Ampullectomy for an unexpected ampullary hamartoma in a heterotaxic patient

MEYER, Jérémy, et al.

Abstract

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.

Reference


DOI : 10.1016/j.ijscr.2013.03.003
PMID : 23608516

Available at:
http://archive-ouverte.unige.ch/unige:29975

Disclaimer: layout of this document may differ from the published version.
Ampullectomy for an unexpected ampullary hamartoma in a heterotaxic patient

Jeremy Meyer a, *, Andrea Rossetti a, Max Scheffler b, Philippe Morel a, Pietro Majno a

a Divisions of Visceral and Transplantation Surgery, University Hospitals of Geneva, Rue Gabrielle-Perret-Gentil 4, 1211 Genève 14, Switzerland
b Division of Radiology, University Hospitals of Geneva, Rue Gabrielle-Perret-Gentil 4, 1211 Genève 14, Switzerland

Article history:
Received 27 February 2013
Accepted 1 March 2013
Available online 16 March 2013

Keywords:
Heterotaxy
Situs ambiguus
Hamartoma
Intestinal malrotation
Polysplenia
Ampullectomy

ABSTRACT

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.

* Corresponding author. Tel.: +41 79 55 33 182.
E-mail addresses: jeremy.meyer@hcuge.ch (J. Meyer), andrea.rossetti@kssg.ch (A. Rossetti), max.scheffler@hcuge.ch (M. Scheffler), philippe.morel@hcuge.ch (P. Morel), pietro.majno@hcuge.ch (P. Majno).
http://dx.doi.org/10.1016/j.ijscr.2013.03.003

© 2013 Surgical Associates Ltd. Published by Elsevier Ltd. All rights reserved.
Laparotomy was performed, facing situs ambiguous 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 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 cholecodochotomy 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, and are mostly represented by adenomas. Non-adenomatous lesions, such as lipomas, lymphangiomas, hemangiomas, leiomyofibromas, and neurogenic tumors account for only 20% of ampullary tumors; hamartomas are the most rare, with 7 sporadic cases described to our knowledge.

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.

To our knowledge, we report the first case of a benign ampullary tumor in a patient with situs ambiguous.

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. In addition, endoscopic biopsies have a high rate of false-negative
results, particularly because of inaccuracies in sampling.\textsuperscript{4,6} Peroperative frozen section histopathological analysis thus remains mandatory in determining the extent of resection.\textsuperscript{1,7} There is no evidence suggesting that the preoperative evaluation is rendered complicated by the presence of heterotaxy.\textsuperscript{18}

Pancreatoduodenectomy is the operation of choice when an invasive ampullary carcinoma is suspected or when preoperative frozen-section histopathological analysis is not available.\textsuperscript{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,\textsuperscript{21} but is not a safe alternative in healthier patients due to high recurrence rates if the tumor is malignant.\textsuperscript{22} Ampullectomy is however an accepted treatment for presumed benign lesions,\textsuperscript{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.

Funding

None.

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.

Acknowledgements

The authors would like to acknowledge Olivier Huber, Jean-Louis Frossard and Laura Rubbia-Brandt for their help in conducting this research.

References


Open Access

This article is published Open Access at sciencedirect.com. It is distributed under the IJSCR Supplemental terms and conditions, which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.