ISSN: 0975-8585

Research Journal of Pharmaceutical, Biological and Chemical Sciences

Sarcomatoid Carcinoma of Small Bowel – A Case Presentation.

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ABSTRACT

Sarcomotoid carcinoma (SCA) is a rare tumour of the GIT. Only 30 cases of SCAs have been reported in literature so far. Small bowel SCAs arise most frequently in jejunum and ileum .Pathologically it can be a monophasic type with predominantly mesenchymal cells or biphasic type with both mesenchymal and epitheliod cells. Pathological findings with immunohistochemistry play a decisive role in diagnosis of the disease. These tumours have been reported under various terminologies in the past, with SCA being the accepted term universally at present. Here, we report a case of 50 year old male diagnosed as SCA involving ileum, caecum appendix and ascending colon.

Keywords: Sarcomatoid carcinoma; sarcomatoid carcinoma of small bowel; Sarcomatoid carcinoma of small intestine; primary carcinoma of small bowel; SCA

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ISSN: 0975-8585

INTRODUCTION

Primary carcinoma of the small bowel is a rare disease with an incidence of 0.5 to 0.8% per 10,000 per year [1]. Among these, Sarcomatoid carcinomas (SCA) constitue only a small propotion. So far only 30 cases of SCA of small bowel have been reported in literature (TABLE I) [2-4]. SCA exhibit mixed histological features of epitheliod and mesenchymal tumours[4]. These tumours have been reported under various terminologies in the past, with SCA being the accepted term universally at present. Here, we report a case of 50 year old male diagnosed as SCA involving ileum, caecum appendix and ascending colon.

Table I - Cases of SCA reported in literature [2][3][4]

AUTHOR & YEAR	NUMBER OF CASES	REGION
Paik et al (1991)	1	Duodenum
Agarwal et al (1999)	1	Ileum
Tsukadaira et al (2002)	1	Jejunum
Reid-Nicholson et al (2004)	1	Jejunum
Moriwaki et al (2009)	1	Jejunum
Lee & Park (2012)	1	Ileum
Padma et al(2012)	1	Ileum
Kang et al (2013)	1	Jejunum
Dikman et al (1973)	1	Ileum
Radi et al (1985)	1	Ileum
Robey caffrey et al (1989)	6	Ileum & jejunum
Bak et al (1989)	2	Jejunum
Jones et al (1991)	3	Ileum
Lam et al (1996)	1	Jejunum
Fukuda et al (1996)	3	Jejunum & Duodenum
Arikan et al (2011)	1	Jejunum
Mittal et al (2012)	1	Jejunum
Nidia Alfonso Puentes (2014)	1	Jejunum
Ning han et al (2013)	1	Jejunum
Fulop emoke et al (2012)	1	Jejunum

CASE HISTORY

Patient, known chronic alcoholic since 20 years, presented with history of diffuse abdominal pain , high coloured urine and abdominal distension. There was no history of bleeding PR, altered bowel habits, vomiting , fever or significant loss of weight. On examination, patient was icteric and abdominal examination revealed ascitis with vague abdominal pain. Liver function test was deranged .While in hospital stay, patient developed intestinal obstruction, and was taken up for emergency laparotomy.

Imaging

USG OF ABDOMEN showed coarse echotexture of liver with nodular surface, dilated portal vein, mild splenomegay and moderate ascitis. CECT abdomen showed thickening at the level of ileo caecal region, with moderate ascitis, multiple peritoneal nodules, multiple enlarged nodes and heterogeneously appearing liver. CT scan of chest revealed mild left pleural effusion. Lung fields were normal and there were no significant mediastinal lymphnodes.

Ascitic Fluid Analysis

Ascetic fluid negative for AFB and malignant cells . NESTED RT PCR for Mycobacterium tuberculosis - negative.

Colonoscopy

Colonoscope could not be passed beyond the sigmoid colon due to technical difficulties.

ISSN: 0975-8585

Surgery

Patient was taken up for emergency laparotomy, since he developed intestinal obstruction. Intra operatively, multiple dense inter bowel adhesions were present , with peritoneum and omentum studded with nodules. Ileocaecal region showed circumferential wall thickening. Right hemicolectomy with ileo caecal anastomosis was performed. Resected specimen was sent for HPE. Patient developed anastomotic leak on 5th post operative day, and was taken up for re laparotmy. End ileostomy with closure of transverse colon was done.

Histopathology

External surface of the resected segment of bowel was congested with multiple tubercles meauriong 0.2 to 0.3 cms in diameter. Multiple strictures with ulceration and hemorrhage were noted within the mucosa. A fistulous tract was seen extending from the ileum into the cecum. Strictures with ulceration were also noted within the mucosa of appendix.

Microscopically the resected specimen showed features of an infiltrating malignant tumour predominantly in the ileo cecal region involving the serosa, mesocolon and muscular layers. The tumour exhibited patternless infiltration composed mainly of dispersed cells, trabeculae and few nests of cells. Cells were highly pleomorphic with spindling, containing round to oval vesicular nuclei with 1-2 prominent nucleoli and abundant bright eosinophilic cytoplasm. Binucleate and multinucleate cells were seen. Areas of hemorrhage were seen. No areas of necrosis were observed (FIGURE I). Resected ileal margin was involved by the tumour. Resected colonic margin was free of tumour. Peritoneal and omental nodules and all the examined lymph nodes showed eveidence of malignancy with similar features.

