Case Report

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Duodenal adenocarcinoma in a 10-year-old boy

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SUMMARY

Gastrointestinal malignancies are extremely rare in the paediatric population and duodenal cancers represent an even more unusual entity. It represents 0.3-1% of all gastrointestinal tumours. A case report of a 10-year-old boy with duodenal adenocarcinoma is reported and the difficulties of diagnosing and treating this rare tumour are discussed.

Key words: Duodenal adenocarcinoma, paediatric population, surgery

INTRODUCTION

Duodenal adenocarcinomas are rare in children. They represents 0.3-1% of gastrointestinal cancers and 25-35% of small bowel malignancies.[1,2] This rarity and the lack of specificity of their symptoms largely explain the delay in diagnosis and especially, their prognosis.

CASE REPORT

A 10-year-old boy presented to our unit in January 2012 with a 1-month history of abdominal pains, repeated vomiting and weight loss. An oesophagogastroduodenoscopy was performed revealing a duodenal ulcer treated medically. The evolution was marked by the persistence of clinical symptoms. A subsequent barium meal showed a bulbar stenosis with a large gastric stasis [Figure 1]. The child was initially operated with the diagnosis of stenosing ulcer of the bulb; he had bilateral vagotomy with pyloroplasty and making a check valve according to Dorr. During the operation, a pyloric lymphadenopathy was found, taken and sent for histological examination.

The histopathological findings reported a lymph node metastasis of adenocarcinoma probably of digestive origin.

Post-operatively, there was a regression of pain with less vomiting. The post-operative gastroscopy showed two polypoid and budding masses: The first in the bulb and the second level in the gastric antrum with respectively 20 and 6 mmdiameter. Biopsies were taken and showed aduodenal adenocarcinomawith same morphology as the node. Immunohistochemical evaluation showed strong staining of the neoplastic cells for Cytokeratin (CK7), Vimentin and CD10. These same cells were negative for CK20 and AFP. This profile suggested a kidney cancer. Abdominal MRI revealed a nodule in the liver [Figure 2], the kidneys were intact.

Patient underwent surgery, during which a 2 cm wide neoplasia was found in the first portion of the duodenum. The tumour extends to D2, the gastric antrum and the pancreas. The liver size, colour and consistency were normal without palpable nodule. The falciform ligament of the liver, which was cut during the first intervention, was retracted exactly in the seat of the nodule visualised by MRI. The essential components of the operation included duodeno-cephalo-pancreatectomy with gastric antrum resection, gastrojejunal end-to-side anastomosis, anastomosis between the jejunum to the common bile duct and pancreatic slice section, lymph node dissection and cholecystectomy.

The histopathological findings reported "well-differentiated adenocarcinoma extending through the duodenal serosa and infiltrating the pancreatic parenchyma with lymph node metastases (5N+/8N); pT4 N2". Post-operative period was uneventful with no

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Figure 1: Bulbar stenosis

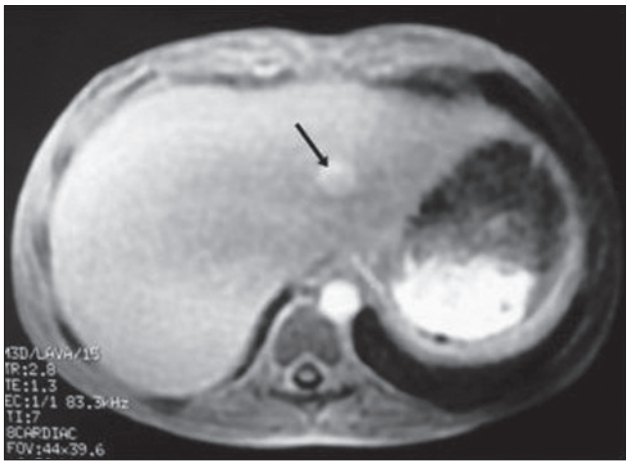


Figure 2: Hepatic nodule

complications. Patient resumed oral feeding on the 8th post-operative day and was discharged on the 15th post-operative day; four cycles of adjuvant chemotherapy were administered after oncological advice. A 1-year clinical-instrumental follow-up showed no locoregional recurrence of the disease.

DISCUSSION

Gastrointestinal malignancies are extremely rare in the paediatric population and duodenal cancers represent an even more unusual entity.[1] It represents 0.3-1% of all gastrointestinal tumours and 25-35% of malignant tumours of the small intestine.[2,3] Associations with familial adenomatous polyposis (FAP), Crohn's disease,[4] Peutz-Jeghers syndrome[5] and neurofibromatosis 1[6] have been reported. Duodenal adenocarcinoma often presents with vague and non-specific symptoms that lead to delayed diagnosis, patients can present with abdominal pain, bleeding, weight loss, obstruction or jaundice and most of the cases are accidentally discovered.[7-9] Oesophagogastroduodenoscopy with biopsy is the diagnostic gold standard. Nevertheless, computed abdominal tomography may be essential to show the neoplasia in the duodenal wall and determine the tumour staging and define the therapeutic strategy. Duodenal adenocarcinoma is known for its low sensitivity to chemotherapy. The only treatment for duodenal adenocarcinoma, which offers the possibility of a cure is excision by radical surgery.[10] Radical pancreaticoduodenectomy (Whipple's operation) is the classical curative operation and is still the treatment of choice for tumours in the first and second parts of the duodenum. Segmental resection is appropriate for selected patients, especially with lesions of the distal duodenum.[11] For unresectable tumours due to mesenteric vascular invasion or a major extension to surrounding organs or peritoneal carcinomatosis or distant metastases only simple bypass, type of gastro-jejunal-anastomosis or biliary-digestive bypass is advised, with a survival of 6 months.[12-14]

The 5-year survival rate for curatively resected adenocarcinomas of the duodenum is of the order of 50-60%.[15] Metastasis to lymph nodes, advanced tumour stage and positive resection margins are associated with decreased survival in patients with duodenal adenocarcinoma.[3]

The role of adjuvant chemotherapy is still unclear likely due to the rarity of this malignant neoplasm and the small patients numbers in most published experiences.[3,9,16,17] Some studies show important statistical benefits for medium- and long-term survival.[9,17] Information in the literature concerning this approach is scarce and needs further evaluation. Radiotherapy is not applicable as the tumour is radio-resistant and the small bowel has poor tolerance to radiation.[18] Hopefully, early diagnosis will correlate with improved long-term survival.

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