International Journal of Pharmaceutical and Bio-Medical Science

ISSN(print): 2767-827X, ISSN(online): 2767-830X

Volume 04 Issue 03 March 2024

Page No: 188-191

DOI: https://doi.org/10.47191/ijpbms/v4-i3-11, Impact Factor: 7.792

An Uncommon Tumor in Stomach in A Young Girl

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ABSTRACT

Introduction: Gastrointestinal stromal tumours (GIST) are rare tumours of the Gastrointestinal tract. They are the most common mesenchymal tumour of the gastrointestinal tract. All GISTs are malignant.Among all Gastrointestinal tract malignancies GIST Comprises 0.1- 3%. The most common location is stomach. They are usually found in elderly and they are not common in pediatric age group.
Case Report: A girl aged 16 years presented with the history of melena, hematemesis with dyspepsia. Physical examination revealed mild tenderness over right hypochondrium. On examination, multiple hypopigmented patches noted over both upper limbs. CECT abdomen showed a well-defined heterogeneously enhancing predominantly exophytic necrotic soft tissue SOL over the greater curvature of the proximal body of the stomach with intraluminal extension. Her complete blood count and routine biochemical tests were within normal limits. Endoscopic biopsy from the polyploid lesion along the greater curvature show fragmented bits of gastric mucosa involved by a tumour composed of sheets and fascicles of round to oval cells having eosinophilic to vacuolated cytoplasm and a few spindle cells. Histopathological examination revealed a mesenchymal neoplasm favouring GIST.. Then wide local excision of the stomach was done and sent for histopathological examination. The stomach mass was diagnosed as Epithelioid GIST which was confirmed by immunohistochemistry.
Conclusion: GISTs are most common mesenchymal tumours of GI tract. They are malignant in nature. The primary treatment of choice is surgery followed by chemotherapy

ARTICLE DETAILS

Published On:
15 March 2024

Available on:
https://ijpbms.com/

KEYWORDS: CECT**, e**pithelioid GIST, immunohistochemistry, spindle cell tumor, surgery.

INTRODUCTION

Gastro-intestinal stromal tumour (GIST) is the most common mesenchymal tumour of the gastrointestinal tract. Among all GI malignancies, 0.1-3% are GIST [1]. Prior to the recognition as distinct tumour, GISTs were most commonly classified as smooth muscle tumours or neural tumours and it was not benign [2]. According to WHO 5th edition all GISTs are malignant [3]. Occurring through the entire GI tract, most common site of GIST is stomach (60%) followed by small intestines (20%-30%) [4,5]

GISTs may rarely occur extra-gastrointestinal sites, where they most commonly occur in the omentum, mesentery and in retroperitoneum [4,5].

However, over the last 20 years with the improvements in the immunohistochemistry and the identification of gain of function mutations, GIST has been recognized as a separate entity [6,7]. 5-40% GISTs are diagnosed incidentally [8]. The most common findings of GIST are sub epithelial lesions seen on endoscopy or contrast-enhancing mass with a smooth margin on CT Scan[4]. There are three main histological patterns of GISTs- Spindle cell type (70%), Epithelioid cell type (20%), and mixed type (10%).[4,9]Spindle cell morphology remains the most common throughout the whole tract**.** Spindle cell type GISTs are described as having cells arranged in short fascicles or whirls, whereas Epithelioid cell type GISTs have cells arranged in a diffuse or nested pattern and mixed cell morphology incorporates both spindle cell and epithelioid cell histologic patterns.[8]

The pluripotent mesenchymal stem cells are known to be the cell of origin, those pluripotent stem cells programmed to differentiate into interstitial cells of Cajal, They are known as the "pacemaker cells" of the GI tract. They are responsible for initiating and co-ordinating GI motility [9]

Gastrointestinal stromal tumours can occur anywhere in the GI tract. Brunner's gland hamartomas of duodenum usually mimic the radiological and endoscopic features of GISTs.[10]

GISTs occur both in male and females in the 5th to 6th decade of life [11].

CASE REPORT

A 16 year old girl presented to the gastroenterology out-patient department with complaints of multiple episodes of melena over a period of 6 months with one episode of hematemesis about 1year ago. She also complained of dyspepsia for the same duration. There was no history of fever, jaundice, haematochezia. She did not complain of asthenia, anorexia and weight loss. No family history of malignancy was present. Her menarche was at 13 years of age.

On examination, there was multiple hypopigmented patches over both upper limbs. She had mild tenderness over right hypochondrium.

