

Rare case of entero-enteric intussusception caused by small bowel metastasis from a cardiac liposarcoma

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Abstract

Primary cardiac liposarcoma is exceedingly rare and its metastatic potential varies based on the actual tumor subclass. Intestinal intussusception is also an uncommon cause of abdominal pain and bowel obstruction in adults and it usually generates at a malignant lead point in this age group. We report a case of a primary cardiac dedifferentiated liposarcoma in a pregnant woman causing small bowel seeding leading to bowel intussusception.

Key words: Liposarcoma; Intussusception; Small bowel metastasis; Enteroenteric; Cardiac

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Core tip: Primary cardiac liposarcoma is exceedingly rare and its metastatic potential varies based on the actual tumor subclass. Intestinal intussusception is also an uncommon cause of abdominal pain and bowel obstruction in adults and it usually generates at a malignant lead point in this age group. We report a case of a primary cardiac dedifferentiated liposarcoma in a pregnant woman causing small bowel seeding leading to bowel intussusception.

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INTRODUCTION

Primary cardiac liposarcoma is exceedingly rare and its metastatic potential varies based on the actual tumor subclass. Intestinal intussusception is also an uncommon cause of abdominal pain and bowel obstruction in adults and it usually generates at a malignant lead point in this age group. We report a case of a primary cardiac dedifferentiated liposarcoma in a pregnant woman causing small bowel seeding leading to bowel intussusception.

CASE REPORT

A 29-year-old female with recent history of surgically resected intra-cardiac liposarcoma presents to the hospital at 27 wk of gestational age with complains of sudden onset of abdominal pain associated with intractable nausea and vomiting.

Three months prior to this presentation, the patient was admitted with severe shortness of breath and dyspnea on exertion. She was found to have a left intra-atrial mass on a transthoracic echocardiogram involving a large portion of the left atrium, anterior leaflet of the mitral valve and pulmonary venous system causing complete occlusion of the right inferior pulmonary vein. Cardiac magnetic resonance imaging confirmed the lesion (Figure 1). The patient underwent resection of the mass and porcine mitral valve replacement. In addition, she also had pulmonary venous endarterectomy. Due to the size of the mass with significant extension into the pulmonary venous vasculature, a complete en-block resection could not be performed. Postoperative course was complicated by a transient 3rd degree atrio-ventricular block. She was discharged on post-operative day 14. Outpatient tumor staging included non-contrast MRIs of the brain, abdomen and pelvis that were negative for any metastatic disease.

On this presentation to the hospital, an magnetic resonance imaging (MRI) of the abdomen and pelvis was performed for severe abdominal pain, which revealed an entero-enteric left upper quadrant intestinal intussusception with proximal small bowel dilation suggestive of a mechanical small bowel obstruction (Figures 2 and 3). After an unsuccessful trial of conservative management with nil per os and naso-gastric tube decompression, she developed acute cardiopulmonary decline associated with significant fetal cardiac distress. A multi-disciplinary meeting was held and she had an emergency caesarian section, exploratory laparotomy and placement of a transcutaneous cardiac pacemaker prior to the surgical interventions. A healthy, viable 950 g female infant was delivered. At exploratory laparotomy, a 10 cm long intussuscepted small bowel segment without evidence of ischemia, beginning at 50 cm from the ligament of Treitz was identified. After several and unsuccessful manual attempts at reducing this bowel segment, partial small bowel resection of the affected area with end-to-end anastomosis was performed.

The surgically resected specimen showed two polypoid

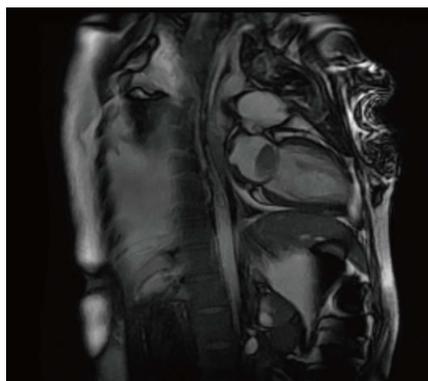


Figure 1 Cardiac magnetic resonance imaging showing a large mass in the left atrium.



Figure 2 Magnetic resonance imaging of abdomen showing entero-enteric intussusception in axial cut.

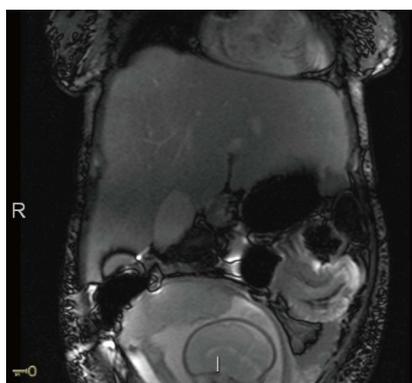


Figure 3 Magnetic resonance imaging of the abdomen showing an entero-enteric intussusception in the left upper quadrant.

lesions, which were 5 cm and 1 cm in their greatest dimension. Pathology was consistent with pleomorphic neoplasm with spindle cells, giant cells, vesicular nuclei with macro nuclei and occasional intra nuclear inclusions (Figures 4 and 5). Immunohistochemical staining was positive for Murine Double Minute (MDM) 2 and CDK 4. The histological features were identical to the original neoplasm seen in the heart 3 mo earlier (Figure 6). Post operatively the patient developed respiratory failure and was not able to be weaned off the ventilator. On post-operative day 8, the family decided to withdraw care.

