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Ampullectomy for an unexpected ampullary hamartoma in a heterotaxic patient

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ABSTRACT

INTRODUCTION: Heterotaxy designates rare congenital disorders of organ positioning in the thoracic and abdominal cavities, which can be associated with numerous anomalies, complicating the surgical management because of the loss of conventional anatomic landmarks.
PRESENTATION OF CASE: A 72-year-old man was found to have asymptomatic cholestasis. Further workup included computed tomography and magnetic resonance cholangiopancreatography that revealed anomalies of lateralization of digestive organs associated with intestinal malrotation and polysplenia, and a stone-like element in the main bile duct. Endoscopic retrograde cholangiopancreatography failed to extract the lesion. Laparotomy found no stone, but a polypoid tumor with ampullary implantation. Pancreaticoduodenectomy was judged unreasonable due to the presence of macroscopic cirrhosis and a complete ampullectomy was performed. Histopathological examination revealed a hamartomatous polyp.
DISCUSSION: The unusual angle of the duodenoscope in a left-sided duodenum may have contributed to the improper pre-operative diagnosis. Endosonography could have recognized the tissular origin of the lesion and prompted a more detailed preoperative planning. It was fortunate that the patient ended up receiving the appropriate treatment despite the absence of an adequate pre-operative diagnosis, as the option of performing an extended resection was ruled out due to the presence of cirrhosis.
CONCLUSION: Although heterotaxy leads to increased technical difficulties in performing usual endoscopic and surgical procedures, it can be safely managed by experienced surgeons as illustrated by the present case. Imaging modalities have limited sensitivity in the diagnosis of small ampullary tumors. As false-negatives are likely to occur, this possibility should guide the choice of the best operation.

1. Introduction

Heterotaxy designates rare congenital disorders of organ positioning in the thoracic and abdominal cavities.[1,2] More precisely, situs inversus totalis defines the full transposition of organs in a mirror image, while situs ambiguus includes any defect of lateralization along the cephalo-caudal axis.[1] Numerous other anomalies may be associated with heterotaxy, in particular concerning the digestive tract, complicating the surgical management because of the loss of conventional anatomic landmarks. Many publications report conventional surgical procedures performed successfully in patients with disorders of laterality, but only a few case reports relate the management of ampullary tumors in the context of these particular anatomic configurations. To our knowledge, we report the first case of limited surgical management of a sporadic ampullary hamartoma in the context of a situs ambiguus associated with bowel malrotation and polysplenia.

2. Case report

A 72-year-old man originating from sub-Saharan Africa was diagnosed with asymptomatic cholestasis during a routine outpatient check-up. A chest radiograph showed a normal left-sided heart. A subsequent computed tomography revealed abnormal lateralization of stomach, duodenum and liver, associated with intestinal malrotation and polysplenia (Fig. 1). There was prominent intra- and extrahepatic bile duct dilatation, which led to magnetic resonance cholangiopancreatography, showing an obstructive 8 mm element in the main bile duct associated with gallbladder stones (Figs. 2 and 3), suspicious of an impacted stone. Endoscopic retrograde cholangiopancreatography with sphincterotomy was performed three times, once in association with extra-corporeal short wave lithotripsy, but failed to extract the lesion. Endoscopic biopsies of the ampulla were unremarkable. A biliary stent was left in place and surgical exploration for failed bile-duct stone extraction was then decided.

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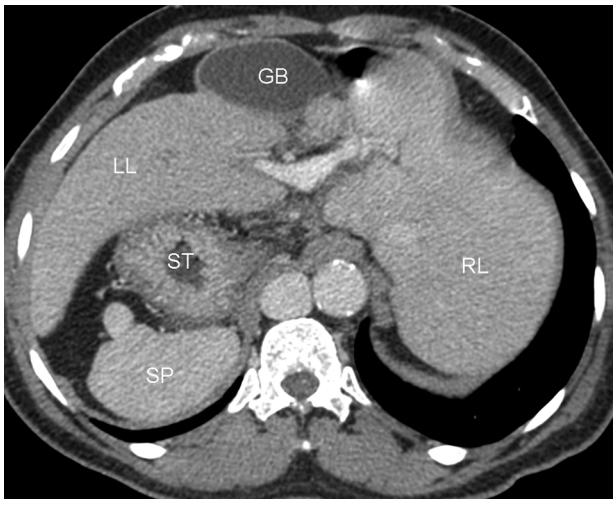

Fig. 1. Axial computed tomography showing anomalies of lateralisation of abdominal organs: LL = morphologic left lobe of liver; RL = morphologic right lobe of liver; GB = midline gallbladder; ST = right-sided stomach; SP = right-sided spleen.

