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Case Report

Small intestine and ovarian metastasis in a patient with a history of cardiac fibrosacoma

Maryam Hassan Esfehani *, **Habibollah Mahmoodzadeh, Ramesh Omranipour**

Department of Surgery, Iranian National Cancer Institute, Tehran University of Medical Sciences, Tehran, Iran

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KEYWORDS

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Abstract Metastatic tumors involving the small bowel are much more common than primary neoplasms. The most common metastases to the small intestine are those arising from other intra-abdominal organs. Metastases from extra-abdominal tumors are rare but may be found in patients with adenocarcinoma of the breast and carcinoma of the lung. Cutaneous melanoma is the most common extra-abdominal source involving the small intestine, with involvement of the small intestine noted in more than half of the patients dying from malignant melanoma [1]. While intestinal metastasis from sarcoma has been described, this is an extremely rare occurrence especially from a rare malignant sarcoma of cardiac origin. The dismal prognosis of cardiac sarcomas results from extensive local invasion at presentation or distant metastasis. Metastasis to the small bowel may cause obstruction, bleeding, or intussusception in which the diagnosis may be delayed because of rarity of the condition and mild and vague abdominal symptoms at early presentation. In this report, a 35 year old woman a known case of cardiac fibrosarcoma was admitted to the emergency ward with abdominal pain and distention, bloody diarrhea, and recurrent nausea and vomiting. Jejuno-jejunal invagination was diagnosed at laparotomy along with tumoral involvement of the left ovary. Histopathological study showed that there was a fibrosarcoma compatible with the earlier diagnosis of primary cardiac tumor. We have described some aspects of diagnosis and treatment of this rare cause of intestinal intussusception.

Case report

A 35-year-old woman was admitted to the emergency department in a tertiary referral hospital with a 10 day history of intermittent upper abdominal pain, nausea, and episodes of bilious vomiting which had become more frequent in recent days. She had rectal bleeding 4 days earlier along with abdominal distention and increased pain intensity over time. She was suffering from obstipation within the last 24 h. In her past medical history, she had an open cardiac surgery for a cardiac sarcoma of atrial origin about 18 months before. There was no history of any other organic or systemic disease or any positive points in family history. In physical examination, she was ill and dehydrated with tachypnea and tachycardia. The abdomen was distended and tender. Results of imaging studies indicated a typical complete small bowel obstruction. Abdominal

* Corresponding author at: Cancer Institute, Keshavarz Blv, Tehran, Iran. Tel.: +98 912 387 62 79/21 6119 2542.
E-mail address: sanaze.esfehani@gmail.com (M.H. Esfehani).

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X-ray showed dilated loops of the small intestine illustrating an obstructive pattern. Abdominal sonography revealed a thickened small bowel wall and a pseudo-kidney appearance of small bowel compatible with intussusception. After resuscitation, the patient was transferred to the operating room for an emergency laparotomy. On exploratory laparotomy, a jujeno-jujenal intussusception and a palpable mass as lead point at about 100 cm distance from the Trietz ligament were found. The manual reduction of intussusceptum was impossible due to intense edema and a nearly gangrenous bowel. On further exploration of the abdomen, a solid tumoral lesion of about 15 cm diameter originating from the left ovary was found. There was not any other intra-abdominal involvement. Segmental small bowel resection with primary anastomosis was performed and then, the left ovary was resected. On the second postoperative day the patient complained of a left thigh pain during ambulation. On a detailed physical examination the pain was perceived to be from both a femoral bone lesion and lumbosacral nerve root irritation. MRI of the lumbosacral region revealed multiple metastatic involvement of the lumbosacral vertebrae. X-ray of her left femoral bone revealed a soft tissue mass with some cortical aberration of the left femoral bone. The post-operative course was uneventful otherwise. Gross and histopathologic examinations of the resected small bowel revealed a neoplastic mass with features of fibrosarcoma compatible with the original cardiac pathology.

Discussion

When a segment of intestine (i.e. Intussusceptum) invaginates into itself the process is called intussusception. Intussusception comprises about 2–3% of bowel obstructions in adults. Intussusception most often presents insidiously with mild abdominal symptoms in adults and rarely presents with vomiting, abdominal pain, and passage of blood through the rectum making an early diagnosis somehow difficult. Intussusception can be diagnosed on abdominal sonography by its virtually characteristic appearance of pseudo-kidney sign on longitudinal view or target sign in transverse view, and on the CT scan it appears as a ''sausage-shaped'' mass in the longitudinal axis, and as a ''target'' mass in the transverse planes [2]. Abdominal CT-scan was not attempted in our case because of the emergent situation which required a prompt surgical intervention.

In contrast to children, more than 90% of intussusceptions in adults have a demonstrable etiology with neoplasms being the leading cause and comprising 60% of the cases (60% malignant, 40% benign). The high rate of pathologic lead points and the high rate of malignancy make surgery mandatory in these cases [2].

Primary cardiac tumors are rare with an incidence of 0.0017–0.019%. Twenty-five percent of cardiac tumors are malignant. Cardiac tumors that have been studied are quite limited and medical literature provides little information on specific histopathological variants and their clinical outcome. Early Cardiac sarcomas are often asymptomatic and produce non-specific symptoms and mimic other pathologies. The poor prognosis of cardiac sarcomas is related to their tendency to infiltrate myocardium locally and or distant metastasis in the course of disease [3]. There are different presenting symptoms according to the location of the tumor and large vessel involvement: cardiac valvular or pump dysfunction, pericardial effusion or tamponade, arrhythmias, embolism to brain, lung, or retina, and symptoms such as dyspnea, chest pain, and syncope [4].

Wide surgical resection remains the cornerstone of treatment for sarcoma. Even with radical resections such as bench surgery and transplantation, although the local recurrence may be reduced to some extent, distant metastasis would be yet anticipated. Adjuvant therapies such as radiotherapy or chemotherapy have not been well evaluated in preventing local recurrences or distant metastasis [3,4].

The most common site of cardiac fibrosarcoma is within the right atrium, but fibrosarcoma can be found anywhere in the heart [5] as it was in the left atrium in our case. Fibrosarcomas are aggressive and can infiltrate myocardium or metastasize to distant sites including the lungs, liver, bone, and skin [6]. Interestingly our case presented with small bowel intussusception due to metastatic fibrosarcoma of jejunum as a lead point and the left ovarian involvement was revealed incidentally during laparotomy. Also, we found metastatic involvement of the lumbar spine and the left femoral bone only after a back pain and bone pain. There was no metastasis to brain, lung, or other intra-abdominal organs in our comprehensive evaluation for most probable metastatic sites of cardiac sarcoma.

In our extensive review of the literature, cardiac fibrosarcoma was a very rare condition and to the best of our knowledge, it is the first report of its metastasis to the small bowel and ovary.

Conclusion

The present patient had the characteristic imaging of small bowel metastasis. Small bowel metastasis from soft tissue sarcomas is rare especially from a cardiac origin where the primary malignant tumors are extremely rare. Fibrosarcomas of cardiac origin are highly aggressive. Widespread multi-organ metastasis should be anticipated at any time even after complete removal of this tumor. Adjuvant therapies for local control or avoiding distant metastasis are disappointing. Unfortunately, short survival of these patients is a rule rather than an exception.

Conflict of interest

The authors declare that they have no conflict of interest.

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