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Case Report

Fibrosarcoma of small bowel in a case of carcinoma ovary post cytoreductive surgery and hyperthermic intra peritoneal chemotherapy: a case report with review of literature

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ABSTRACT

Fibrosarcoma is a rare, malignant tumor of mesenchymal cell origin which derives from pathologically transformed spindle shaped fibroblasts with an excessively high division rate. There is no reported case of fibrosarcoma of the small intestine as a form of recurrence in carcinoma ovary post cytoreductive surgery and hyperthermic intra peritoneal chemotherapy (CRS and HIPEC). A 35-year-old female who is a known case of recurrent carcinoma ovary previously treated with cytoreductive surgery and HIPEC presented to emergency with features suggestive of intestinal obstruction. Upon exploration dense bowel adhesion and a segmental stricture with micro perforation without any leakage of intestinal content was discovered at 200 cm from DJ flexure. A diagnosis of fibrosarcoma was made on histopathological analysis. Recurrent carcinoma ovary patient treated with secondary CRS and HIPEC presenting with fibrosarcoma in small intestine is a rare entity and hardly any report in the literature. But appropriate diagnosis and management is the key for the successful outcome which we maintained in our case and managed successfully.

Keywords: Fibrosarcoma, Small bowel, Carcinoma ovary, CRS, HIPEC

INTRODUCTION

Fibrosarcoma is a rare, malignant tumor of mesenchymal cell origin. It derives from pathologically transformed spindle shaped fibroblasts with an excessively high division rate.[1] It is commonly reported to arise from the deep soft tissues of the lower extremities, particularly the thigh, but occasionally reported to arise also, from the chest wall, axilla, inguinal region, buttock, neck, the mediastinum, and brain. Abdominal fibrosarcoma is extremely rare, it had been reported to arise from small bowel mesentery, colon, falciform ligament, retroperitoneum, and the ovary.[2-4] They are commonly seen in the young or middle-aged adult, although it can occur even at the extremes of life. There is no reported case of fibrosarcoma of the small intestine as a form of recurrence in carcinoma ovary post CRS and HIPEC. We are reporting a rare case of fibrosarcoma of the small intestine presented as intestinal obstruction in a known case of recurrent carcinoma ovary treated with secondary CRS and HIPEC.

CASE REPORT

A 35-year-old female was diagnosed with complex right adnexal cystic mass for which right salpingoophorectomy was done in a different hospital in 2015. HPR showed high grade papillary carcinoma and mucinous cystadenoma. She subsequently underwent cytoreductive surgery followed by six cycles of paclitaxel and carboplatin. After disease free interval (DFI) for two years recurrence she was treated with six cycles of paclitaxel and carboplatin followed by secondary CRS and HIPEC in September 2018 in our center.

In October 2021 she presented to emergency with features of subacute intestinal obstruction. CECT abdomen showed dilated proximal bowel loops with acute cut off around jejuno-ileal junctional area.

On exploration dense bowel adhesion and a segmental stricture with micro perforation without any leakage of intestinal content was discovered at 200 cm from DJ flexure. There was no evidence of peritoneal recurrence. Resection of the diseased segment keeping 5 cm margin each side and double barrel ileostomy was done.

HPR showed spindle cell tumor with atypical mitosis (3-4/10 HPF) causing ischemic transmural infarct and bowel perforation. The spindle cells were immune-positive for SMA and CD34, and focally for desmin. They were immune-negative for cytokeratin, S100, MDM2, ALK and ERG proteins. Based on overall features, fibrosarcoma was diagnosed.

