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A RARE CASE OF SIGMOID COLON GIST- IMAGING PRESENTATION

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Abstract : Gastrointestinal stromal tumours (GIST) are the most common mesenchymal tumour of the gastrointestinal tract. They account for approximately 5 percentage of all sarcomas. They respond remarkably well to ST-571 (Imatinib). A 48 year old male presented with dull aching pain localized to the right lumbar and right iliac region for 1 month duration. CECT abdomen and pelvis in a 4 slice CT was performed. An imaging diagnosis of sigmoid colon GIST was made which was confirmed post operatively with Histopathology and Immunohistochemistry.

Keyword : Gastrointestinal stromal tumour (GIST), large bowel, sigmoid colon, exophytic, atypical imaging features

INTRODUCTION

Gastrointestinal stromal tumours (GIST) are the most common mesenchymal tumour of the gastrointestinal tract. GISTs are rarely seen in patients before the age of 40. Many tumours are incidentally identified on imaging for other indications. Some tumours on the other hand are aggressive and present with metastases or symptoms relating to local disease. Additionally, and more so in large tumours, ulceration seen in 50 percentage of tumours and haemorrhage may occur, with presentation relating to gastrointestinal tract bleeding. GISTs are believed to arise from the the interstitial cells of Cajal. Common sites of involvement include: stomach: 70 percentage, small intestine:20 to 25 percentage, anorectum: 7 percentage, oesophagus: 5 percentage. Sigmoid colon is a very rare site.

CASE REPORT

A 48 year old male presented with dull aching pain localized to the right lumbar and right iliac region for 1 month duration. He had no complaints of loss of weight or appetite and altered bowel habits. Per abdomen examination revealed mild fullness in the umbilical region and pelvis. USG revealed a hypoechoic mass with areas of calcification posterior to the bladder and extending into the pelvis. Colonoscopy was performed and it was normal. CECT with rectal contrast revealed a 12 x 4 cm well defined, soft tissue dense enhancing lesion with multiple foci of calcification which was seen predominantly in the pelvis posterior to the bladder and anterior to the rectum. The lesion was arising exophytically from the sigmoid colon causing extrinsic compression of the sigmoid colon. However there was no obvious intraluminal mass or bowel wall thickening in the large bowel. No evidence of lymph nodal or other secondaries were present. An imaging diagnosis of exophytic GIST arising from the sigmoid colon was given.

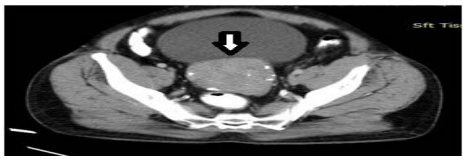

FIGURE 1

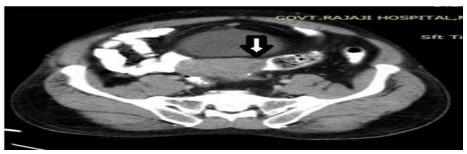

FIGURE 2

FIGURE 1 AND 2 : Axial CECT images with rectal and intravenous contrast shows a well defined, soft tissue dense enhancing lesion with multiple foci of calcification arising exophytically from the sigmoid colon causing extrinsic compression of the sigmoid colon

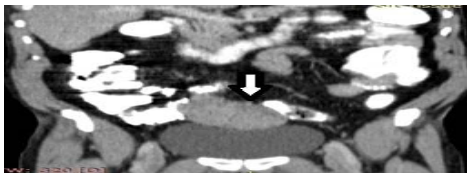

FIGURE 3

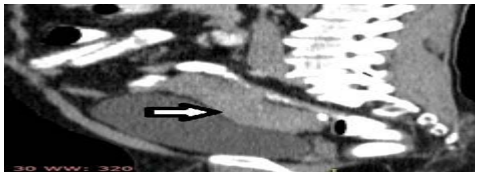

FIGURE 4

Figure 3 and 4 : Coronal and Sagittal reconstructed CECT images well depicting the exophytic GIST arising from the sigmoid colon causing extrinsic compression of sigmoid

Intra operative findings confirmed an exophytic, well defined mass lesion arising from the sigmoid colon. The lumen was intact. Sigmoid colectomy was performed with anastomosis. Gross specimen was a well defined, lobulated mass lesion arising from the sigmoid colon. Histopathology report revealed spindle cell type of GIST. Immunohistochemistry was positive for KIT (CD 117). Hence the diagnosis of GIST was confirmed. The patient had no complaints in the postoperative period.

