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Gangliocytic paraganglioma leading to duodeno-jejunal intussusception: A case report ☆,☆☆

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ABSTRACT

The intussusception of the small bowel is rarely encountered in adult patients and is frequently associated with a lead point that is often malignant. In a 69-year-old female patient with an episode of gastrointestinal (GI) bleeding, computed tomography (CT) showed a duodenal-jejunal intussusception caused by an intraluminal mass. Open polypectomy and reduction of intussusception were performed and the diagnosis of gangliocytic paraganglioma was made at pathological evaluation. It would be important to consider neoplasms like gangliocytic paraganglioma in the setting of adult small bowel intussusception.

Introduction

Intussusception is defined as the telescoping of a proximal segment of the gastrointestinal (GI) tract (intussusceptum) into the lumen of an adjoining segment of the GI tract (intussuscipiens). In children, this condition is mainly idiopathic, whereas in 90% of adult cases, it occurs due to the presence of intraluminal solid masses, which are commonly of malignant nature [1]. We present a case of upper GI bleeding caused by

Abbreviations: GI, Gastrointestinal; GP, Gangliocytic paraganglioma; WHO, World Health Organization; CT, Computed tomography; MRCP, Magnetic resonance cholangiopancreatography; ERCP, endoscopic retrograde cholangiopancreatography; CBD, common bile duct.

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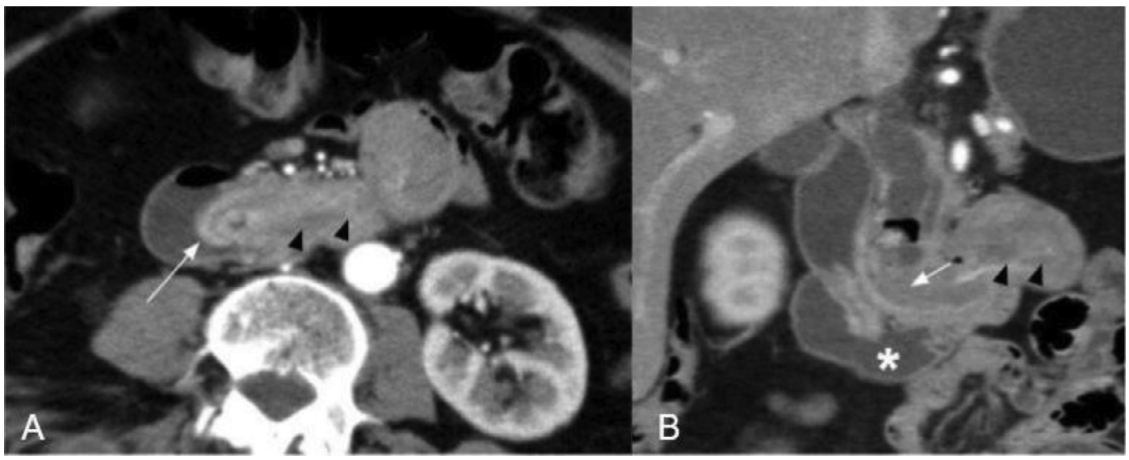

Fig. 1 – (A) Axial computed tomography (CT) scan (arterial phase). Duodeno-jejunal intussusception with a "target" sign is seen (white arrow) with contrast-enhanced tissue inside bowel lumen (black double arrowhead). (B) Coronal CT scan (arterial phase). Solid endoluminal tissue is more evident (black double arrowhead) in the duodenum distal tract, resulting in duodeno-jejunal intussusception (white asterisk) with "sausage" sign (white arrow).

an ulcerated duodenal mass associated with duodeno-jejunal intussusception. At the histological evaluation, the diagnosis of gangliocytic paraganglioma (GP) was made. The presenting symptoms of duodenal GP vary from GI bleeding, as in the case reported here, which occurs in nearly half of the patients, to less specific presentations such as abdominal pain or pancreatitis [2].

Although the World Health Organization (WHO) has classified the lesion as benign, lymph node metastasis and even distant metastasis have been reported.

Almost all patients with GP achieve good outcomes without relapses; however, a unique case of aggressive biological behavior resulting in a malignant clinical course after surgical resection and chemoradiotherapy was described [3].

Case presentation

A 69-year-old woman was referred to our intensive care unit presenting with melena for 3-4 days, epigastric discomfort, and nausea, with spontaneous and progressive escalation just before hospital admission. Upon physical examination, tenderness over the right hypochondrium in deep inspiration was present with an absence of masses, and peristalsis was normal. The patient had previously suffered from recurring deep vein thrombosis, which was treated with oral anticoagulants.

The admission blood test demonstrated severe anemia (hemoglobin level of 6.9 g/dL) with a serum urea level of 69 mg/dL (normal range: 17-48 mg/dL), while liver and kidney function were normal. Serum amylase levels were not elevated. The patient underwent upper GI endoscopy and colonoscopy, both of which were negative.