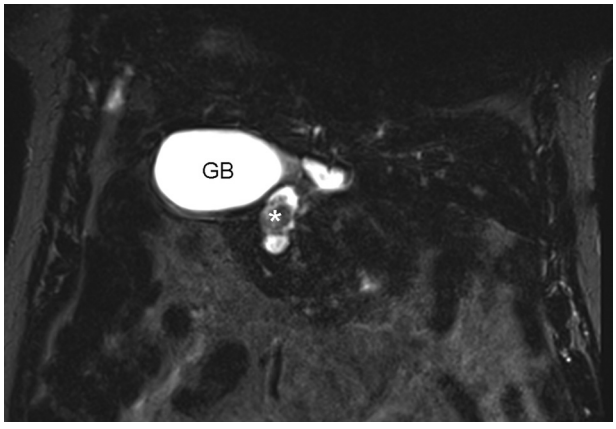

Fig. 2. Coronal magnetic resonance cholangiopancreatography suggesting a stone-like lesion (*) in the ampulla of Vater. GB = gallbladder.

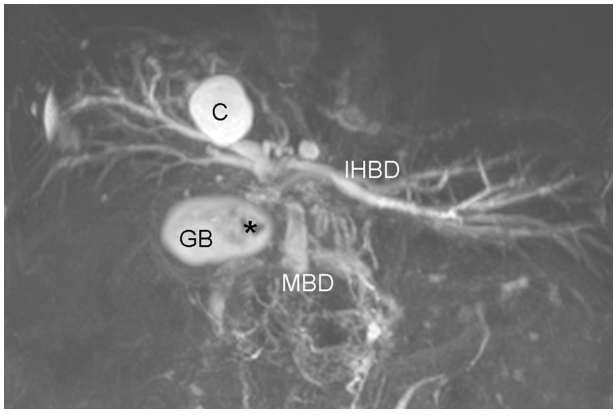

Fig. 3. Coronal magnetic resonance cholangiopancreatography: IHBD = slightly widened intrahepatic bile ducts; C = hepatic cyst; MBD = main bile duct; GB = gallbladder with stones (*) in the Hartmann's pouch.

Laparotomy was performed, facing situs ambiguus with a left-sided liver, a gallbladder situated on the midline, a duodenum displaced on the right, in association with polysplenia and bowel malrotation, represented by a common mesenteric implantation with the entire colon located in the right hemiabdomen. The liver

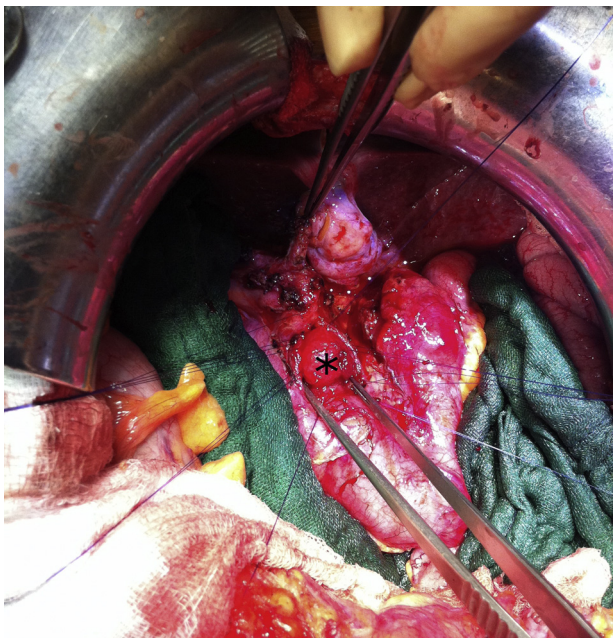

Fig. 4. Duodenectomy showing polypoid tumor (*) with ampullary implantation.