On Upper GI endoscopy (UGIE), a sub epithelial lesion of size 4x4 cm with ulceration and central depression seen in the greater curvature of the stomach in the proximal body just distal to gastroesophageal junction. Endoscopic Ultrasonography (EUS) showed a hypoechoic mass in fundus which was confirmed by CECT.

CECT abdomen showed a heterogeneous large polypoidal SOL arising from the fundus and greater curvature of the stomach. A second CECT abdomen was done after 3 months. There was a well-defined heterogeneously enhancing predominantly exophytic necrotic soft tissue SOL of size 5.4x5.5x7.2 cm (AP x TR x CC) seen over the greater curvature of the proximal body of the stomach with intraluminal extension. Mass effect was seen over the body of the pancreas with preserved fat plane.

Endoscopic biopsy from the stomach was done and sent for histopathological examination. The sections show fragmented bits of gastric mucosa involved by a tumour composed of sheets and fascicles of epithelioid cells having bland nuclei and eosinophilic to vacuolated cytoplasm. Occasional mitotic figures noted. Features were in keeping with the mesenchymal neoplasm favouring GIST.

Immunohistochemistry was performed and the tumour cells strongly and diffusely positive for CD34, CD 117, DOG1, while they are negative for S100, SMA, Desmin and CK (AE1/ AE3).

Her CBC and routine biochemical tests were within normal limits except the haemoglobin level which was 9.0 g/dl. So she received 2units of PRBC per-operatively.

Wide local excision done under GA. Intra-operatively one 8x5 cm. mass arising from posterior wall of stomach along the greater curvature. The defect was closed with primary repair. The margins of stomach wall was marked with sutures and sent for histopathological examination..

The tumor measured 7x7x4 cm. and it situated at the greater curvature of the stomach. The growth was well-encapsulated. The cut surface was lobulated, tan colored and fleshy and solid. (Fig. 1)

The tumor was unifocal. Microscopically, round cells with eosinophilic cytoplasm arranged in sheets (Fig. 2). Occasional spindle cells were also seen. Focal areas of necrosis present. The mitotic rate was more than 5 per $5mm^2$. The surgical margins were free from tumor invasion. The histological features were consistent with epithelioid GIST. The Immunohistochemistry for CD34 (Fig. 3) and CD117 (Fig.4), DOG1 were positive and S100 was negative

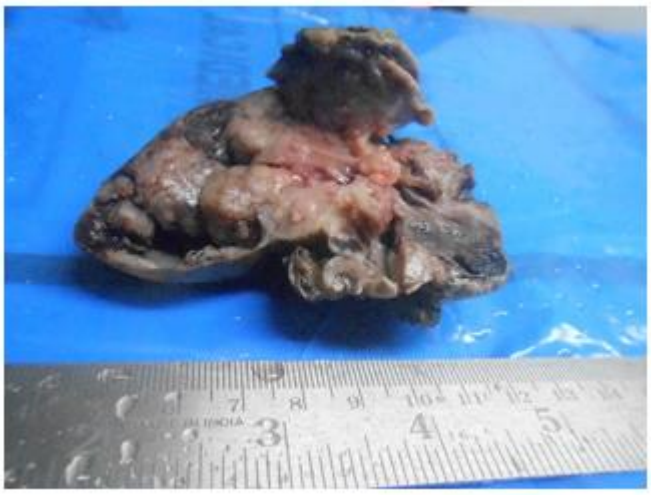

Fig 1 -Cut surface shows greyish white solid tumor

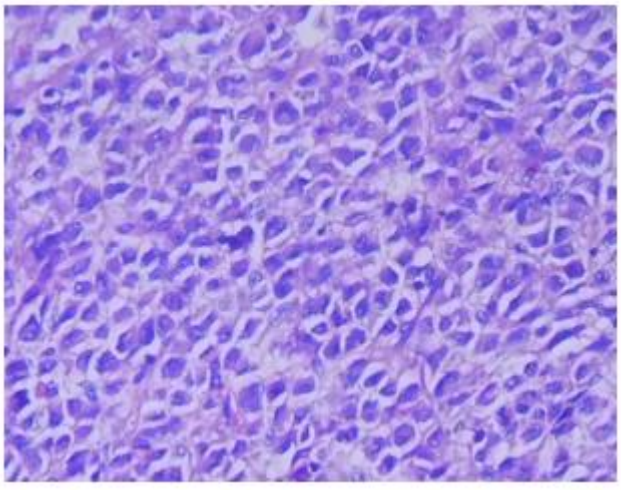

Fig. 2 Epithelioid cells in sheet (H&E 400X)