The patient also underwent capsule endoscopy, which demonstrated stenosis of the distal part of the duodenum secondary to a large ulcerating mass, resulting in almost complete occlusion of the lumen. She was further investigated with computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP), and endoscopic retrograde cholangiopancreatography (ERCP).

CT of the abdomen with contrast enhancement revealed a well-defined intraluminal mass (8.5 cm × 3.5 cm), causing duodenal intussusception of the D3-D4 tract into the proximal jejunum.

Also reported was dilatation of the intrahepatic biliary ducts and common bile ducts (CBDs) (axial diameter of 16 mm, and subsequent relief likely resulted from the involvement of the ampulla of Vater and distal CBD in the GI intussusception (Fig. 1). MRCP also confirmed the presence of CBD dilatation and mild dilation of the intrahepatic biliary tract.

The mass was further evaluated by ERCP, revealing a pedunculate ulcerating "polyp-like" lesion in the third part of the duodenum and intussusception into the proximal jejunum beyond the duodeno-jejunal junction; unfortunately, the biopsy performed on this mass did not result in a diagnosis.

At this point, surgery was performed with bi-subcostal laparotomy, cholecystectomy, drainage of the cystic duct, duodenotomy, removal of the mass in the duodenum, and excision of enlarged lymph nodes (Fig. 2).

During the surgery, intraoperative GI endoscopy and cholangiography were performed, particularly to locate the lesion and identify the ampulla with its anatomical correlation. Pathological evaluation of the specimen showed a polypoid mass 5.6 cm in diameter, with a tan-gray color cut surface and well-demarcated margins. Histologically, 3 different neoplastic populations were seen: epithelioid cells, ganglion-like cells and spindle cells. The former being the most representative with a moderate cytological atypia and a neuroendocrine architecture and morphology. The immunohistochemistry also

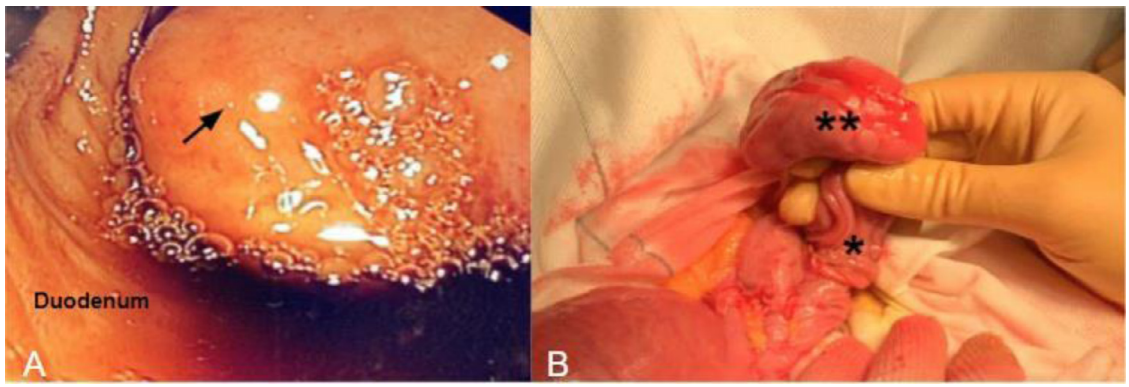

Fig. 2 – (A) Intraoperative esophagogastroduodenoscopy. Lumen of the third portion of the duodenum is occupied by a large mass (black arrow). (B) Intraoperative findings. Stick of polyp (black asterisk) and head of polyp after reduction of intussusception (double black asterisk).

