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Exophytic Gastrointestinal Stromal Tumor with Cystic Changes in the Pancreas

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Abstract:

The term stromal tumor was coined in 1983 by Clark and Mazur for smooth muscle neoplasm of the gastrointestinal tract (GIT). Gastrointestinal stromal tumors (GIST) are nonepithelial tumors arising from the interstitial cells of Cajal, which express KIT protein-CD117 on immunohistochemistry. GIST can arise anywhere in the GIT, including the mesentery, omentum, and retroperitoneum. We present a case of multiple stomach gist with cystic changes in the pancreas.

Keywords: ***Stomach GIST, Pancreatic cyst.***

Introduction

Before 1983, gastrointestinal stromal tumors (GIST) were classified as smooth muscle tumors, along with leiomyoma, leiomyoblastoma, and leiomyosarcomas. With advances in electron microscopy and immunohistochemistry, it is now known that they are nonepithelial tumors arising from the interstitial cells of Cajal. Gastrointestinal stromal tumor (GIST) is the most common type of mesenchymal tumor in the gastrointestinal (GI) tract, with a disease incidence of 10-20 per million individuals worldwide.[1-3] GIST can occur in any region of the digestive tract and the incidence of GIST in the stomach, small intestine, large intestine and esophagus is reported to be 60–70, 20–30, 18.1 and 1.4%, respectively. According to the tumor location, GISTs are classified as endoluminal, exoluminal, intramural and mixed types.[4] Immunohistochemical findings and the ultramicrostructure of GIST cells are similar to those of Cajal cells, which are autonomous nerve-related GI pacemaker cells that regulate intestinal motility.[5,6] On diagnosis, immunohistochemical analysis revealed the presence of cluster of differentiation (CD)117-positive and CD34-positive/negative tumor cells. The most typical characteristic of malignancy is infiltration of neighboring organs or lymph node metastasis. Infiltration of the lamina propria mucosae or the muscular layer is an important indicator for the diagnosis of malignant GIST. In addition, manifestation of the malignancy includes a large tumor size (diameter of >5 cm for gastric tumors and >4 cm for small intestine tumors), obvious mitosis count [>5/50 high-power fields (HPFs)],[7] high density of cells, infiltration of the lamina propria mucosae, presence of coagulative tumor necrosis,[8] high Ki-labeling index (>5%),[9,10] recurrence and metastasis.

