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Recurrent intussusception of small bowel in a young patient due to metastases from cardiac undifferentiated pleomorphic sarcoma: A first ever case report

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ABSTRACT

INTRODUCTION: Undifferentiated metastatic pleomorphic sarcoma (Malignant Fibrous Histiocytoma) is a rare entity in the small intestine, especially when the primary tumor is of cardiac origin.

CASE REPORT: We report a case of metastatic intestinal undifferentiated pleomorphic sarcoma in a young patient with a history of primary cardiac tumor in the left atrium and recurrent small bowel intussusception. He was admitted for abdominal pain and constipation. A segmental resection of the small intestine was performed with side-to-side entero-enteric anastomosis.

DISCUSSION: Intussusception of the small bowel is rare in adults and it represents about 1–3% of intestinal obstructions. It mainly affects the fifth decade with a male/female ratio of 1:5. More than 60% of patients with intussusception have a tumor with 50% being malignant. This type of intussusception can be diagnosed on the CT abdominal scan. Radiological features include a typical “target” sign with overdistention of the proximal intestine and air-fluid levels, but the diagnosis of certainty is made by exploratory laparotomy.

CONCLUSION: The metastatic tumors that cause intussusception represent a rare clinical condition in adult patients, but much more common than primary ones. Metastasis to the small intestine are part of differential diagnosis in patient with a history of tumor who present with intussusception.

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1. Introduction

Metastatic undifferentiated pleomorphic sarcoma (Malignant Fibrous Histiocytoma) is a rare entity in the visceral organs. The most common metastasis in the small intestine are those originating from other organs in abdomen, rarely they arise from lung or breast [1,2]. We report a case of a metastatic primary cardiac undifferentiated pleomorphic sarcoma which presented with a recurrent small bowel intussusception in a young man. He was admitted for abdominal pain and constipation. A segmental resection of the small intestine was performed with entero-entero-anastomotic reconstruction. To our knowledge, there are no other similar cases in the literature and it is reported in line with the SCARE criteria [3,4].

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2. Presentation of case

A 22-year-old man was admitted to our emergency department with a 2-day history of abdominal distension and pain with constipation, nausea and multiple episodes of biliary vomiting. In his past medical history he had cardiac surgery for an undifferentiated pleomorphic sarcoma in the left atrium, about 3 years before with resection of atrial wall and mitral valve plasty. On cardiology follow-up he had a normal cardiac function with no assumption of anticoagulants and the only surgical residue was a hypodense formation (2 cm of diameter) on chest CT (Fig. 1a). Furthermore, he had previously undergone two partial resections of the small intestine with side-to-side entero-enteric anastomosis in our department for large pleomorphic sarcoma metastases causing recurrent intussusceptions (Fig. 1b and c). Family history was negative for other diseases. On admission physical examination revealed distended abdomen, constipation for about two days, pallor and tachycardia. The patient refused the insertion of the nasogastric tube. Laboratory findings showed a WBC count of $15 \times 10^3 \mu\text{L}$ and neutrophil count of 82.5%. Contrast-enhanced CT abdominal scan detected the typical target sign of intussusception of a tract of 10-cm length ileum with dilation of the proximal intes-