was hard and nodular, suggesting cirrhosis. After cholecystectomy, a complete Kocher maneuver was performed, complicated by the presence of a duodenal diverticulum and a short duodenal papilla located at approximately one centimeter from the pylorus. Exploration of the main bile duct by a vertical choledocotomy found no stone, but a polypoid tumor with ampullary implantation. Pancreaticoduodenectomy was judged unreasonable due to the presence of cirrhosis, instead of which a complete ampullectomy through a duodenal incision was performed (Fig. 4). The main bile duct was sutured around a T-tube. Derotation of the bowel with section of Ladd's bands was then achieved. Histopathological examination revealed a hamartomatous polyp of the juvenile subtype without dysplasia. Liver cirrhosis of undetermined origin was confirmed on the peroperative liver biopsy. The postoperative course was complicated by an early incisional dehiscence that required repair, but was otherwise uneventful.

3. Discussion

Tumors of the ampulla of Vater are rare, with a yearly incidence of less than 1/100,000,[3] and are mostly represented by adenomas.[4] Non-adenomatous lesions, such as lipomas, lymphangiomas, hemangiomas, leiomyofibromas, and neurogenic tumors account for only 20% of ampullary tumors[5–7]; hamartomas are the most rare, with 7 sporadic cases described to our knowledge.[8–12]

The literature reports only a few studies on patients suffering from digestive cancers associated with disorders of laterality. Concerning ampullary tumors, the experience is even more anecdotal, since to date only two publications reported ampullary carcinoma as part of a situs inversus totalis, and this without evidence of a possible association between both diseases.[13,14] To our knowledge, we report the first case of a benign ampullary tumor in a patient with situs ambiguus.

Definitive treatment of ampullary tumors of all histologic types remains complete surgical excision. The extent of surgical resection, however, is primarily dictated by the presence or absence of invasive carcinoma. This differentiation is difficult to establish during the preoperative period due to the limited sensitivity of conventional imaging techniques in noninvasive tumor staging.[15] In addition, endoscopic biopsies have a high rate of false-negative

results, particularly because of inaccuracies in sampling.[4,16] Perioperative frozen section histopathological analysis thus remains mandatory in determining the extent of resection.[17] There is no evidence suggesting that the preoperative evaluation is rendered complicated by the presence of heterotaxy.[18]

Pancreatoduodenectomy is the operation of choice when an invasive ampullary carcinoma is suspected or when preoperative frozen-section histopathological analysis is not available.[19,20] Ampullectomy is associated with less mortality and morbidity than pancreatoduodenectomy and remains indicated in selected patients (such as ours) who are at high risk in maximally invasive operations,[21] but is not a safe alternative in healthier patients due to high recurrence rates if the tumor is malignant.[22] Ampullectomy is however an accepted treatment for presumed benign lesions,[19–23] with the limits of preoperative histological analysis mentioned above.

In the present case, the finding of an ampullary tumor instead of a stone was a surprise. The unusual angle of the duodenoscope in a left-sided duodenum may have contributed to the false preoperative diagnosis. Endosonography would have avoided the erroneous presumptive diagnosis, prompted a more detailed preoperative planning, and should be used liberally in complex patients, like those with heterotaxy. It was fortunate that the patient received the correct operation as, despite the absence of an adequate pre- and peroperative diagnosis, the option of performing an extended resection was ruled out due to the presence of cirrhosis. No frozen section histopathological analysis was therefore performed.

4. Conclusion

Although heterotaxy leads to increased technical difficulties in performing usual endoscopic and surgical procedures, it can be safely managed by experienced surgeons, as illustrated by the literature and the present case. Imaging modalities have limited sensitivity in the diagnosis of ampullary tumors, as well as endoscopic biopsies. As false-negatives are likely to occur, this possibility should guide the choice of the best operation, even if the possibility of a more favorable histological diagnosis should be borne in mind, and confirmed by frozen-section examination if the answer is going to influence the choice of the surgical procedure.

Conflict of interest statement

The authors disclose no conflict of interest.

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Ethical approval

The patient passed away due to an unknown cause in his home country several months after his hospitalisation. No consent for the present case report could therefore be obtained. His family remained unreachable by conventional means.

Author contributions

All the authors have participated in acquisition of data, critical revision and final approval of the article, and gave their overall responsibility. Meyer J., Rossetti A., and Majno P. had written the article.

