Journal of Clinical and Biomedical Sciences

Case Report

Synchronous presentation of gastrointestinal stromal tumor of stomach and neuroendocrine tumor of Jejunum as acute intestinal obstruction.

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Abstract

Neuroendocrine tumors (NETs) arise from the diffuse components of the endocrine system A patient with jejunoileal NETs usually presents with vague abdominal complains which may progress to intestinal obstruction. Gastrointestinal stromal tumors (GIST) are mesenchymal tumors arising from interstitial cell of Cajal or pacemaker cells. GIST is the most common mesenchymal tumor of the abdomen. We are here by reporting a rare synchronous tumor for awareness and better management of patents.

Keywords: Synchronous, Gastrointestinal tumors, stomach, Jejunum, NETs.

Introduction

Neuroendocrine tumors (NETs) arise from the diffuse components of the endocrine system. Small intestine is the most common site for NETs within the gastrointestinal tract (GI), accounting for nearly 50% of all GI NETS. [1, 2, 3] The peak incidence of GI NETs is in the sixth decade, but they may appear at any age. Symptoms are due to hormones released by the tumor cells. A patient with jejunoileal NETs usually presents with vague abdominal complains which may progress to intestinal obstruction.[4, 5, 6]

Gastrointestinal stromal tumors (GIST) are mesenchymal tumors arising from interstitial cell of Cajal or pacemaker cells. GIST is the most common mesenchymal tumor of the abdomen, with an annual incidence between 11 and 20 per 1 million people.[2, 3, 7] More than half of these tumors occur in the stomach. GISTs are diagnosed at a peak age of 60 years, with fewer than 10% occurring before 40 years of age. Approximately 75% of all GISTs have oncogenic, gain of-function mutations in the receptor tyrosine kinase KIT. Another 8% of GISTs have mutations that activate the related receptor tyrosine kinase, platelet-derived growth factor receptor α (PDGFRA).[3, 6, 7]

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Conflict of Interest: None
Financial Aid: Nil

Case report

60 year old female presented with pain abdomen, vomiting and constipation. These symptoms were present on and off since six months and worsened from the past five days. Pain abdomen was exacerbated by eating and relieved by vomiting. Past history was not significant. Vital signs were within normal limits. On examination, tenderness was present in the pereumbilical region. On per rectal examination, there was no tenderness.

Laboratory investigations revealed leucocytosis with neutrophilia with normocytic blood picture. Biochemical investigations were within normal limits. Abdominal x-ray revealed a dilated small bowel loop with intestinal pneumatosis. Upper Gastrointestinal endoscopy revealed large nodular mass in lesser curva-

ture of stomach which is bulging into the mucosa. Contrast enhanced computed tomography (CT) of upper abdomen showed a large nodular lesion in the lesser curvature of the stomach. CT also showed dilatation of the small bowel loop with intestinal pneumatosis and wall thickening.

Patient underwent exploratory laparotomy; wedge resection of gastric tumor was done along with segmental enterectomy. Thickened and dilated segment of small bowel was resected and side to side anastomosis was performed.

Pathological examination

Wedge resected gastric mass measured 7x5x5 cm, gastric mucosa showed focal ulceration. Beneath the mucosa, there was a well circumscribed grey white lesion with cut surface showed whorling pattern (Figure 1). Microscopic examination of this lesion showed well circumscribed lesion, composed of spindle cells having elongated bland nuclei with indistinctive cytoplasm. These cells were arranged in fascicles. Areas of collaginization, myxoid change and focal inflammatory cells were also seen in the section studied. Mitosis was less than 2 per 10 high power fields. These features were of gastrointestinal stromal tumour of uncertain malignant potential (Figure.2).

Small intestinal segment measured 23cm, identified a stricture. Also observed a well-defined grey white lesion measuring 1x1cm in the wall of the small intestine, corresponding to the area of stricture. Microscopy showed monomorphic tumor cells having salt and pepper nuclear chromatin, scanty cytoplasm arranged in nesting pattern. Tumor cells were seen beneath the sub mucosa extending up to sub serosa (Figure 3 & 4). Diagnosis of Neuroendocrine tumor grade one was given.

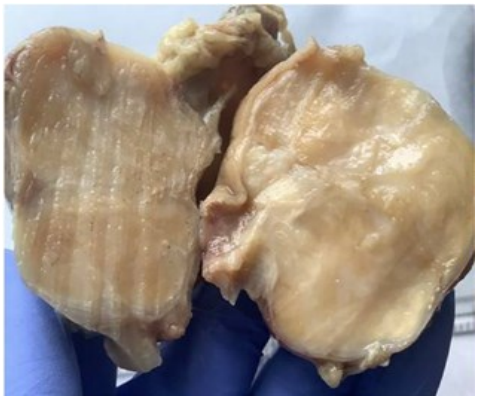

Figure 1: Well circumscribed lesion beneath the gastric mucosa

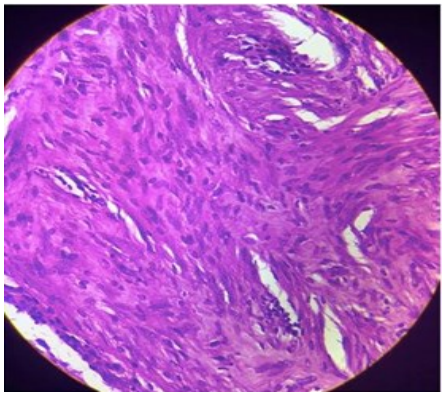

Figure 2: Spindle cells with elongated cigar shaped nuclei, indistinct cytoplasmic margins, arranged in fascicles. $400_{X,}$ H&E.

