

Synchronous Development of Gastrointestinal Stromal Tumor and Arteriovenous Malformation in the Jejunum

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Abstract

This case report is about synchronous development of gastrointestinal stromal tumors (GISTs) with AV malformations (AVMs). A 23-year-old pale woman presented with melana and anemia. Abdominal computed tomography Non Enhanced Contrast (CT-NEC) revealed a gut wall thickness in left hypochondrium and multiple focal calcifications. Contrast enhanced computed tomography (CECT) revealed non-homogenous enhancement with circumferential gut wall thickening and narrowing lumen plus AV malformations in the gut wall giving impression of mass in the jejunum with AV malformation. An exploratory laprotomy was performed. The resected jejunum showed a protruding solid mass. The mass was confirmed to be a spindle cell type GIST and was intermingled with the AVM. To our knowledge, this is the second case of an AVM associated with a GIST. First case was reported in 2008 by Shim SH et al ¹.

key words: Gastrointestinal stromal tumor; Arteriovenous malformation; Jejunum.

Introduction

Vascular malformations are abnormalities in the architecture of vessels. They include angiodysplasia, deulafof's lesion and Arteriovenous malformation. The most common being angiodysplasia ¹. AVMs are relatively rare in the gastrointestinal tract. The usual presentation is recurrent or obscure upper or lower GI bleeding due to vascular lesion. GIST affects GIT and consists of CD34 positive and c-kit signaling-driven cells ². GISTs originate from the interstitial cells of Cajal ³. Diagnosis is made radiographically (CT scan, MRI or angiogram).

By imaging studies, GISTs on CT are well-defined tumors showing homogeneous contrast enhancement ⁴ and usually as hypoechoic, well-demarcated round masses by ultrasonography (US) ⁵ but, these findings may vary according to malignant potential of tumors ⁶. The synchronous occurrences of vascular malformation and neoplasia in the GI tract have been rarely reported in the English literature and till date there is only one case of AVM associated with GIST have been reported ¹. Since this is an unusual and interesting case, we are reporting it with review of the literature.

Case Report

A 23-years-old pale woman was admitted with complaint of painless, intermittent melana and anemia from last 12 years. She had previous history of oral iron supplements and multiple blood transfusions but she was intolerant to oral iron therapy. No history of dyspepsia, hematemesis, menorrhagia or previous surgical intervention. On examination, there was a mass palpable in left hypochondrium and lumbar region.

Laboratory investigation revealed iron deficiency anemia: hemoglobin 9.7 g/dl; hematocrit 30.2%, red blood cells $3.96 \times 10^6 / \mu\text{l}$, peripheral film showed hypochromic microcytic red cells with few target cells. Hepatitis B, C was negative.

Upper GI endoscopy and sigmoidoscopy was normal. Abdominal computed tomography with I/V contrast revealed small bowel hemangioma. Abdominal computed tomography non contrast enhanced (CT-NCE) revealed a gut wall thickness in left hypochondrium and multiple focal calcifications. Contrast enhanced computed tomography (CECT) revealed non-homogenous enhancement with circumferential gut wall thickening and narrowing lumen plus AV malformations in the gut wall giving impression of mass jejunum with AV malformation. An exploratory laparotomy was performed. The

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jejunum showed a protruding, solid mass adherent to the wall. The jejunum along with mass lesion was excised and sent for histopathology.

The resected jejunum was received in pathology department of PIMS hospital in a jar containing 10% formalin. The specimen was left in formalin for 24 hrs for fixation. The resected segment of the jejunum measured 14x2.2x2.2cm with a solid mass, protruding out of the intestinal wall measuring 9x8x5 cm. Cut section showed solid, nodular whitish areas having whorled appearance. Focal areas show hemorrhagic areas (fig 1).



Figure1: Photomicrograph of Cut section of tumor showing whorled appearance.

Microscopically, the tumor revealed a well circumscribed spindle cell neoplasm in the submucosa, extending beyond the muscularis and serosa. The tumor was arranged in interlacing fascicles of spindle cells separated by fibrous septa.

The cells were uniform having pale eosinophilic, indistinct cytoplasm and oval to short spindled nuclei. Areas of vascular malformation in the form of dilated and congested blood vessels admixed with medium to large sized nerve bundles were also seen. Surrounding tissue showed chronic inflammatory infiltrate comprising of lymphocytes and plasma cells (fig 2 & 3).

On immunohistochemistry tumor show positivity for CD 117 and vimentin, (fig 4 & 5) and negativity for S100, SMA Final diagnosis was Low grade gastrointestinal tumor with Arteriovenous malformation.

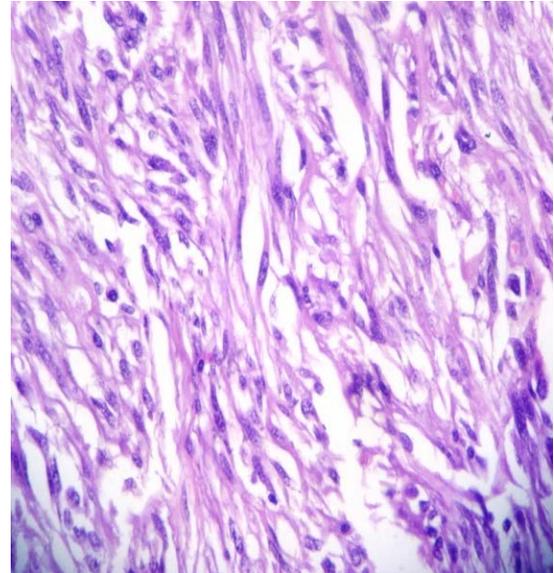


Figure 2: Photomicrograph of GIST showing spindle shape cells. (Hemotoxylin-eosin stain, x400).

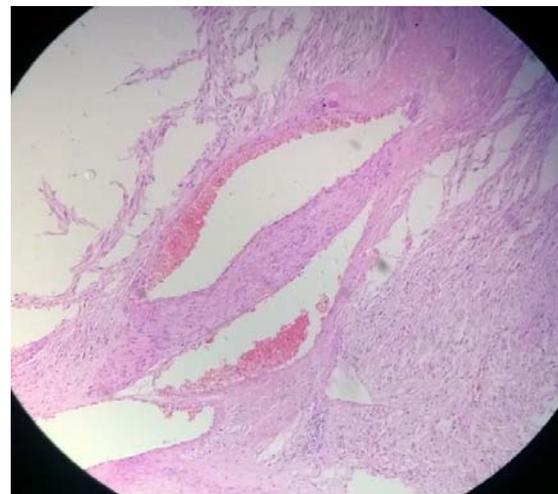


Figure 3: Photomicrograph of GIST showing AV malformation. (Hemotoxylin-eosin stain, x400).

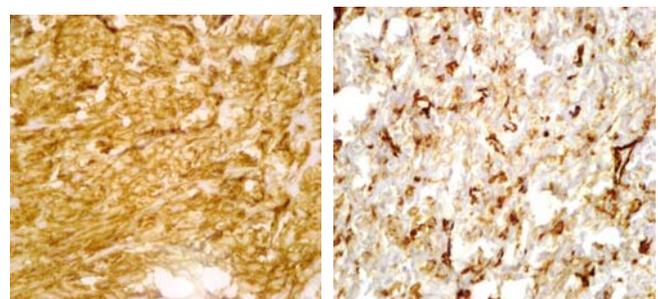


Figure 4 & 5: Strong and vimentin immunopositivity reactivity for GIST.

Procedure	Laparotomy and excision
Tumor site	Jejunum
Tumor size	9x8x5 cm
Tumor focality	Unifocal
GIST subtype	Spindle cell
Mitotic rate	<5/50HPF
Necrosis	Not identified
Histologic grade	G1: Low grade
Risk assessment	Intermediate risk
Margins	Proximal resection margin: Not involved by tumor Distal resection margin: Not involved by tumor Circumferential margin: Involved by tumor
Pathologic staging (pTNM)	pT3: tumor more than 5cm but not more than 10cm pNx: Cannot be assessed
Ancillary studies	Immunohistochemical studies: KIT(CD117)-Positive
Preresection treatment	Not known

Discussion

GISTS are although rare but most common mesenchymal tumors of GIT. The Incidence of GIST is approximately 10-20/million people per year ⁷. Common site of AVMs of the GI tract are cecum and the right colon 78%, followed by 10.5% in the jejunum, 5.5% in the ileum, 2.3% in the duodenum, 1.4% in the stomach and 0.9% in the rectum. Only 0.9% was found in the pancreas ⁸. AVMs of the GI tract may be asymptomatic, or may present with pain or GI bleeding. Bleeding usually originates from a ruptured esophageal or gastric varix secondary to portal hypertension. The prognosis of GISTS depends upon tumor site, size and mitosis.

Conclusion

Although many improvements have been made in diagnostic modalities, but in cases of small intestinal AVMs diagnosis is still very challenging. In our case, CECT shows AVM in wall of gut along with a mass lesion which was diagnosed as gastrointestinal stromal tumor with focal areas of AVM on histopathological specimen. AVM was the cause of bleeding, and the concomitant GIST may have contributed to the symptomatology of the AVM and made it more easily seen on imaging.

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