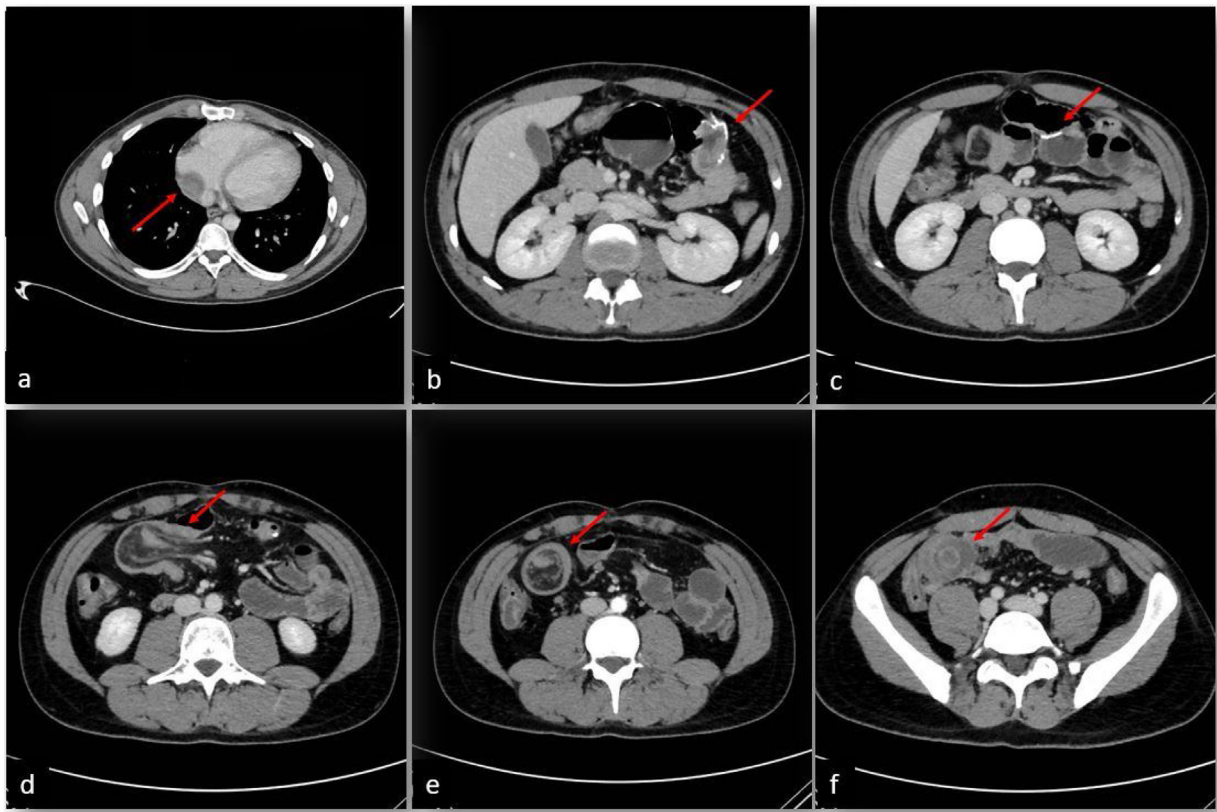


Fig. 1. preoperative CT scan. a) residual scar of cardiac surgery as a hypodense formation of 2 cm of diameter; b, c) radiological picture of small bowel obstruction with multiple air-fluid levels; the arrows indicate the previously constructed intestinal anastomoses that appeared well established; d–f) typical target sign of intussusception of a tract of 10-cm length ileum.

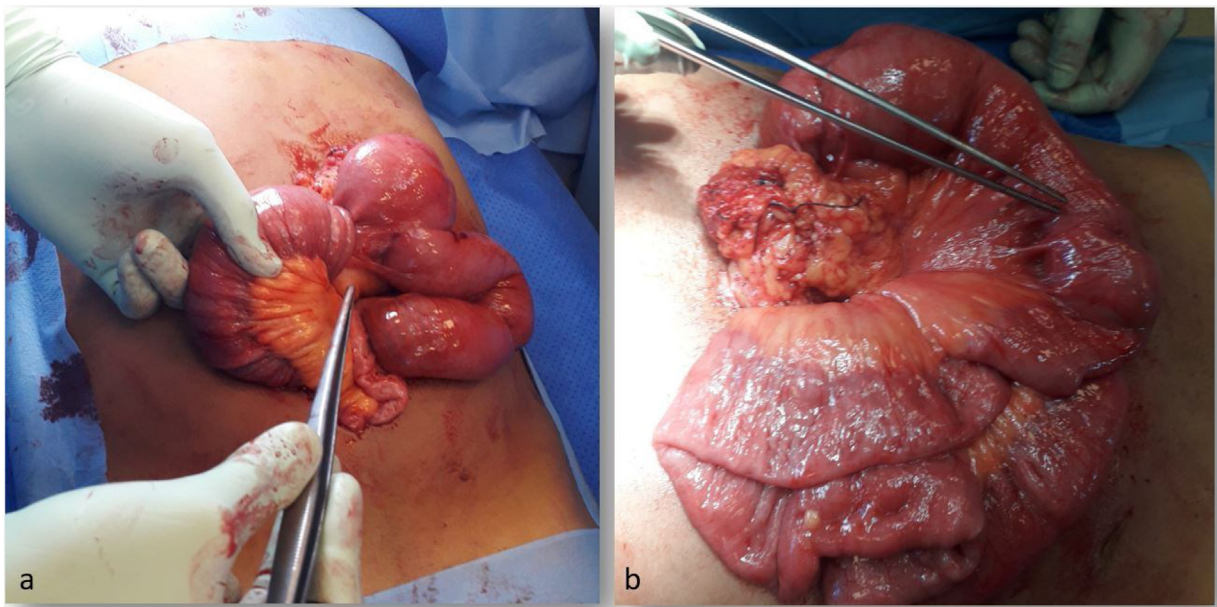


Fig. 2. intraoperative findings. a) intussusception of a 15-cm section of small intestine caused by a 4-cm intraluminal mass; b) the surgical forceps indicates the site of the intraluminal neoplasm.

