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Leiomyosarcoma Of The Intestine With Osseous Differentiation- A Rare Presentation

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Running Title: Leiomyosarcoma of the intestine with osseous differentiation
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ABSTRACT:
Heterotopic ossification (HO) within tissues involved by a pathologic process is a well-recognized phenomenon. It is most frequently observed in atherosclerotic plaques, in soft tissue around joints, and in the central nervous system. Less frequently, carcinomas and some benign neoplasms will undergo heterotopic ossification. We report on a case of high-grade Leiomyosarcoma with osseous differentiation. The identification of areas with osseous differentiation in soft tissue sarcomas can be of clinical importance because of a possible change in oncologic treatment strategies.

Key Words: Leiomyosarcoma, Intestine, Osseous Differentiation.

INTRODUCTION

Leiomyosarcoma (LMS) of small intestine is an extremely rare neoplasm (incidence ~ 1% of all gastrointestinal tract tumours) that usually presents with vague abdominal symptoms.[1] The highest incidence is seen in patients in their 6th decade with a slight predilection for males.[2] Owing to the rarity of the lesion and relative indolent clinical course, most of these tumours are diagnosed late in the course of the disease.

CASE SUMMARY

A 33-year-old Indian male who was admitted with pain abdomen and vomiting for 2 days- symptoms mimicking acute intestinal obstruction. No previous history of malena, other

gastrointestinal symptoms or weight loss was present. Preoperative ultrasound revealed a hyperechoic intramural growth of size 5 x 6 cms in the distal small bowel. At laparotomy, obstruction was identified at 10 cm proximal to ileo-cecal junction with a grey-white soft mass infiltrating the serosa of the small bowel. Resection of the involved ileal segment and adjoining omentum, followed by ileostomy was done. The regional lymph nodes and omentum were unremarkable. Imprint smear of the mass showed scattered population of plump to spindle shaped cells with oval hyperchromatic nucleus with distinct nucleoli and pulled out eosinophilic cytoplasm. (Figure 1) Histopathological examination revealed that the tumour comprised of malignant spindle cells with marked nuclear atypia, increased mitosis (>4 per 10 high power fields), focal necrosis and rich vascularity. (Figure 2) Widespread areas of osseous metaplasia were also present in between the tumour cells – another unusual finding that prompted this report. (Figure 3) Immunohistochemistry showed positivity for smooth muscle actin in the neoplastic cells (Figure 4) while CD34, CD117 and S-100 were negative. The final diagnosis of LMS of small bowel was offered. Post-operative adjuvant therapy of Cisplatinum 50 mg/m2 and 5 Flurourasil 500mg/m2 intravenous x 6 cycles were given. The patient is doing well after 12 months of follow up.

DISCUSSION

Ileum is the commonest site of intestinal LMS. Ahmad et al studied 28 patients of intestinal LMS and in 15 of them the tumour was in ileum.[3] They further observed that cases with presenting symptoms more than one year, tumour size <9 cm and without local or distant metastasis were significantly associated with better prognosis.[3] Patients of LMS present with vague complaints of dyspepsia, anorexia, malaise, dull abdominal pain and anaemia.[2]

Histologically, leiomyoma and gastrointestinal stromal tumours (GIST) should be considered in the differential diagnosis of LMS. The former tumour is much more common in the ileum and has benign smooth muscle fascicles without nuclear atypia, pleomorphism or tendency of invasion. GIST can mimic LMS morphologically; however,

immunohistochemically, 70-100% GIST are positive for CD117 and CD34 while LMS are not. Also, LMS is almost always immunoreactive to desmin and smooth muscle actin while only 5-10% GISTs show this feature. [4,5] Heterologous osteoid elements are extraordinarily rare in the intestinal leiomyosarcomas.[6]

Prognosis of LMS of intestine is poor. Spread by haematogenous (to liver, lungs and other parts of intestine) (69% cases), lymphatic routes (13% cases) and local peritoneal seedings (18% cases) significantly reduce the chances of response to treatment and likelihood of disease free survival.[7,8] Surgery remains the mainstay of treatment for the primary tumour and also for metastatic nodules. Radiotherapy has shown almost no role and chemotherapy have low response rate in this disease.[9,10]

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LEGENDS TO FIGURES

Figure 1: Imprint smear of the mass showed scattered population of plump to spindle shaped cells with oval hyperchromatic nucleus with distinct nucleoli and pulled out eosinophilic cytoplasm. H&E x 40X