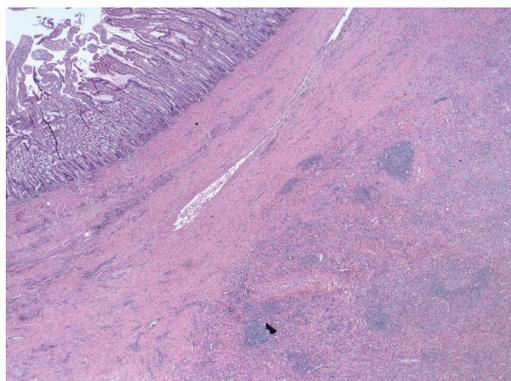


Figure 4 Normal small bowel mucosa (left hand corner) in contrast to infiltrative area of increased cellularity (right lower hand corner).

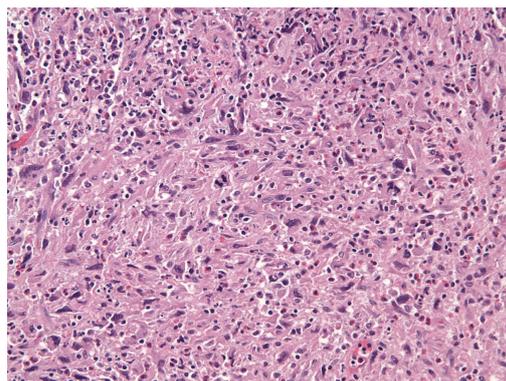


Figure 6 Poorly differentiated malignant neoplasm composed of variably spindled polygonal or histiocytoid cells and irregular vesicular nuclei consistent with a dedifferentiated liposarcoma.

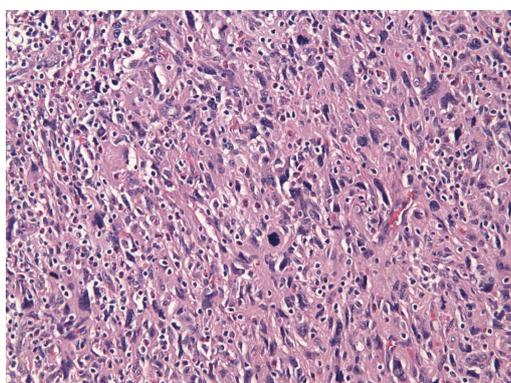


Figure 5 High power: Population of poorly differentiated malignant cells that are high grade (pleomorphic, hyperchromatic and contain increased mitotic activity) and are similar to the intracardiac dedifferentiated liposarcoma.

Unfortunately, the patient passed away the same day.

DISCUSSION

Primary cardiac neoplasms are exceedingly rare. Secondary (metastatic) cardiac lesions are 20-40 times more common than primary tumors^[1,2]. Malignant cardiac neoplasms constitute between 15%-25% of these primary lesions and sarcomas are found to be the most common malignancy^[3,4]. Liposarcoma accounts for about 13% of primary malignant tumors of the heart^[5]. There are five histological subtypes of these adipocyte precursor tumors including well differentiated, dedifferentiated, myxoid or round cell type, pleomorphic and mixed variant. Dedifferentiated liposarcoma commonly rises from the retroperitoneum and extremities and carries a lower incidence of distal seeding compared to the pleomorphic variant^[6-8]. Most common sites of metastasis include the lung, liver, bone, brain and soft tissues. Metastasis to the small bowel is rare with only few previously reported cases in the medical literature.

Surgical management is the preferred approach for cardiac liposarcoma, given its metastatic potential and the significant associated cardiorespiratory morbidity.

Intestinal intussusception is a rare cause of intestinal

obstruction in adults accounting for 1%-5% of the cases^[9]. It is defined as telescoping or prolapsing of a proximal intestinal segment into the adjacent more distal bowel segment secondary to peristaltic movements. Depending on the location of intestinal segment involved, intussusceptions are classified as enteroenteric, ileocolic, ileocecal or colocolonic. Majority of cases in adults are related to pathologic conditions that serve as a lead point for initiation of the prolapsing process. Intestinal diverticulum, luminal strictures and benign or malignant neoplasm are some of the more common causes^[9,10]. Enteroenteric intussusceptions due to malignant conditions accounts for up to 30% of all cases^[11]. Metastatic disease, adenocarcinoma and small bowel lymphomas are some of the leading metastatic causes^[12].

Clinical presentation varies depending of the degree of obstruction and ranges from chronic abdominal pain to intractable nausea, vomit and frank hematochezia in cases of complete obstruction associated with intestinal ischemia. Computer tomography scan of the abdomen and pelvis is the preferred imaging modality, with reported accuracy rates form 30%-100%^[12]. Abdominal Ultrasound and abdominal MRI are two other alternatives and can be used in pregnancy to minimize radiation exposure to the fetus.

Surgical resection of the intussuscepted area is the treatment of choice in the adult population^[9]. Although surgical resection was performed in our patient, with appropriate treatment of intestinal obstruction, her simultaneous pregnancy, heart block and respiratory condition increased her overall risks for mortality.

There have been few reported cases of liposarcoma causing metastasis to the small bowel. To the best of our knowledge, this is the first reported case of a primary cardiac dedifferentiated liposarcoma causing metastasis to the small bowel with associated intussusception.

COMMENTS

Background

Primary cardiac liposarcoma is exceedingly rare and its metastatic potential

varies based on the actual tumor subclass.

Research frontiers

Intestinal intussusception is also an uncommon cause of abdominal pain and bowel obstruction in adults and it usually generates at a malignant lead point in this age group.

Innovations and breakthroughs

The authors report a case of a primary cardiac dedifferentiated liposarcoma in a pregnant woman causing small bowel seeding leading to bowel intussusception.

Applications

On this presentation to the hospital, a magnetic resonance imaging of the abdomen and pelvis was performed for severe abdominal pain, which revealed an entero-enteric left upper quadrant intestinal intussusception with proximal small bowel dilation suggestive of a mechanical small bowel obstruction.

Peer-review

The case is interesting, well written and easy to read. References are new and adequate.

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