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References

1. Kosaki K, Casey B. Genetics of human left–right axis malformations. *Seminars in Cell & Developmental Biology* 1998;**9**(1):89–99.
2. Sutherland MJ, Ware SM. Disorders of left–right asymmetry: heterotaxy and situs inversus. *American Journal of Medical Genetics Part C: Seminars in Medical Genetics* 2009;**151C**(4):307–17.
3. Hsing AW, et al. Rising incidence of biliary tract cancers in Shanghai, China. *International Journal of Cancer* 1998;**75**(3):368–70.
4. Beger HG, et al. Tumor of the ampulla of Vater: experience with local or radical resection in 171 consecutively treated patients. *Archives of Surgery* 1999;**134**(5):526–32.
5. Denjoy R. Benign tumors of Vater's ampulla. *Journal de Chirurgie* 1968;**95**(2):211–26.
6. Chu PT. Benign neoplasms of the extrahepatic biliary ducts; review of the literature and report of a case of fibroma. *Archives of Pathology* 1950;**50**(1):84–97.
7. Sobol S, Cooperman AM. Villous adenoma of the ampulla of Vater. An unusual cause of biliary colic and obstructive jaundice. *Gastroenterology* 1978;**75**(1):107–9.
8. Bujanda L, et al. Chronic anemia and intestinal pseudoobstruction as presentation form of hamartomatous polyp of the Vater's ampulla. *Revista Espanola de Enfermedades Digestivas* 1997;**89**(8):644–6.
9. Moyana TN, Miller GG, Keith RG. Sporadic ampullary hamartoma simulating cancer. *Canadian Journal of Surgery* 1997;**40**(3):227–30.
10. Venu RP, et al. Ampullary hamartoma: endoscopic diagnosis and treatment. *Gastroenterology* 1991;**100**(3):795–8.
11. Chahal P, et al. Endoscopic resection of nonadenomatous ampullary neoplasms. *Journal of Clinical Gastroenterology* 2007;**41**(7):661–6.
12. Allgaier HP, et al. Ampullary hamartoma: a rare cause of biliary obstruction. *Digestion* 1999;**60**(5):497–500.
13. Fu H, et al. Periampullary carcinoma with situs inversus totalis: case report and review of the literature. *Zhonghua Wei Chang Wai Ke Za Zhi* 2007;**10**(2):134–7.
14. Bilimoria MM, et al. Pancreaticoduodenectomy in a patient with ampullary carcinoma and situs inversus. *Surgery* 2001;**130**(3):521–4.
15. Cannon ME, et al. EUS compared with CT, magnetic resonance imaging, and angiography and the influence of biliary stenting on staging accuracy of ampullary neoplasms. *Gastrointestinal Endoscopy* 1999;**50**(1):27–33.
16. Menzel J, et al. Tumors of the papilla of Vater – inadequate diagnostic impact of endoscopic forceps biopsies taken prior to and following sphincterotomy. *Annals of Oncology* 1999;**10**(10):1227–31.
17. Clary BM, et al. Local ampullary resection with careful intraoperative frozen section evaluation for presumed benign ampullary neoplasms. *Surgery* 2000;**127**(6):628–33.
18. Garcia-Fernandez FJ, et al. ERCP in complete situs inversus viscerum using a "mirror image" technique. *Endoscopy* 2010;**42**(Suppl. 2):E316–7.
19. Roggin KK, et al. Limitations of ampullectomy in the treatment of nonfamilial ampullary neoplasms. *Annals of Surgical Oncology* 2005;**12**(12):971–80.
20. Hornick JR, et al. A single-institution review of 157 patients presenting with benign and malignant tumors of the ampulla of Vater: management and outcomes. *Surgery* 2011;**150**(2):169–76.
21. Gray G, Browder W. Villous tumors of the ampulla of Vater: local resection versus pancreatoduodenectomy. *Southern Medical Journal* 1989;**82**(7):917–20.
22. Asbun HJ, Rossi RL, Munson JL. Local resection for ampullary tumors. Is there a place for it? *Archives of Surgery* 1993;**128**(5):515–20.
23. Posner S, et al. Safety and long-term efficacy of transduodenal excision for tumors of the ampulla of Vater. *Surgery* 2000;**128**(4):694–701.