"Multi-cystic gist of stomach: an unusual presentation as obstructive jaundice."

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Abstract:
A 42 yr old male presented with pain in upper abdominal fullness and pain and jaundice. Upper GI endoscopy suggestive of extra-luminal compressing mass at antrum. On laparotomy there was multiple cystic swelling along the lesser curvature largest cyst 6 x 4 cms. There was three sub-mucosal swelling near antrum causing narrowing. The patient was histo-pathologically reported as gastrointestinal stomal tumor(GIST), which is present 0.1% of all colorectal cancer. GIST is mostly identified incidentally during endoscopic screening in the absence of symptom. GIST in comparison with leiomyoma and leiomyosarcoma is on the basis clinic-pathologic, immunohistochemical and molecular genetic study. Mostly malignant GIST run a very slow course and have recurrence and metastasis over 10-15 years. Thus long term follow-up is necessary. Unless proved other-wise gastric and colonic lesions are considered as malignant.

Key words: Gastrointestinal stomal tumor, gastric malignancy, CD117 antigen, Cajal cell tumor, S-100.

Background: The gastrointestinal stomal tumors (GIST) may arise any-where in the tubular gastrointestinal tract, from esophagus to the rectum. In-addition, it has been appreciated in extra-GI locations, principally mesentery, omentum and retro-peritoneum. In terms of detailed distribution, 5% in the esophagus, 50-60% in the stomach, 20-30% in the small intestine, 10% in the large bowel, 5% else-where in the abdominal cavity (gallbladder, bladder, mesentery, omentum and retro-peritonium).

Case Report:
A 42 yr old male presented with pain in upper abdominal fullness and pain and jaundice. Patient was investigated for that, had normal hematological reports except raised serum bilirubin 6 IU which was suggestive of obstructive jaundice. Upper GI-scopic shows narrowing at antrum of stomach with normal mucosa. CT abdomen suggestive of cystic swelling 6 x 4 cm size between head of pancreas and lesser curvature of stomach. Large cyst compressing over lower common bile duct and partial narrowing at antrum. On laparotomy, there was multiple cystic swelling along the lesser curvature largest cyst 6 x 4 cms. There was three sub-mucosal swelling near antrum causing narrowing. Distal gastrectomy with Bilroth II procedure was done. There was no rupture of any cyst during surgery.

The cysts contained clear fluid. On HPR the cyst was positive for low grade Malignant GIST. Immunohistochemistry showed c-kit-positive CD34-
positive smooth muscle actin (SMA)-negative, and S-100-negative staining of tumor cells. The final diagnosis was gastrointestinal stromal tumor (GIST). Postoperative period was uneventful and patient discharged on 8th day. Since last 8 months patient is on Imatinib. After Six month CT not show any evidence of recurrence.

**Discussion:** The GIST is present 0.1% of all cancer. Sixty five % of cases are identified incidentally during endoscopic screening in the absence of symptom. Most of the tumors are silent until it reaches a large size, at which point it may cause non-specific abdominal pain, discomfort or become recognized as a palpable lump. The diagnosis of GIST is often suspected histological & imuno-histochemistry, because the majority of cases have remarkably uniform appearance. The cystic GIST have a benign to malignant ratio of 1:2.4,9 Unless proved otherwise gastric and colonic lesions are considered as malignant.10,11 Mucosal invasion is very rare and it can be diagnosed as extra-luminal compression on endoscopy. Invasion between smooth muscle fibers and bundles is a characteristic growth pattern. This may be related to the GISTs origin from Cajal cells or related stem cells, the non smooth muscle elements between the layers of the muscularis propria.12 The low cellularity has emerged as a favorable prognostic feature. The mitotic activity >5/50 HPFs or the tumor >5 cm size have a high risk for intra abdominal or hepatic metastasis. Nuclear atypia is generally not prominent in GISTs, and its significance is unclear.13 But prognostic relevance of cell type is seems limited. CT-abdomen is used to define correctly the site, size and structure of lesions in all cases and to identify signs of invasion of neighboring structures in some cases.14 On immune-histochemistry, in contrast to true smooth muscle tumors, GISTs are usually positive for expression of the KIT receptor tyrosine kinases (detected as CD117 antigen) and CD34, variably positive for smooth muscle actin and usually negative for desmin. Unlike Shwanomas GISTs are usually negative for S100 protein. The prevalence of c-KIT mutation in GIST is as high as 90%.15 Imatinib (STI-571) selectively inhibits the ABL, BRC-ABL, KIT and PDGF receptor tyrosine kinases. The Imatinib is a targeted therapy directed against the apparent fundamental and pathogenic defect in the GIST. Adjuvant therapy with STI-571 will improve outcome if applied earlier in course of GIST.15, Radiotherapy (5040 cGy) has limited role, in positive microscopic margins in gastric and rectal GIST.16,17

The primary goal of surgery is complete resection of the disease with avoidance of tumor rupture. Unlike intestinal adeno-carcinoma GIST rarely metastases to lymph-nodes, so lymphadenectomy is seldom warranted. The disease specific survival rate of asymptomatic GIST patient is 81.7% at 5 years. The patient with primary tumor without metastasis underwent complete gross resection having 54% 5 year disease specific survival rate, but if the tumor of size >10 cm, it will only 20%.16 Prognosis of the GISTs depends on anatomic sites, tumor size, mitotic rate and patient age and tumor rupture during surgery.11,16 Most malignant GISTs run a slow course, with recurrence and metastasis developing over
years, give rise to peritoneal cavity or hepatic metastasis. Sometimes it may be 10 to 15 years after primary surgery. Thus long term follow-up is necessary.

References:


