoma, which was first coined by Eugen Albrecht in 1904. Although the etiology is unclear, several possible mechanisms have been proposed which may explain ectopic liver at various sites. This includes the development of an accessory lobe with atrophy of the original bridge to the main liver, or migration of pars hepatica to the

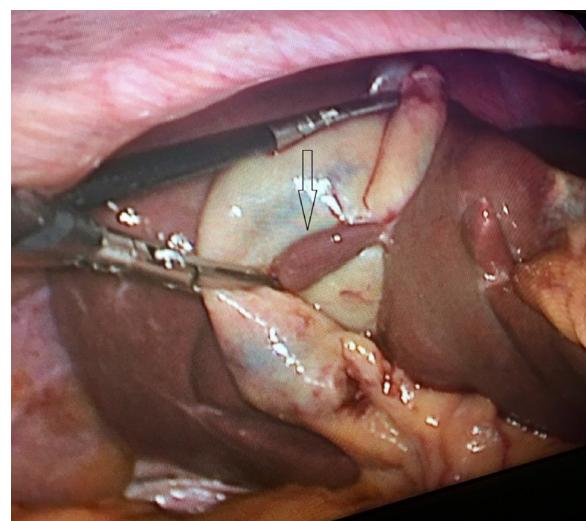


Fig. 3. Intraoperative finding of hepatic choristoma.

rudiment of various organs [8]. Ectopic liver lobes are commonly detected in perinatal cases, with a reported incidence of 11.5% [9]. However, they are rarely found to persist into adulthood [10].

There have been several proposed theories as to the cause of development of ectopic liver tissue at various locations within the body, such as atrophy or regression of the original bridge to the main liver, migration during embryological development of the liver, dorsal budding of hepatic tissue before closure of the pleuroperitoneal canals, trapping of hepatocyte-destined mesenchyma in different areas or entrapment of nest cells in the region of the foregut following closure of the diaphragm or umbilical ring [11].

The most common location of liver ectopia is associated with the gallbladder, ranging from microscopic tissue to a few cm in size [4,5]. Ectopic liver can also be found above and below the diaphragm. The close relationship of the developing hepatic parenchymal cell cords to the pars cystica and early fetal duodenum explains why ectopic liver tissue could be found in the wall of the gallbladder, the gastrohepatic ligament, the umbilical cord, the adrenal glands, the diaphragm, the pancreas, the pylorus and the splenic capsule if a portion of the pars hepatica is displaced. Dorsal budding of hepatic tissue before closure of the pleuroperitoneal canals may explain how ectopic liver develops in the thoracic cavity in locations such as esophagus, pericardium, and in the pleural cavity [6].

Hepatic choristomas are occasionally associated with other congenital anomalies such as biliary atresia, agenesis of the caudate lobe, omphalocele, bile duct cyst or cardiac anomalies, but not when the heterotopic tissue is in the gallbladder [12,13]. Although the ectopic tissue is usually attached to the serosa of the gallbladder or lies within its wall, it can also occur in the gallbladder lumen [14].

Hepatic choristomas are rarely symptomatic. However, when they do occur, ectopic livers have been reported to cause recurrent abdominal pain due to torsion, compression of adjacent organs, intraperitoneal bleeding, as well as obstruction of the esophagus, portal vein, neonatal gastric outlet and pylorus [4,15]. Few cases of symptomatic ectopic livers have been reported in the literature, some of which were described in infants [16]. The number of reported cases of ectopic liver giving rise to acute symptoms was fewer still. Our patient presented with right upper quadrant pain in the absence of gallbladder pathology, her symptoms were relieved after surgery.

Hepatocytes in ectopic livers usually mimic normal hepatocytes and show relatively the same pathological pattern. Thus, ectopic liver tissue usually show normal histological architecture and are

subject to the same risk factors and pathological processes as native liver tissue [17]. Fatty infiltration and alpha-1-antitrypsin deficiency have been described in ectopic liver [18]. However, the development of HCC in a hepatic choristoma is of utmost significance. In fact, HCC can be observed in about 46% of ectopic liver tissue encountered outside the liver, with only 2.4% in gallbladder associated hepatic choristomas [19]. The high incidence of neoplastic change in ectopic livers is probably explained in that they possess different functional architecture with incomplete vascular and/or ductal systems. This results in longer exposure of ectopic liver tissues to carcinogenic substances thereby propagating its malignant transformation [1]. The reverse situation, wherein a HCC develops in the parent liver but not in the ectopic liver is exceedingly rare. Gallbladder associated hepatic choristomas may have a lower rate of malignant transformation as they occur later during biliary development and is well differentiated [20].

