Management of ampullary tumours in children: still a challenge

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ABSTRACT

This study describes the case of the youngest patient ever reported with ampullary adenoma. The incidence of ampullary adenomas in childhood is unknown. Endoscopic retrograde cholangiopancreatography (ERCP) and endoscopic ultrasound are used in adults to assess and treat these lesions, although there are no instruments designed specifically for use in young children. A six-year-old girl was admitted for abdominal pain, vomiting, pruritus and weight loss. Abdominal ultrasound showed biliary tree (8mm) and pancreatic duct dilatation (4mm). Magnetic resonance cholangiopancreatography and computed tomography confirmed these findings, and also showed displacement of the ampulla to the left upper quadrant. An upper endoscopy confirmed a large ampullary adenoma.

A laparotomy was performed and a 5cm villous tumour arising from the ampulla was excised. The postoperative course was uneventful. The histology demonstrated adenoma of the ampulla (intestinal type) without low-grade dysplasia. All clinical and radiological parameters are normal at 20 months follow-up.

We describe the case of the youngest patient ever reported with ampullary adenoma. Pancreaticoduodenectomy carries high morbidity and mortality rates, and therefore it should be avoided in absence of histologically proven malignancy. We believe that surgical ampullectomy is a safe and oncologically correct procedure until better endoscopic instruments for paediatric use will be designed.

KEYWORDS

Ampullary tumours – Ampullectomy – Paediatric tumour – Endoscopic ultrasound – Bowel obstruction

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Introduction

The incidence of ampullary adenoma in childhood is unknown, although children with familial adenomatous polyposis (FAP), Lynch syndrome and Peutz–Jeghers syndrome have a particular increased risk.^{1,2} Ampullary adenomas are more frequently identified in adults and they are considered to be premalignant lesions that need extirpation before completion of the adenoma–carcinoma sequence. Endoscopic retrograde cholangiopancreatography and endoscopic ultrasound are routinely used in adults to assess and treat these lesions; there are no instruments designed specifically for use in young children.^{3,4}

Case history

A six-year-old girl was admitted for abdominal pain, vomiting, pruritus and weight loss. She had history of a mild anaemia, which had started three months earlier. Physical examination revealed tympanic epigastric distension and elevated serum alkaline phosphatase (1547 u/L, normal range 80.0–90.0 u/L) was detected at blood tests. Abdominal ultrasound showed intra- and extrahepatic biliary tree dilatation (8mm) with gallstones. The pancreatic duct was mildly dilated (4mm). A magnetic resonance cholangiopancreatography was performed, which confirmed these findings and also showed displacement of the ampulla to the left upper quadrant (Fig 1). Computed tomography showed

Figure 1 Magnetic resonance imaging (axial view) shows gastric distension (blue arrow) with dilatation of the gallbladder and the biliary tree. Mild pancreatic duct dilatation can also be noticed (red arrow). The ampulla is displaced on the left side of the abdomen (yellow arrow).

Figure 2 Computed tomography (sagittal view). Intussusception of the duodenum with possible intraluminal mass can be noticed (blue arrow). Bile duct (yellow arrow) and pancreatic duct (red arrow) can be identified.

gastric distension and a jejunal obstruction at the ligament of Treitz (Fig 2). An upper endoscopy confirmed a large villous tumour mass extending from the duodenum to the first jejunal loop, suggestive of an ampullary tumour.

A laparotomy was performed and a jejunal intraluminal mass was confirmed at manual palpation. Bowel exploration revealed no other findings. Following a complete Kocher manoeuvre and cholecystectomy, the bile duct was cannulated through the cystic stump, confirming the presence of the probe within the mass. A duodenotomy was made along the lateral wall of the third portion, overlying the area of the ampullary tumour. A large villous tumour (5 × 4 × 3 cm) with no macroscopic signs of malignant transformation was detected. Full thickness resection of the ampulla was performed by sharp dissection. The pancreatic duct was identified. A papiloplasty was carried out with 5-0 PDS® sutures and the dilated duodenum was closed with an interrupted suture.

The postoperative course was uneventful and drains were removed after confirming normal drain amylase levels (less than three times serum level). Histology demonstrated adenoma of the ampulla (intestinal type) without low-grade dysplasia and negative margins. The young patient was discharged on postoperative day eight. She was subsequently diagnosed with Peutz–Jeghers syndrome. Retrospectively,

perioral freckling was also noticed. All clinical and radiological parameters were normal at the 20-month follow-up and a capsule endoscopy showed no evidence of further polyps.

Discussion

The incidence of ampullary adenoma in children is unknown, with most cases being described in association with familial adenomatous polyposis and other syndromes.^{1,2} We report the case of the youngest patient ever described with ampullary adenoma. In this particular situation, ampullary tumours can lead to two challenging problems: how to assess and how to treat these patients. Endoscopic retrograde cholangiopancreatography and endoscopic ultrasound can be used in adults for the evaluation of ampullary obstruction and to assess local tumour extension and suitability for resection, although there are no instruments designed specifically for use in very young children.⁵ There is no doubt that these are important limitations for decision making in ampullary malignancies, especially in case of Peutz–Jeghers syndrome, when duodenal polyps can markedly modify the normal anatomy.⁴ Endoscopic or surgical treatments are available, depending on the tumour

extension and local invasion.^{5,6} The American Society for Gastrointestinal Endoscopy guidelines suggest that endoscopic resections should only be used for lesions less than 4 cm and occupying less 33% of the circumference of the bowel⁴ but these guidelines are designed for adult patients. On the other hand, pancreaticoduodenectomy carries high morbidity and mortality and therefore should not be performed in the absence of histologically proven malignancy. Postoperative endoscopic surveillance is mandatory.⁴ Until better instruments are designed specifically for paediatric use, we believe that surgical ampullectomy can be carried out safely and with a good long-term outcome. This procedure should therefore be considered as the best treatment option for children with ampullary adenomas and no signs of invasive tumour.

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