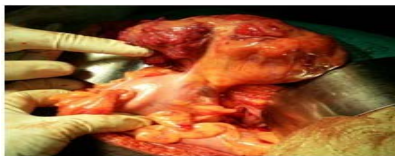

FIGURE 5

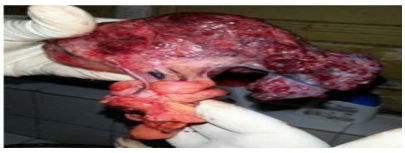

FIGURE 6

Figure 5 and 6: Intraoperative images showing an exophytic mass lesion arising from the sigmoid colon and sigmoid colectomy done

DISCUSSION

Gastrointestinal stromal tumors are the most common mesenchymal tumours arising from the gastrointestinal tract. They are common in the age group 50 to 60. They are rarely seen below the age group of 40. They are usually sporadic. When they present in a younger age group and in multiple numbers syndromic associations must be considered. The common associations are with Carney triad (extra adrenal paraganglioma, pulmonary hamartoma and GIST) and Neurofibromatosis

1. Clinical presentation varies. It may be asymptomatic or present as tumour induced bleed leading to anemia. Symptoms may relate to the mass effect caused by the tumor such as dysphagia, abdominal mass, bowel obstruction and rarely as bowel perforation. Common sites of involvement include: stomach: 70 percentage, small intestine: 20-25 percentage, anorectum: 7 percentage, oesophagus: 5 percentage. Sigmoid colon is a very rare site. GIST can also arise from the mesentery, omentum and retroperitoneum, which is called extra-gastrointestinal GIST. Gross pathology : Size vary between 1 to more than 20cm.It may be a Submucosal, intramuscular or subserosal mass. Usually, it is well circumscribed but lacks a true capsule. The cut surface is gray to pink in colour. Areas of cystic degeneration, infarction and hemorrhage and necrosis may be seen. They are prone to surface ulceration and bleeding. The diagnosis of GIST is based on cellular morphology and immunophenotype. Histologically, there are three types of GISTs: spindle cell (70 percentage), epithelioid (20 percentage), and mixed . Spindle cells type: They are short, uniform cells, blunt ended with eosinophilic cytoplasm. Paranuclear vacuole may be present. They are arranged in sheets, fascicles whorled, storiform or palisaded patterns. Most gastric GISTs, some of small and large bowel GISTs are of spindle cell type. Epithelioid type: Epithelioid GISTs were previously were known as epithelioid leiomyoblastoma. They are usually noted in gastric antrum. Cells have abundant cytoplasm, round nuclei, cytoplasmic vacuoles and are arranged in sheets or nests. Clear cell, signet ring, oncocytic and plasmacytoid variants may be noted. Immunohistochemistry : Approximately 95 percentage of GISTs carry an activating somatic mutation of CD117 (c-kit) . CD117 is a transmembrane receptor tyrosine kinase encoded by the proto-oncogene c-kit located on chromosome 4 .Positive immunohistochemical staining for CD117 is a defining characteristic of GISTs. Tumor size and mitotic index are the two most important prognostic factors used for risk stratification of GISTs. Malignant potential of GIST is increased if the tumor size exceeds 5 cm and a mitotic index of >5/50 hpf. GIST arising from the stomach has a comparatively less chances of progressing to malignancy when compared to other sites. GIST arise from the interstitial cells of Cajal. 95 percentage are positive for CD 117(c-kit) and 70 percentage for CD 34. Imaging findings are variable in case of GIST. CECT is the best modality for diagnosing GIST. It presents usually as a relatively rounded/ovoid mass with well defined borders. It may be endoluminal or exophytic. Homogenous enhancement pattern is seen in small tumors. Heterogenous enhancement is seen in larger masses representing areas of necrosis and heamorrhage. Calcification is rarely seen in GIST. In MRI, GIST are T1 : hypointense, T2 : iso to hypointense, with areas of hyperintensity which represent necrosis. T1 post contrast image shows enhancement of the solid component and non enhancing areas of necrosis. 15 to 45 percentage of patients present with metastasis. Liver is the most common site followed by peritoneum and omentum. Liver metastasis are commonly cystic in nature. Lymph node, bone and brain metastasis are rare. Treatment includes resection and Imatinib therapy. PET plays an important role in assessing the response of the tumor to chemotherapy. Imaging differential diagnosis includes gastrointestinal leiomyoma, gastrointestinal lymphoma, gastrointestinal schwannoma and calcifying fibrous tumour. Leiomyoma commonly arises from the esophagus presenting as ovoid intramural solitary mass with a smooth surface and the presence of calcifications is almost pathognomonic. The typical presentation of a gastrointestinal lymphoma is a thick walled infiltrating mass with aneurysmal dilatation without obstruction. Aneurysmal dilatation is based upon destruction of the bowel wall and the myenteric nerve plexus.

The radiologic features are classified as mucosal nodularity, endo-exoenteric mass, intraluminal mass, mural infiltration, and mesenteric invasion. Schwannoma is very rare mesenchymal tumor which is typically seen as a well-defined, rounded, mural mass with homogeneous attenuation and tends to lack cystic change and haemorrhage. It most commonly arises in the stomach, colon. Calcifying fibrous tumor (CFT) is a rare benign mesenchymal tumor which presents a densely calcified well defined mass.

SUMMARY

This is a rare case of GIST arising from the sigmoid colon. The imaging features consisted of a well defined, homogenously enhancing mass lesion arising exophytically from the sigmoid colon and which had multiple foci of calcification ,which is a relatively rare presentation . Thus it was a case of GIST, which is the most common mesenchymal tumour arising in the gastrointestinal tract but arising from a rare location – sigmoid colon and with atypical imaging presentation of multiple foci of dense calcification within the mass. This explains the variable imaging presentation of GIST. Hence a diagnosis of GIST may be considered even when it arises from uncommon locations and presents with atypical imaging findings.

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