Enteroenteric Intussusception Secondary to Metastatic Cardiac Pleomorphic Sarcoma

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Introduction

Primary cardiac intimal sarcoma (PCIS) is a rare type of primary cardiac tumor that is characterized by poorly-differentiated spindle-shaped cells with similar appearing smooth muscle. Intimal sarcomas are undifferentiated mesenchymal tumors arising from different tissues, large blood vessels versus cardiac tissues. PCIS is rare and often caught late in the diagnosis resulting in an average survival rate of 12-15 months if amenable to surgical resection. Recently, there has been some suggestion that PCIS and intimal sarcomas are subtypes of the same neoplasm. Diagnosis tends to be late as patients often present with metastatic disease. Cardiac neoplasms visualized on imaging often appear benign or similar-appearing to a thrombus, adding to the diagnostic challenge. Currently, there is no modality to screen for PCIS. Our case report outlines an uncommon presentation of cardiac pleomorphic sarcoma as a metastatic small bowel intussusception, as well as the challenges faced with diagnosing and treating this rare and aggressive malignancy.

Case Presentation

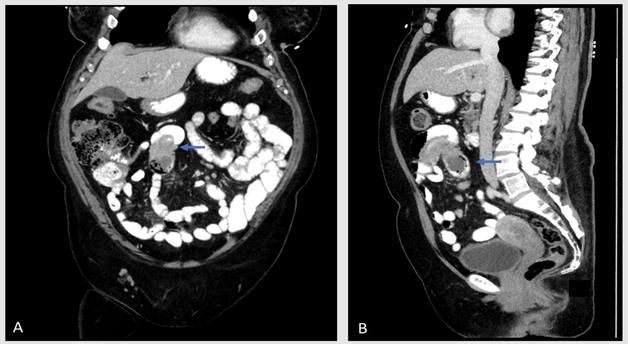
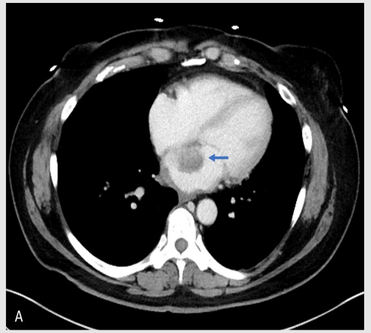


Figure 1. Axial (A) and sagittal (B) view of intussusception noted on CT abdomen and pelvis with IV and oral contrast.

67 year old female with past medical history of anemia, diabetes mellitus type II, hypertension, hyperthyroidism, and smoking history presented to her primary care physician with 1-week of intermittent abdominal pain. Outpatient CT abdomen and pelvis with IV and oral contrast revealing concerns for small bowel intussusception as a possible lead point for small bowel obstruction (Figure 1) and left atrial cardiac filling defect (Figure 2).

Patient underwent diagnostic laparoscopy where a mass was found 70 cm distal to Ligament of Treitz. A segmental small bowel resection was performed (Figure 3).

Treatment Course



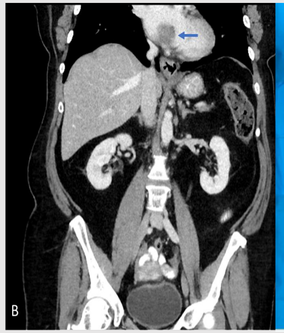


Figure 2. Axial (A) and coronal (B) views of incidentally noted 2.3 x 3 x 2.6 cm cardiac filling defect of the left atrium on CT abdomen and pelvis with IV and oral contrast.

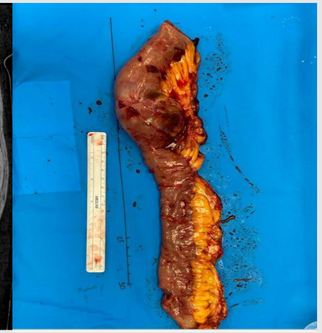


Figure 3. Surgically resected small bowel mass

Clinical Timeline and Treatment Course

- Acute onset of abdominal pain
  - CT abdomen/pelvis with concern for intussusception, cardiac mass 2.3 x 3.0 x 2.6 cm filling defect in the cardiac left atrium (Figure 1 and 2)
  - Echo: echodense structure located in the left atrium measuring 2.0 x 2.9 cm which myxoma cannot be excluded
- Diagnostic laparoscopy: 39 cm of small bowel resected (Figure 3)
- Cardiac mass work-up at University of Michigan sarcoma clinic
  - CT Chest: 2 masses in the attached to the left atrial septum measuring 2.6 and 1.5 cm noted
  - MRI: Large left atrial mass attached to the mid left atrial septum. Findings most consistent with a myxoma with probable intra-tumor necrosis/ thrombosis
  - Cardiac PET scan: left atrial mass consistent with myxoma
  - Cardiac catheterization without evidence of coronary artery disease
- Excision of left atrial mass and multiple satellite masses in left atrium by cardiothoracic surgery
  - Path: Grade 3, 4cm intimal sarcoma / cardiac pleomorphic sarcoma
- 6 cycles of doxorubicin and ifosfamide
  - ifosfamide held during cycles 5 and 6 due to acute kidney injury

Discussion

There are significant challenges faced with diagnosing and treating this rare and aggressive malignancy. When a cardiac pleomorphic sarcoma is suspected as a diagnosis, it requires an extensive workup to evaluate for possible metastasis as these are often late to be diagnosed. As with our patient, she presented with symptoms related to metastatic disease to her small bowel with initial pathology inconclusive of the origin until the cardiac mass was resected.

Initial imaging did demonstrate the cardiac lesion, however, without knowledge of malignancy, one could oversee this filling defect as a benign lesion. She underwent several cardiac imaging studies; however, only after surgical resection of the cardiac lesions was it able to be confirmed as a cardiac pleomorphic sarcoma. Additional diagnostic challenges to consider is the difficulty in obtaining tissue sampling of the intracardiac lesion, further contributing to the diagnostic and therapeutic dilemma.

Conclusion

PCIS/cardiac pleomorphic sarcoma is a difficult to diagnose and difficult to manage malignancy, with late diagnosis and an overall high mortality rate. Importantly, there are no current screening tools. From our literature review, there have been 4 other described cases of metastatic cardiac pleomorphic sarcoma to small bowel, however, given there is very little data regarding this pathology, we presented cardiac pleomorphic sarcoma as a metastatic small bowel intussusception.

References

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