Case Reports and Reviews

Case Report

A very rare case of jejuno-jejunal invagination caused by epithelioid sarcoma: A case report and literature review

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Keywords

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Abstract

Rationale: Intestinal invagination or intussusception is a rare event in adult and it is linked to a specific pathology abnormality in 70%-90% of cases often a benign lesion of jejunal wall being malignant tumors rare.

Objective: We report our experience about a 71 year-old man presented to emergency with three days history of generalized abdominal pain and swelling, vomiting and partial bowel obstruction, ability to gas.

Findings: A total body computed tomography (CT) scan detected one thickened ansa in the proximal jejunum, which suggested an invagination. Moreover evident gastrectasis with duodenal fluid distension was reported. The patient underwent laparoscopy that revealed a strictured area of the jejunum with a mass lesion. Owing to technical difficulties during surgery, the procedure was converted to open laparotomy, that showed jejuno-jejunal invagination caused by an intestinal tumor. Reduction was impossible and resection was performed. Histopathological examination revealed a malignant neoplasm with the features of mesenchymal tumor. Malignant cells with pleomorphic and large vesicular nuclei, prominent nucleoli and ample of eosinophilic cytoplasm were described. Histochemical markers supported the diagnosis of epithelioid sarcoma. One month after surgery, TC scan was performed finding lung and liver metastases also confirmed at PET scan, therefore the patient received adriamycin chemotherapy without efficacy confirming poor prognosis of this tumor.

Conclusion: Being intussusceptions in adult a infrequent condition underlyning neoplastic cause should always ruled out.

Introduction

Intestinal invagination or intussusception is the leading cause of intestinal obstruction in children, but in adults it accounts for only 5% of all intussusceptions, up to 5% of all cases of adult intestinal obstructions and 0.003%–0.02% of all adult hospital admissions. In contrast to childhood intussusception, which is idiopathic in 90% of cases, adult intussusceptions is linked to a specific cause which is pathologic abnormality in 70%–90% of cases, often a benign lesion of jejunal wall, being malignant neoplasms rare [1]. Sarcomas of the small bowel (SSB) are rare and represent around 8-15% of primary small bowel neoplasms [2]. Before the identification of gastrointestinal stromal tumors (GIST) as specific neoplasm, the majority of SSB were classified as leiomyoma, leiomyoblastoma or leiomysarcoma. Nowadays, GIST (85%) are the most common gastrointestinal tract sarcomas followed by leiomyosarcoma. Others non-GIST sarcomas include liposarcoma, fibrosarcoma, Kaposi's sarcoma and angiosarcoma [3]. Epithelioid sarcoma (ES) is a distinct clinicopathologic entity. It is a rare soft tissue neoplasm of unknown histogenesis and usually occurs in the distal extremities of young adults [4].

Case Presentation

A 71 year-old Caucasian man presented to emergency department with three days history of generalized abdominal pain and swelling, vomiting and partial bowel obstruction, ability to gas. She reported abdominal pain, constipation and moderate weight loss for several months, but had not carried out any medical examination. A total body computed tomography (CT) scan detected one thickened ansa in the proximal jejunum, which suggested an invagination. Moreover evident gastrectasis with duodenal fluid distension was reported.

The patient underwent laparoscopy that revealed a strictured area of the jejunum with a mass lesion. Owing to technical difficulties during surgery, the

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Desmin	Absent
S 100	Absent
CD 31	Absent
CD 34	Absent
CD 117	Absent
CD 68	Absent
Vimentin	Present
MNF II6	Present
AE I/AE3	Present
CAM 5.2	Present

Table 1: Immunohistochemical panel.

Discussion

In adult gastrointestinal invaginations are a rare events and the most common location on the small bowel is at distal end, ileo-ileal or ileo-colic sites. Acute clinical presentation is characterized by complete mechanical obstruction and sometimes with strangulating injury. Conversely some patients report relapsing abdominal pain and hematochezia for a long time. Often, the diagnosis is achieved during laparotomy performed for acute symptoms. From 80% to 90% of all intestinal invaginations develop at the site of structural lesions of the bowel wall and about 15% of those are malignant tumors. Small bowel tumors are infrequent, about 3% of all gastrointestinal neoplasms and sarcomas are extremely rare. GIST are the most common followed by leiomyosarcoma. ES is a rare soft tissue sarcoma, described for the first time by Enzinger in 1970 [5]. Two variants are known, the conventional distal type often found in the distal extremities of young adults and the more recent proximal-type/axial type, described by Guillon in 1977 [6]. It affects older patients, mostly in axial or deep location and has an aggressive clinical behavior with poor outcome. Visceral location is extremely rare and is described in the colon, bladder and perineal sites. Radical surgery is the treatment of choice although the tendency to local relapse and pulmonary metastasis that worsen the prognosis.

Conclusion

Here, we presented an extremely rare case of small bowel intussusception linked to ES. Being intussusceptions in adult a infrequent condition underlyning neoplastic cause should always ruled out.

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