tine and air-fluid levels (Fig. 1). Despite our experience in elective and emergency laparoscopy [5–10] we chose an open approach with a 15-cm mini-laparotomy because the patient had already undergone two median laparotomies and the CT abdominal scan did not show the presence of adhesion syndrome. The procedures

were performed by a young surgeon in urgent setting. Laparotomy by a small midline incision performed on the same day identified an intussusception of a 15-cm section of small intestine caused by a 4-cm intraluminal mass, about 40 cm away from Treitz ligament and distal to previous anastomosis (Fig. 2). We carried out a segmen-

tal small bowel resection and made an antiperistaltic side-to-side entero-enteric anastomosis with a linear mechanical stapler. At a systematic exploration there was no evidence of other intrabdominal neoplasia recurrences and the previously constructed intestinal anastomoses appeared well established. The patients were satisfied with the treatment received, the postoperative course was uneventful and the patient was discharged on the postoperative day 4. Histopathological examination of the tumor mass proved that it had a submucosal location. Immunohistochemistry analysis demonstrated a weak positive for SMA (Smooth Muscle Actin) and MDM2 (Mouse Double Minute 2 homolog) but there was no expression of desmin, myogenin, S100, CD34, CD31, Melan-A, PanCK, CD45, CD68, CD14, CD30 and ALK. Proliferation rate was 20% (Ki-67+). The pathological diagnosis was undifferentiated pleomorphic sarcoma.

3. Discussion

Intestinal intussusception refers to the invagination of an intestinal segment into the one immediately following due to abnormal peristalsis and which hinders the transit of intestinal contents and ultimately compromises the vascular flow [11]. Intussusception of the small bowel is rare in adults and it represents about 1–3% of intestinal obstructions. It mainly affects the fifth decade with a male/female ratio of 1:5 [12]. The metastatic tumors that cause intussusception represent another rare clinical conditions in adult patients. Generally, causes of intussusception in adult are benign lesions (lipomas, adenomas, Meckel's diverticula and fibromas) [13–15]. More than 60% of patients with intussusception have a tumor with 50% being malignant. The percentage of malignant tumors is lower in patients with enteric intussusception than in patients with intussusception involving the colon [12]. Usually the symptoms are nonspecific, such as abdominal pain and distension, fever, diarrhea or constipation and vomiting. Intussusception of the small bowel can be diagnosed on the CT scan. Radiological features are very helpful with a typical “target” sign with overdistention of the proximal intestine and air-fluid levels, but the diagnosis of certainty is made by surgery. Our case presented a recurrent small bowel intussusception, for which our patient has previous undergone two intestinal resections. In his past medical history he had a cardiac surgery for an undifferentiated pleomorphic sarcoma in the left atrium. The incidence of primary cardiac tumor is rarer than metastases. They have an incidence lower than 1%, 25% of these are malignant tumors [1,16]. Undifferentiated pleomorphic sarcoma is a soft tissue sarcoma of mesenchymal origin [15,17]. Most frequently it occurs in abdominal wall, retroperitoneum and in the extremities, but rarely it involves the visceral organs [18]. It is a very rare neoplasm of soft tissue, especially when we consider small bowel metastases originating from cardiac sarcoma.

4. Conclusion

Metastasis intussusception in the small intestine is a very rare condition and they are part of differential diagnosis in patient with a history of tumor who present with intussusception. In particular, establishing the real incidence of metastatic intussusception is quite challenging in many contexts so the published intussusception rates may be overestimated or underestimated. We hope that in-depth studies will be carried out to add to the existing literature in order to be able to better treat patients with these rare malignancies.

Declaration of Competing Interest

Di Buono Giuseppe and other co-authors have no conflict of interest.

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Di Buono Giuseppe and other co-authors have no study sponsor.

Ethical approval

Ethical Approval was not necessary for this study. We obtained written patient consent to publication.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Di Buono Giuseppe: study design, data collections, data analysis and writing.

Randisi Brenda: study design, data collections, data analysis and writing.

Romano Giorgio: data collections.

Ricupati Federica: data collection.

Buscemi Salvatore: study design, data collections, data analysis and writing.

Agrusa Antonino: study design, data collections, data analysis and writing.

Registration of research studies

Not applicable.

Guarantor

Di Buono Giuseppe.

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