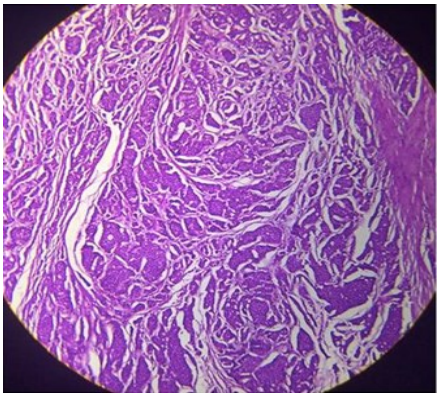

Figure 3: Tumor cells infiltrating muscular layer in nests. $100_{X,}$ H&E.

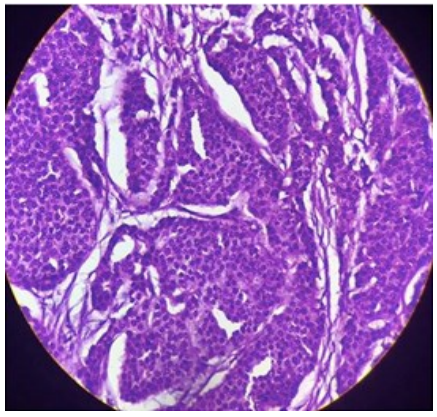

Figure 4: Tumor cells arranged in nests, $200_{X,}$ H&E.

Discussion

Gastrointestinal stromal tumors (GIST) are mesenchymal neoplasms of the gastrointestinal tract-interstitial cell of Cajal. GISTs can arise anywhere along the GI tract. Stomach (50 -62%) is the commonest site followed by small intestine (20-30%), colon (11%) and rectum (7%).[3,6] Occur in all age groups, Peak incidence seen in 5th - 8th decade, with a median age of 60 years.[5,6] 25% of gastric GISTs are clinically malignant, accounts to 2.2 % of all malignant gastric tumors. GISTs composed of thin elongated cells are classified as spindle cell type**,** while GISTs dominated by plumper, epithelial-appearing cells are termed epithelioid type.[5,7,8] The most useful diagnostic marker is KIT (CD117), which is immunohistochemically detectable in Cajal cells and 95% of gastric GISTs.[9,10] Patients with GISTs usually presents with symptoms related to mass effects.

The prognosis depends on tumor size, mitotic index, and location. Gastric GISTs are less aggressive than those arising in the small intestine. Recurrence or metastasis is rare for gastric GISTs smaller than 5 cm but may be seen in mitotically active tumors larger than 10 cm.[10,11]

NETs are group of tumors arising from neuroendocrine cells which secret many bioamines and peptides. Incidence of small intestinal NETs ia about 1.2 per 100000 persons. GIT is the primary site for about 67% of NETs; Jejunal NETs are rare acunting to only 11%, heterogeneous, may be large and locally invasive.[6,7,9] Most patients with small intestinal NETs presents with abdominal pain, intestinal obstruction and metastasis. Carcinoid syndrome develops in fewer than 10% of patients and is caused by vasoactive substances secreted by the tumor into the systemic circulation.[6] The most important prognostic factors for GI neuroendocrine neoplasms are degree of histological differentiation, mitotic rate, and Ki-67 proliferative index. These tumors are rarely more than 2 cm, deep infiltration in to muscular layer and sub serosa is commonly seen. Most patients will have prolonged survival due to low proliferative rate.[5, 6,10]

Synchronous GIST and neuroendocrine tumors of jejunum are extremely rare. The pathogenesis of NENs associated with other secondary malignancies remains unclear. Diverse theories have been developed such as, existence of a common carcinogenic effect that stimulates the growth Of NETS and secondary primary tumor or by a common stem cell which may undergo similar

Genetic mutations giving rise to different types of gastrointestinal malignancies.[3, 7, 6] Other theory proposed is that the paracrine or autocrine growth effects by secretary peptides by NET cell tumors influence tissue growth with subsequent transformation into neoplastic cells.[11, 12] Role of Helicobacter pylori is also related to the coexistence of these two tumors.[3] The most common type of secondary primary malignancy seen with NETs are adenocarcinomas (49.4%), followed by GIST (13.5%), other NENs in different GIT segments (7.9%), lymphomas (6.8%) and Squamous cell carcinoma(4.5%).[6,13] Knowledge of occurrence and pattern of synchronous GIST and other neoplasm's is important for both histopathologists and surgeons for patient's management.

Conclusion

Synchronous GIST and Neuroendocrine tumors of jejunum are extremely rare. Awareness of synchronous GIST and other neoplastic tumors is important for both the surgeons and Pathologist for appropriate patient management.

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