Pathologically the tumour was staged as pTx pN1 pM (N.A)

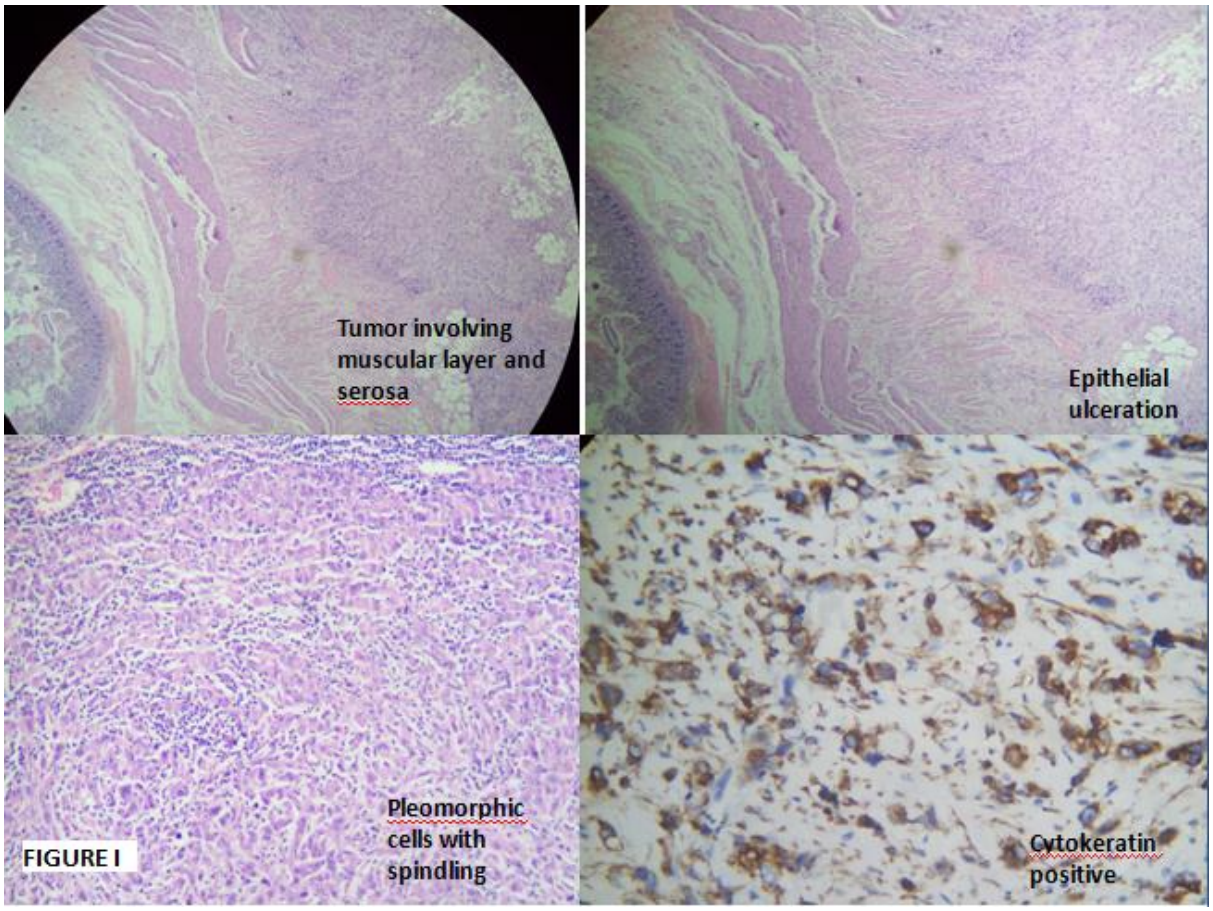

FIGURE I

ISSN: 0975-8585

Immunohistochemistry

The tumor was strongly positive for cytokeratin. It was negative for CD74, CD 45 and S100.

DISCUSSION

Sarcomotoid carcinoma is a rare tumour of the GIT [4]. Only 30 cases of SCAs have been reported in literature so far[2] .

SCA may present with symptoms such as intestinal obstruction, intestinal bleeding, abdominal pain, palapable mass , acute peritonitis due to perforation and anemia[2][5]. SCAs is found mainly in the middle ages (mean age – 57 yrs), and more common in the males[3]

CT scan of the abdomen is not conclusive of the disease. Usual findings are asymmetrical wall thickening , Irregular ulcerative or proliferative lesion [2]. Small bowel SCAs arise most frequently in jejunum and ileum [1][4].Pathologically it can be a monophasic type with predominantly mesenchymal cells or biphasic type with both mesenchymal and epitheliod cells [4]. SCA are usually strongly positive for cytokeratins and vimentin. Immunohistochemistry is principal in differentiating SCAs from other tumours such as leomyosarcoma (muscle specific actin and Des) [3] and GIST (c-kit and cd 34)[2].

Curative resection seems to be the mainstay of treatment with very less role for adjuvant chemotherapy or radiotherapy [3]. Our patient was taken up for emergency surgery since he developed intestinal obstruction. Compared to other histologic types of tumours of small bowel , sarcamatoid carcinomas have a far worse prognosis, due to highly invasive nature of the disease[3].

CONCLUSION

In conclusion SCA occurring in the small bowel are very rare tumor. It presents with non specific clinical symptoms and imaging findings. Pathological findings with immunohistochemistry play a decisive role in diagnosis of the disease. We report this case due to its rarity and peculiar clinico pathological features.

Ethical Statement

We warrant that the patient's rights and confidentiality have been well protected in all aspects and he consented to the study described in the Work. All relevant ethical safeguards have been met in relation to patient protection.

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