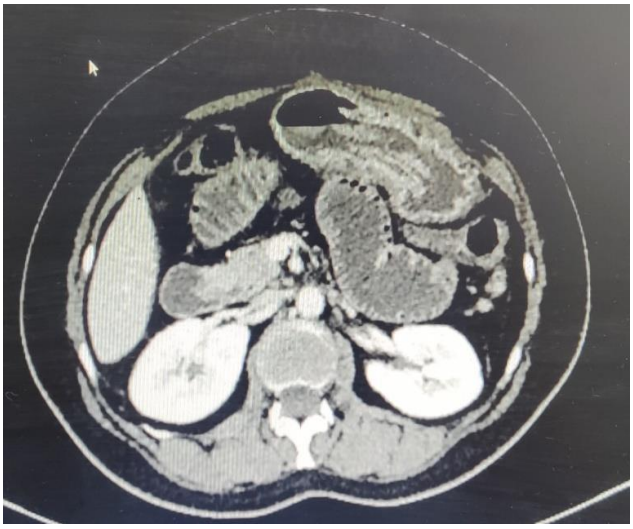

Figure 1: CT abdomen of acute intestinal obstruction

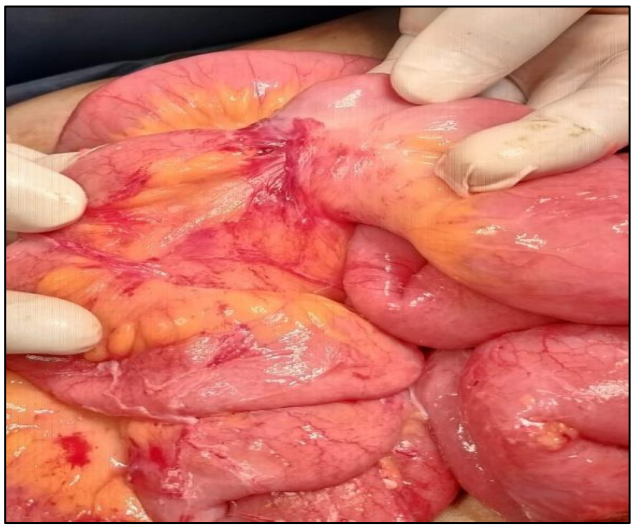

Figure 2: Intra-operative image of small bowel stricture.

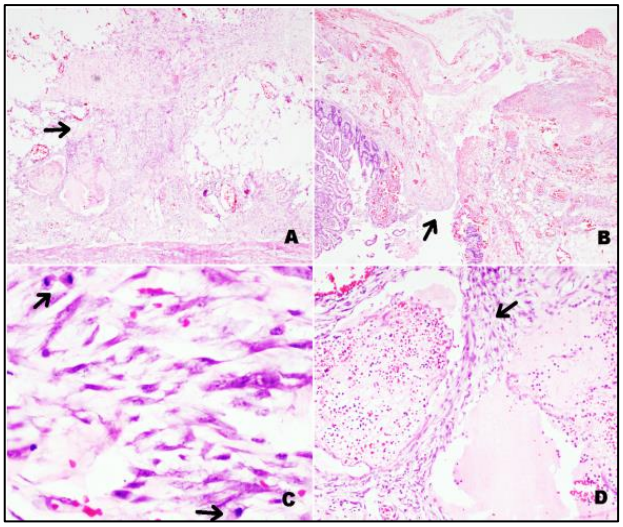

Figure 3 (A-D): Sections examined from thickened area of the serosa shows an irregular area of fibrous thickening (arrow) [A ×40]. Just adjacent to this area the intestine shows transmural ischemic infarct and perforation (arrow) [B ×40]. On high power examination the spindle cells were moderately pleomorphic with brisk mitotic figures (arrows) [C ×200]. The spindle cells (arrow) were also entrapping sub serosal blood vessels with dilated lymphatic spaces [D ×100].