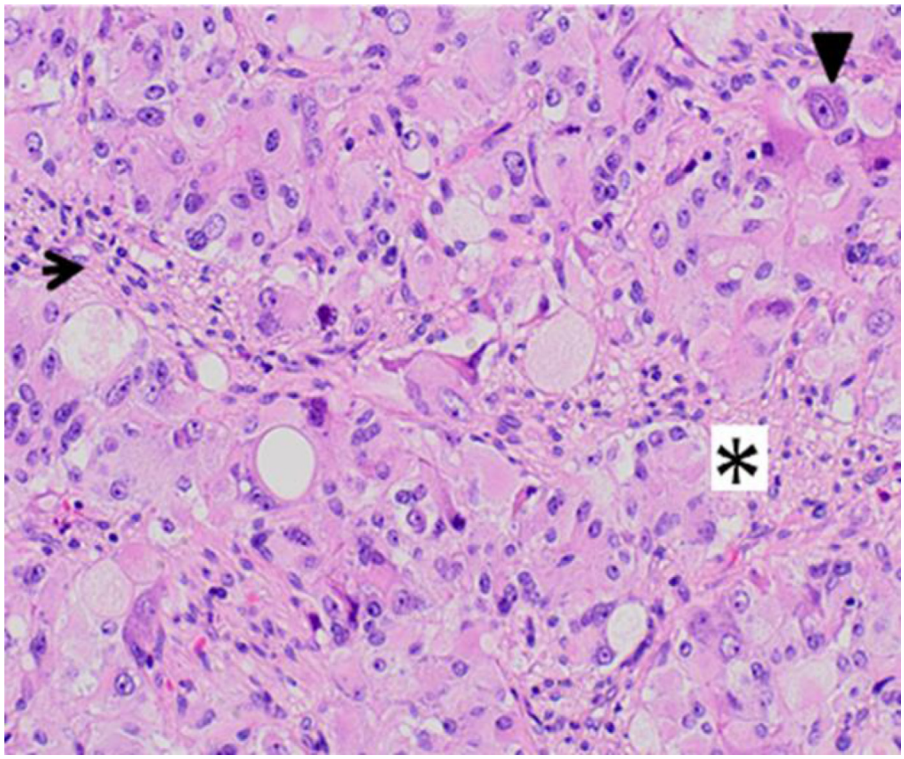

Fig. 3 – Ganglyocityc paraganglioma. High power field of the 3 neoplastic population: epithelioid cell (black astrerisk), ganglion-like cell (head arrow) and spindle cell (black arrow).

confirmed these 3 different types of cells. Spindle cells stained positive for S100 protein, ganglion cells for NF, chromogranin and synaptofisin. Epithelioid cells were also positive for the last 2 as well as CK 8/18 and somatostatin. The histological data combined with the immunohistochemical information made the diagnosis of Gangliocytic Paraganglioma possible (Fig. 3).

Also, one of the enlarged lymph nodes revealed to be metastatic, with the same histological pattern of the primary tumor. The patient was discharged 12 days after surgery, in good clinical condition.

Discussion

The term “intussusception” was conceived by John Hunter in 1789 and is defined as full-thickness invagination of a proximal portion of the bowel into the distal portion [2].

Such bowel obstruction is very unusual in adults, who represent 5% of all cases, with an occurrence of 2-3 cases per million adults per year. In children, almost all intussusceptions are idiopathic; however, in adults, the origin is detectable in 90% of cases [4]. The clinical manifestations of intussusception in adults are nonspecific: nausea, vomiting, GI bleeding, constipation, and intermittent abdominal pain.

The ileocolic junction is the most prevalent site for intussusception, followed by the ileoileal and colocolic regions. The duodenum is rarely involved in intussusception due to its fixed anatomic position; duodenal involvement has occurred in only 27 patients reported in the literature [5]. When it occurs at this site, the condition is almost always linked with a subordinate lead point, such as an intraluminal mass. Typically, a lead point consists of both benign and malignant neoplasms, lipomas, leiomyomas, Brunner's gland adenomas, adenomas, or stromal tumors [6].

Abdominal CT is considered the most reliable modality for preoperative diagnosis of intussusception and identification of lead point lesions when visible, showing the presence of typical bowel telescoping signs, including "target", "doughnut", or "sausage-shaped" signs [7].

Gangliocytic paragangliomas have been recognized to provoke intussusception in other parts of the intestine [8]; to the best of our knowledge, our patient represents the first reported case of duodeno-jejunal intussusception subordinate to GP.

GPs are a subclass of NETs, which occur nearly entirely in the second portion of the duodenum and were first illustrated as "duodenal ganglioneuroma" by Dahl et al. in 1957 [9].

However, remarkable cases of GPs emerging in the third or fourth portion of the duodenum have been described [10].

These are characterized by triphasic cellular differentiation giving rise to ganglion-like cells, epithelioid neuroendocrine cells, and spindle-shaped cells with Schwann cell differentiation; this heterogeneity can lead to misdiagnosis based on biopsy [2].

The use of CT in diagnosis is notably important, showing an intramural or, most frequently, a pedunculated mass with homogeneous enhancement and the eventual presence of metastasis [7].

Differential diagnosis covers lipoma, fibrolipoma, GIST, hamartoma, Brunner's gland adenoma, leyomioma, adenoma, villous and tubulovillous adenoma, duodenal membrane, duplication cyst, adenocarcinoma, duodenal ulcera, and pancreatic head cancer [2,11–21].

The proper treatment of duodenal GP is still disputed. Some authors report the need for radical resection with pancreaticoduodenectomy for better lymph node clearance and prevention of metastasis, while others suggest that surgical resection is adequate [2].

Endoscopic resection of duodenal gangliocytic paraganglioma appears to be reliable and effective in cases where the tumor can be removed in its entirety by endoscopic techniques [2,22,23].

Radiation therapists have advocated the use of adjuvant radiotherapy at the post-surgical bedside in cases of lymph node metastases [23].

There is no information on the use of chemotherapy in the management of this condition.

Conclusion

In the setting of GI bleeding and intermittent abdominal pain, it is uncommon to consider the diagnosis of small bowel intussusception caused by a neoplasm in an adult.

We report, here, a rare case of duodenal GP with regional lymph node metastasis occurring in an adult patient leading to a duodeno-jejunal intussusception that was successfully treated using a laparoscopic approach.

Ethical statement

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Human and animal rights

This article does not contain any studies with animals performed by any of the authors.

Informed consent

Informed consent was obtained from all individual participants included in the study.

Consent for publication

Consent for publication was obtained for every individual person's data included in the study.

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