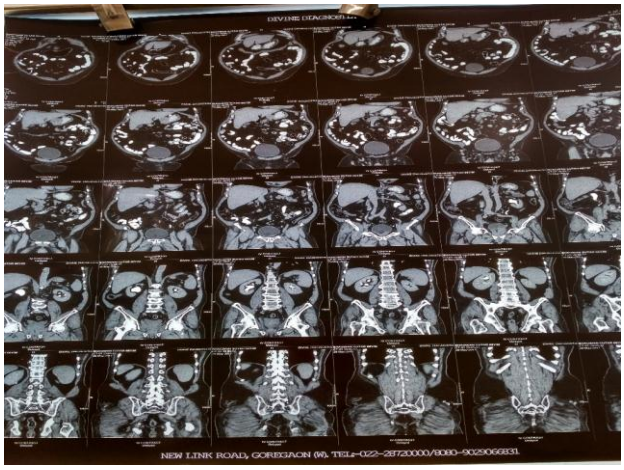

Figure 1: Showing the Multiple GIST with pancreatic cyst

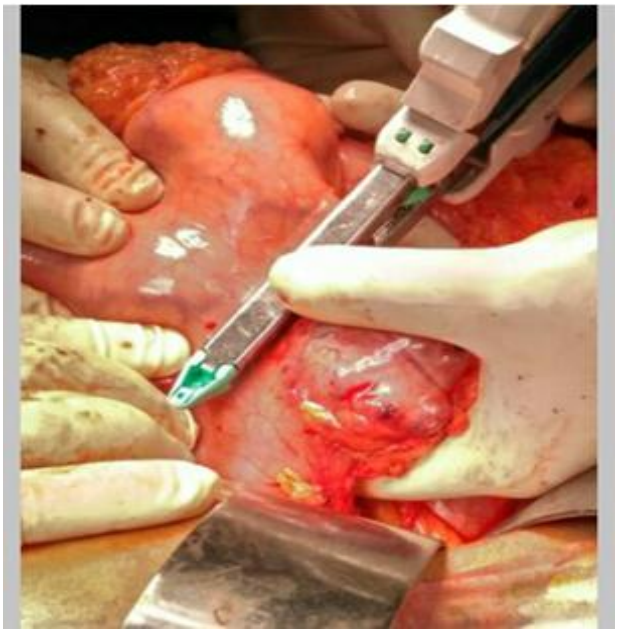

Figure 2: Showing the stomach GIST

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Case Report

66 yrs old male came with complaints of pain in abdomen and lump in abdomen since 3 months. On examination a lump of 4x3x1 cm was felt in the epigastric region. CT scan report was s/o enhancing soft tissue mass lesion arising from posterior medial wall of stomach with non-enhancing areas of calcification s/o GIST [Figure 1]. 2 triangular lobulated non enhancing hypodense areas involving distal body and tail of pancreas s/o walled off necrosis vs cystic neoplasm of pancreas.

Pt was admitted and thoroughly investigated. EUS was done, report was s/o gastric GIST with serous cystadenoma of pancreas. Fluid was aspirated from cyst and sent for CEA and amylase. Both were within normal limits. Biopsy was taken s/o spindle cell tumourfavouring GIST. Serum tumour markers were sent ie CEA, AFP, CA 19-9 all were within normal limits. Pt was posted for exploratory laparotomy. Abdomen opened in layers with a midline incision. Omentum, peritoneum and rest of the bowel were normal. Two gist of size 4x4 and 2x2 cm noted along the lesser curvature [Figure 2]. Lesser and greater sac opened, posterior wall of stomach assed and was normal. Pancreas exposed and two small simple cyst noted.75 blue stapler used to excise the gist from the lesser sac. Drain placed and the abdomen closed in layers. The patient tolerated the procedure well and was discharged the 8th postoperative day.

Discussion

Gastrointestinal stromal tumors (GISTs) are rare neoplasms, with an annual incidence of approximately 4 per million.[3] Historically, these tumors were classified as leiomyomas, leiomyoblastomas, and leiomyosarcomas, because of a mistaken belief that they originated from smooth muscle in the wall of the gastrointestinal tract.[2]

The cellular origin of GIST recently has been proposed to be the interstitial cell of Cajal, an intestinal pacemaker cell. This postulate is supported by the finding that GISTs display positivity for cell markers similar to those of the normal cell of Cajal.[4-6] The majority (approximately 95%) of GISTs express the CD117 antigen (KIT), a protooncogene product. CD34, a commonly expressed human progenitor cell antigen, is also frequently found positive in GISTs.[3] More than half of the GISTs are located in the stomach followed, by the small intestine, colon and rectum, and esophagus.[1,3,7]

Complete tumor resection with disease-free resection margins is the treatment of choice for primary nonmetastatic tumors. Lymphadenectomy is not recommended because lymph node involvement is rare. Wedge resection allows full-thickness resection of the stomach wall containing the tumor, with negative resection margins.[1,6]

Grossly, GISTs vary greatly in size and can be more than 30 cm in diameter. These tumors are usually well circumscribed and unencapsulated. GIST can grow in an endophytic or exophytic pattern. They are usually solid. Small cysts are frequently observed, presumably as a consequence of cystic degeneration or necrosis. Larger stromal tumors usually degenerate, and cysts are formed.[8-11]

In the present case, the large size of the cyst obscured the origin from the stomach. Imaging showed that the tumor was not originating from the pancreas or any other organ, so the exact origin of the tumor could not be determined preoperatively. Imaging demonstrated no vascularity of the lesion. Since malignancy could not be excluded in our case and the origin of the tumor could not be determined by imaging, surgery was indicated.