The vascular supply of a hepatic choristoma is an important consideration during operative planning to avoid vascular injury during dissection or traction of the specimen. With regards to gallbladder associated hepatic choristomas, there are mainly three different vascular supply patterns of gallbladder associated HC. (a) Artery arising from cystic artery (b) Vascular pedicle (with/without its own vein) arising from liver parenchyma substance (c) Vascular structures embedded in a mesentery lying from the hepatic site to ectopic liver tissue [21]. In one study Koh and Hunt reported a case of HC on the gallbladder wall in which its artery was derived from cystic artery and bile duct drainage to cystic duct [22]. Catani et al. reported that ectopic liver nodule seemed drizzled by an arterial branch that runs along the front part of gallbladder [23].

Detection of ectopic liver tissue before surgical intervention or autopsies by means of imaging studies is rare [7]. This may be due to the small size of most ectopic livers, the lack of awareness of this unusual condition amongst radiologists, difficulty interpreting imaging and the frequent lack of symptoms. The diagnosis of hepatic choristomas should be considered when radiologists identify a soft tissue mass on the gallbladder wall during imaging, whether by ultrasound, CT scan, magnetic resonance imaging or as an incidental finding during laparoscopy. Percutaneous biopsies should be avoided due to the risk of bleeding and the possibility of seeding the needle tract due to the risk of HCC. To the best of our knowledge, a preoperative diagnosis of gallbladder-associated hepatic choristoma was made in only three reported cases [5–7]. In the patient described in this report, the ultrasonographic examination prior to surgery showed thickening of the anterior wall of the gallbladder mimicking a lesion, insufficient to suggest a hepatic choristoma. Nevertheless, CT scan delineated its presence and excluded any other findings.

According to the literature, a pattern emerges wherein surgeons are prone to resect the ectopic tissue if encountered during cholecystectomy for symptomatic gallbladder to avoid surgical manipulation if seen incidentally during other procedures. Despite the low incidence of hepatic choristomas and its unlikely detection preoperatively, an awareness of this entity and its associated risk of carcinogenesis may lower the rate of misdiagnoses and complications. Resection of hepatic choristomas occurring outside of the liver should strongly be considered due to its high risk of transformation, whereas the management of gallbladder associated hepatic choristomas warrant further study due to its low risk in comparison.

Conflicts of interest

The authors declare no conflict of interest.

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Ethical approval

The case report was approved by the local ethic committee of the Al-Amiri teaching hospital. LEC-project number 52-2017.

Consent

Written informed consent was obtained from the patient and is available upon request. No patient identifying material was used in this manuscript.

Author contribution

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Analysis and case management: Salah Termos* and Ahmad Alsaleh.

Writing and revising of the final version of the manuscript: Salah Termos, Abdullah AlDuwaisan and Mohammad Alali.

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The work has been reported in line with the SCARE criteria [24].