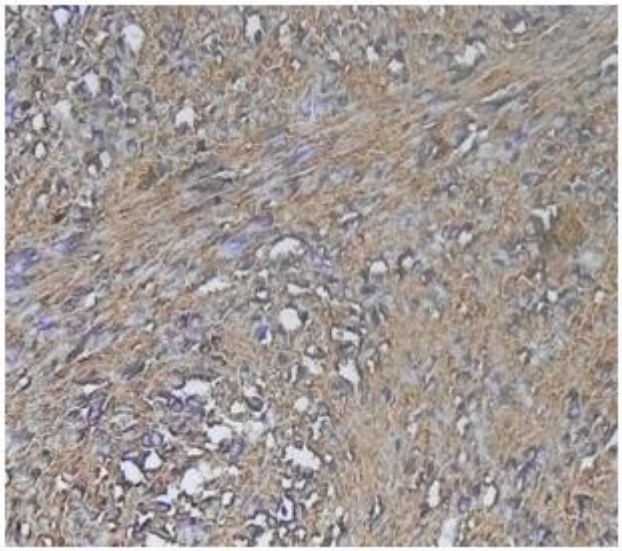

Fig 3- CD 34 positive in tumor cells (400X)

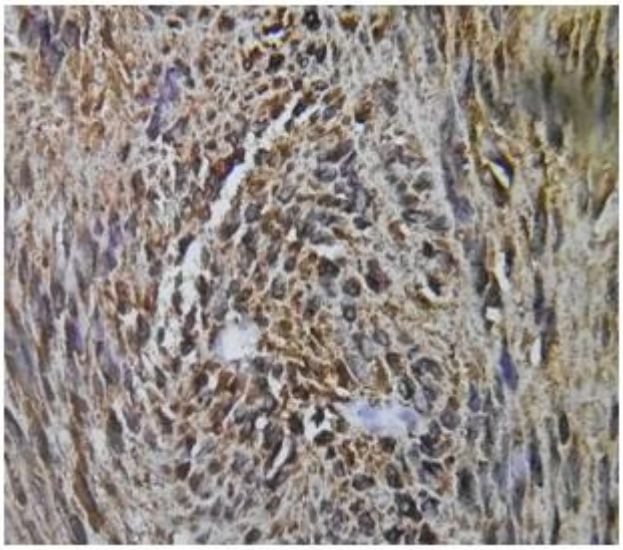

Fig 4- CD 117 positive in tumor cells (400X)

DISCUSSION

GIST is a rare tumor in the gastrointestinal (GI) tract and it is more rare in pediatric age group [12]. Our patient was also in pediatric age group.Among imaging modalities CT scan play an important role. CT scan usually done before surgery which shows a solid, contrast-enhancing mass with smooth margins. It is more helpful than USG [13]. MRI is not superior for diagnostic purpose when compared to CT. USG usually used for endoscopic biopsy before final surgery [13] . In the present case the tumor was diagnosed by CECT as heterogeneously enhancing soft tissue SOL . USG guided Endocopic Biopsy was done.

Pediatric GIST represents show female predominance and they are SDH deficient. They usually show epithelioid histology [3]. This pediatric patient was also female and histopathology revealed epithelioid histology .

The presenting symptoms depends on the anatomic location of gastrointestinal stromal tumors . In most cases there are vague abdominal pain. Some present with symptoms of gastrointestinal bleeding [14]. Here the presenting symptom was melena and one episode of hematemesis.

Endoscopy is the preferred method of evaluation for patients presenting with gastrointestinal bleeding. On the endoscopic exam, GISTs most commonly appear similar to other sub-epithelial lesions (SEL) with a smooth bulge covered with normal-appearing mucosa. Endoscopic ultrasound (EUS) has proven to be an important tool to help differentiate GISTs from other SELs. On EUS a GIST will appear as a hypoechoic solid mass **[15]**. But hypoechoic mass also seen in cases of Leiomyoma, schwannoma, neuroendocrine tumor, etc [15]. Here the EUS presented a hypoechoic mass arising from fundus of stomach[15]. In this case also CECT showed a sub epithelial lesion in the stomach on the greater curvature.

Surgical resection of the tumor with wide locl excision is the main treatment. Then the patients are given Imatinib to prevent recurrence of the tumor[15]. In this case also wide local excision of the tumor was done. She also received Imatinib treatment for last one year and she is still continuing the treatment. Now she is now doing well.

CONCLUSION

GIST isa rare malignant tumour. It is the most common mesenchymal tumour of the GI tract. It is very rare in pediatric population. Surgery with wide local excision the treatment of choice followed by Imatinib therapy.

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