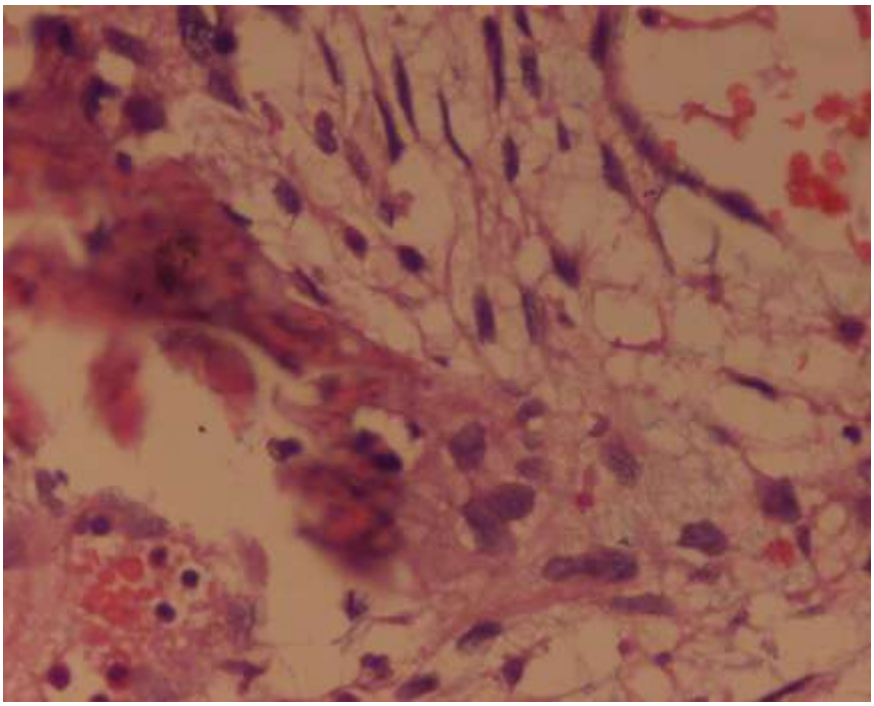

Figure 2: Histopathological examination revealed that the tumour comprised of malignant spindle cells with marked nuclear atypia, increased mitosis (>4 per 10 high power fields), focal necrosis and rich vascularity. H & E x40X.

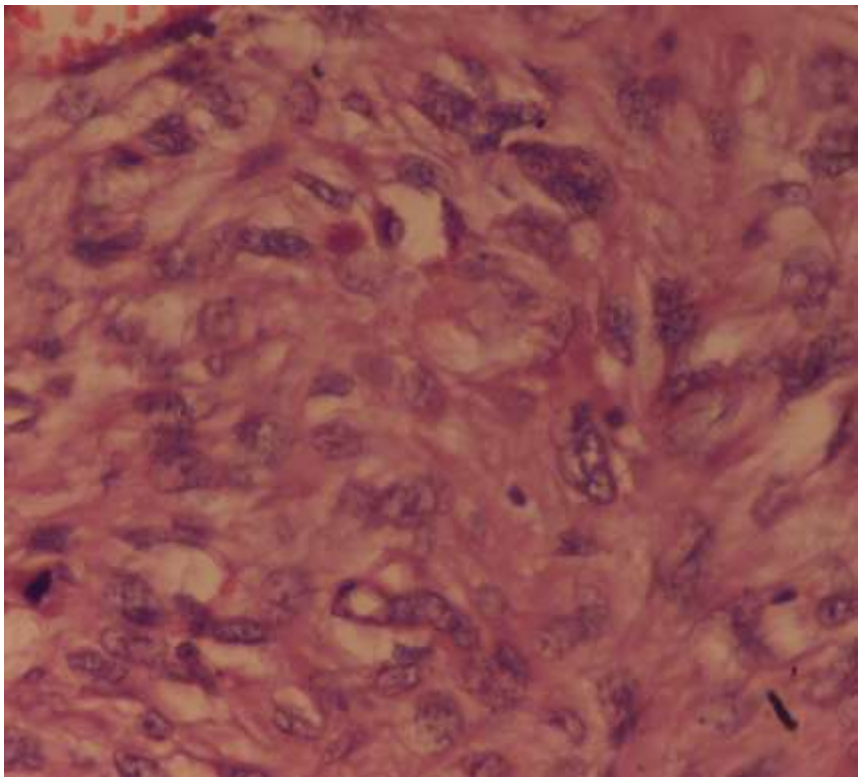

Figure 3: Widespread areas of osseous metaplasia seen in between the tumour cells. H & E x40X.

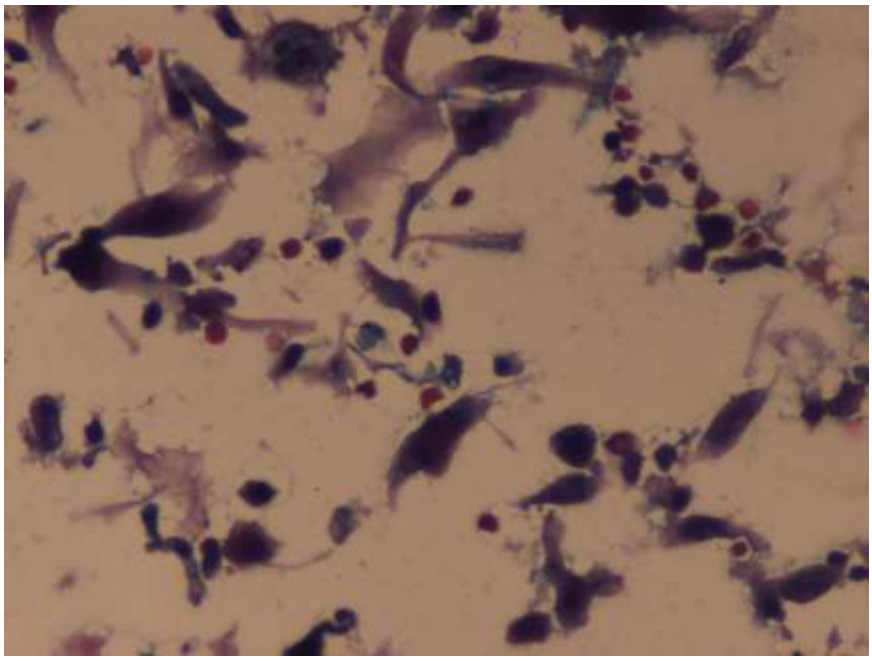

Figure 4: Immunohistochemistry showed diffuse cytoplasmic positivity for smooth muscle actin in the neoplastic cells. IHC SMA x 40X.