References

- [1] M. Arakawa, Y. Kimura, K. Sakata, Y. Kubo, T. Fukushima, K. Okuda, Propensity of ectopic liver to hepatocarcinogenesis: case reports and a review of the literature, *Hepatology* 29 (1999) 57–61.
- [2] P. Eiserth, Beiträge zur Kenntnis der Nebenleber, *Virchows Archiv* 307 (1940) 307–313.
- [3] S. Sato, M. Watanabe, S. Nagasawa, M. Niigaki, S. Sakai, S. Akagi, Laparoscopic observations of congenital anomalies of the liver, *Gastrointest. Endosc.* 47 (1998) 136–140.
- [4] M. Watanabe, T. Matsura, Y. Takatori, K. Ueki, T. Kobatake, M. Hidaka, H. Hirakawa, S. Fukukimoto, Y. Shimada, Five cases of ectopic liver and a case of accessory lobe of the liver, *Endoscopy* 21 (1989) 39–42.
- [5] J. Lundy, E. Johnson, K. Edwards, D. Rivera, Laparoscopic management of gallbladder associated ectopic liver, *JLSL* 9 (2005) 485–487.
- [6] S. Hamdani, R. Baron, Ectopic liver simulating a mass in the gallbladder wall: imaging findings, *Am. J. Roentgenol.* 162 (1994) 647–648.
- [7] M. Catani, R. De Milito, F. Romagnoli, Ectopic liver nodules: a rare finding during cholecystectomy, *G. Chir.* 32 (2011) 255–258.
- [8] Y. Collan, A. Hakkiuoto, J. Hastgacka, Ectopic liver, *Ann. Chir. Gynaecol.* 67 (1978) 27–29.
- [9] Wang, Ectopic liver tissue in the gallbladder, *Zhonghua Gan Zang Bing Za Zui* 14 (2006) 369.
- [10] W. Parke, H. Settles, P. Bunger, R.E. van Demark, Malformations of the liver: some prenatal and postnatal developmental aspects, *Clin. Anat.* 9 (1996) 309–316.
- [11] A. Sakarya, Y. Erhan, H. Aydede, E. Kara, O. İlkgül, C. Ciftdogan, Ectopic liver (choristoma) associated with the gallbladder encountered during laparoscopic cholecystectomy: a case report, *Surg. Endosc.* 16 (July (7)) (2002) 1106.
- [12] J. Asada, S. Onji, Y. Yamashita, S. Okada, M. Morino, M. Kanoaka, Y. Ohta, Ectopic liver observed by peritoneoscopy; report of a case, *Gastroenterol. Endosc.* 24 (1982) 309–312.
- [13] J. Shapiro, L. Metlay, Heterotopic supradiaphragmatic liver formation in association with congenital cardiac anomalies, *Arch. Pathol. Lab. Med.* 115 (1991) 238–240.
- [14] T. Natori, S. Hawkin, M. Aizawa, T. Asai, Y. Kamed, K. Iky- yo- hashi, Intra-cholecystic ectopic liver, *Acta Pathol. Jpn.* 36 (1986) 1213–1216.
- [15] M. Hadda, A. Currie, M. Honeyman, Pyloric obstruction by ectopic liver tissue, *Br. J. Surg.* 72 (1985) 917.
- [16] Y. Tomooka, M. Torisu, T. Fujimura, N. Sakaguchi, A. Taira, Symptomatic accessory lobe of the liver associated with hyperthyroidism, *J. Pediatr. Surg.* 23 (1988) 1055–1056.

- [17] M. Dettmer, G. Cathomas, N. Willi, Alpha 1-antitrypsin retention in an ectopic liver, *Diagn. Pathol.* 28 (6) (2011) 16, <http://dx.doi.org/10.1186/1746-1596-6-16>.
- [18] I. Triantafyllidis, L. Papapavlou, N. Nikoloudis, A. Economou, E. Andreadis, M. Chrissidou, K. Georgakis, T. Chrissidis, Ectopic liver tissue attached to the gallbladder wall: a case report, *Cases J.* 2 (29) (2009) 6786, <http://dx.doi.org/10.1186/1757-1626-2-6786>.
- [19] M. Arakawa, Y. Kimura, K. Sakata, Y. Kubo, T. Fukushima, K. Okuda, Propensity of ectopic liver to hepatocarcinogenesis: case reports and a review of the literature, *Hepatology* 29 (1999) 57–61.
- [20] J. Griniatsos, A. Riaz, A. Isla, Two cases of ectopic liver attached to the gallbladder wall, *J. Int. Hepato-Pancreato-Biliary* 4 (2002) 191–194.
- [21] A. Bal, S. Yilmaz, B. Yavas, et al., A rare condition: ectopic liver tissue with its unique blood supply encountered during laparoscopic cholecystectomy, *Int. J. Surg. Case Rep.* 9 (2015) 47–50.
- [22] C.E. Koh, R. Hunt, Ectopic liver encountered during laparoscopic cholecystectomy, *Asian J. Surg.* 3 (2007) 227–229.
- [23] M. Catani, R. De Milito, F. Romagnoli, P. Mingazzini, V. Silvestri, V. Usai, Ectopic liver nodules: a rare finding during cholecystectomy, *G. Chir.* 5 (2011) 255–258.
- [24] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, the SCARE Group, The SCARE statement: consensus-based surgical case report guidelines, *Int. J. Surg.* 34 (2016) 180–186.

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