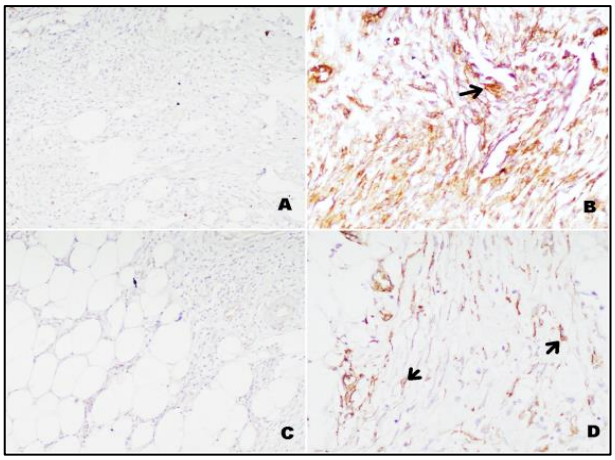

Figure 4 (A-D): The spindle cells were negative for cytokeratin [A ×100], positive for smooth muscle actin (arrow) [B ×100], negative for MDM2 stain [C ×100] and with focal; positivity for desmin stain (arrows) [D ×100].

DISCUSSION

Fibrosarcoma is a malignant tumor of mesenchymal cell origin which is commonly reported to arise from the deep soft tissues of the lower extremities, particularly the thigh.[1-2] The incidence of small bowel tumors is 22.7 cases per million, with sarcomas accounting for only 1.2% of the cases.[5] Sarcoma of the small bowel are extremely rare entities, to extent that there is limited literature available.

The etiology of sarcoma remains largely unknown, although there are certain genetic defects and environmental factors that have been linked to the development of sarcoma. Mostly sarcomas are sporadic and idiopathic. The diagnosis of small bowel sarcoma can be difficult, as the presentation can be delayed until complications are present.[6] The initial investigational modalities for ominous symptoms such as weight loss, constipation and rectal bleeding consists of upper gastrointestinal endoscopy and colonoscopy. Small bowel is not assessed by both modalities, and further imaging is therefore warranted.[7] Currently, magnetic resonance enterography (MRE), CT colonography (CTC) and wireless capsule endoscopy (WCE) have all been shown to be effective in arriving at a diagnosis.[8] Fibrosarcoma is a diagnosis of exclusion. It is further divided into subtypes by immunohistochemical and molecular techniques which can be very similar in their morphology, tumor genetics and clinical manifestation. These include low-grade fibromyxoid sarcoma (LGFMS, Evans's tumor), sclerosing epithelioid fibrosarcoma and myxofibrosarcoma. Moreover, other spindle-type tumors such as the monophasic fibrous synovial sarcoma, malignant peripheral nerve sheath tumor (MPNST), solitary fibrous tumor (SFT), aggressive fibromatosis as well as spindle-cell types of angiosarcoma, rhabdomyosarcoma, leiomyosarcoma and epithelioid sarcoma should be distinguished from fibrosarcoma.[9-11] In fibrosarcoma, vimentin, is often the only positively stained marker. Sometimes muscle specific antigen (MSA) and/or smooth muscle actin (SMA) can be detected as a sign of myofibroblastic differentiation. In those fibrosarcomas which arise secondarily from either solitary fibrous tumor (SFT) or dermatofibrosarcoma, CD34 can sometimes be detected.[9]

Ovarian cancer is more common in patients older than 50 years. Patients often present with nonspecific pelvic or abdominal symptoms. Standard treatment includes cytoreductive surgery (CRS) followed by chemotherapy. Recently hyperthemic intraperitoneal chemotherapy (HIPEC) has emerged as a promising modality for the treatment of carcinoma ovary as it has longer recurrence-free survival and overall survival compared to surgery plus adjuvant chemotherapy.[12] Extensive literature search did not reveal that either adjuvant chemotherapy (paclitaxel and carboplatin) or HIPEC is associated with fibrosarcoma of the small bowel. The possible role of HIPEC in occurrence of fibrosarcoma of the small intestine should be further investigated.

CONCLUSION

Fibrosarcoma of the small intestine in a patient of recurrent carcinoma ovary post CRS and HIPEC is extremely rare and warrants further investigation to ascertain the role of HIPEC in its pathogenesis. Surgical resection remains standard of care.

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