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Lesions that should be considered in the differential diagnosis of a cystic abdominal mass on radiologic imaging (CT and MRI) include gastric or bowel duplication cysts, cystic mesothelioma, cystic lymphangioma, cystic mucinous retroperitoneal tumors, cystic pancreatic tumors, pseudocysts of the pancreas or peritoneum, cystic teratoma, and GIST.[12-14]

In this case, tumor cells showed diffuse and strong positivity for CD117 (KIT) and CD34, which was consistent with a diagnosis of GIST.

Wang et al. recently published a series of 7 patients with cystic GISTs and analysis of c-kit and PDGFRA gene. Gene mutation of exon 11 of c-kit was identified in 3 cases.[15] PDGFRA mutant GISTs arise almost exclusively in the stomach, whereas KIT mutant tumors occur at a variety of sites along the gastrointestinal tract. PDGFRA exon 14 mutations may be associated with a reduced risk of recurrence. Limited clinical data are published, but PDGFRA exon 14 mutant GISTs appear; sensitive to imatinib, the sensitivity is similar to KIT exon 11 mutants.[16-18]

In summary, GISTs with cystic appearance clearly should be considered in the differential diagnosis of cystic abdominal tumors. Most GISTs (95%) express Kit (CD117) and CD34 (70%). In case of doubt gene mutation analysis is necessary. KIT and PDGFRA genotyping is important for GIST diagnosis and assessment of sensitivity to tyrosine kinase inhibitors.

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Conflict of Interest: Non to Declare

References

[1] Kim KM, Kang DW, Moon WS, Park JB, Park CK, Sohn JH, Jeong JS, Cho MY, Jin SY, Choi JS, Kang DY Gastrointestinal Stromal Tumor Committee; Korean Gastrointestinal Pathology Study Group. Gastrointestinal stromal tumors in Koreans: it's incidence and the clinical, pathologic and immunohistochemical findings. J Korean Med Sci. 2005; 20:977-984. [PMC free article] [PubMed].

[2] Tryggvason G, Gíslason HG, Magnússon MK, Jónasson JG. Gastrointestinal stromal tumors in Iceland, 1990–2003: the icelandic GIST study, a population-based incidence and pathologic risk stratification study. Int J Cancer. 2005; 117:289-293. [PubMed].

[3] Goettsch WG, Bos SD, Breekveldt-Postma N, Casparie M, Herings RM, Hogendoorn PC. Incidence of gastrointestinal stromal tumours is underestimated: results of a nation-wide study. Eur J Cancer. 2005; 41:2868–2872. [PubMed].

[4] Miettinen M, Lasota J. Histopathology of gastrointestinal stromal tumor. J SurgOncol. 2011; 104:865–873. [PubMed].

[5] Hirota S, Isozaki K, Moriyama Y, et al. Gain-of-function mutations of c-kit in human gastrointestinal stromal tumors. Science. 1998; 279:577–580. [PubMed].

[6] Heinrich MC, Corless CL, Duensing A, et al. PDGFRA activating mutations in gastrointestinal stromal tumors. Science. 2003; 299:708–710. [PubMed].

[7] Emory TS, Sobin LH, Lukes L, Lee DH, O'Leary TJ. Prognosis of gastrointestinal smooth-muscle (stromal) tumors: dependence on anatomic site. Am J SurgPathol. 1999; 23:82–87. [PubMed].

[8] Lerma E, Oliva E, Tugués D, Prat J. Stromal tumours of the gastrointestinal tract: a clinicopathological and ploidy analysis of 33 cases. Virchows Arch. 1994; 24:19–24.[PubMed].

[9] Rudolph P, Gloeckner K, Parwaresch R, Harms D, Schmidt D. Immunophenotype, proliferation, DNA ploidy, and biological behavior of gastrointestinal stromal tumors: a multivariate clinicopathologic study. Hum Pathol. 1998; 29:791-800. [PubMed].

[10] Ando N, Goto H, Niwa Y, et al. The diagnosis of GI stromal tumors with EUS-guided fine needle aspiration with immunohistochemical analysis. GastrointestEndosc. 2002; 55:37-43. [PubMed].

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