We recommend ERCP and stenting in the initial management of such patients.

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Periampullary gastrointestinal stromal tumor presenting with obstructive jaundice

Introduction

GIST is the most common mesenchymal neoplasm of the gastrointestinal (GI) tract representing 0.1-3% of all gastrointestinal malignancies. GIST cells are thought to arise from a common precursor cell which also gives rise to intestinal cells of Cajal. GIST results from activating mutations in receptor protein kinase: either KIT (CD 117) or PDGFRA (platelet derived growth factor receptor alpha). The stomach and intestine are common sites of GIST. The periampullary region is a very rare site for GIST. To the best of our knowledge, only three cases of periampullary GIST have been reported before.[1,2,3]

Methods

A 48-year-old man presented with progressive jaundice, anorexia and weight loss for 2 months, fever and abdominal pain for 7 days. On examination the patient was pale and icteric. There was firm, tender hepatomegaly 5 cm below the subcostal margin. Investigations showed microcytic hypochromic anemia (hemoglobin-6.3g%) with leucocyte count 12900cells/mm^3 and neutrophils 85%. The stool was positive for occult blood. The patient had conjugated hyperbilirubinemia with total and direct bilirubin of 4.6 mg/dL and 4.0 mg/dL, respectively and alkaline phosphatase of 693U/L. Ultrasonography of the abdomen showed liver size 16 cm with dilatation of IHBR and common bile duct and 8.8×4.8×5.5 cm heterogenous hyperechoic mass near the head of pancreas in the periampullary region. CECT abdomen revealed 8.3 cm x8.1 cm well defined lesion in the region of the head of pancreas (**Figure a**). MRCP showed a well-defined lobulated hyperintense lesion in the region of head and uncinate process of pancreas with sudden cutoff of common bile duct (**Figure b**) and pancreatic duct at the lesion, which was suggestive of malignancy. Upper gastrointestinal endoscopy (**Figure c**) was done and multiple biopsies were taken from the mass. Endoscopic retrograde cholangiopancreatography failed to relieve biliary obstruction because of the large mass at the ampulla.

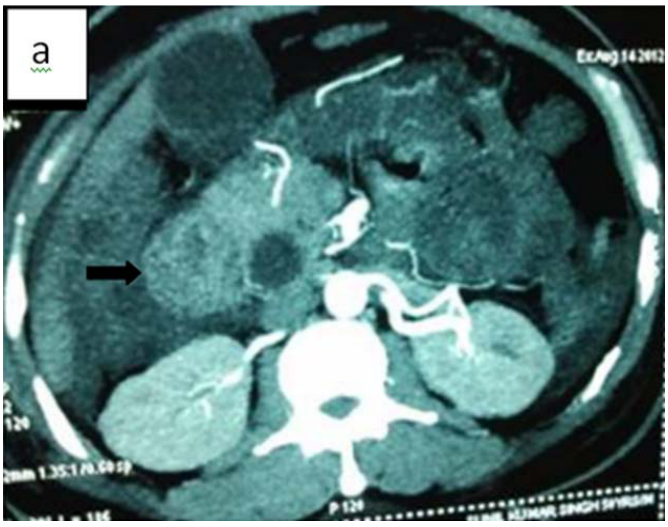

Figure a: CECT abdomen showing 8.3x8.1cm periampullary tumor (arrow)

Figure b: MRCP Image showing dilated CBD and CBD cut off sign.

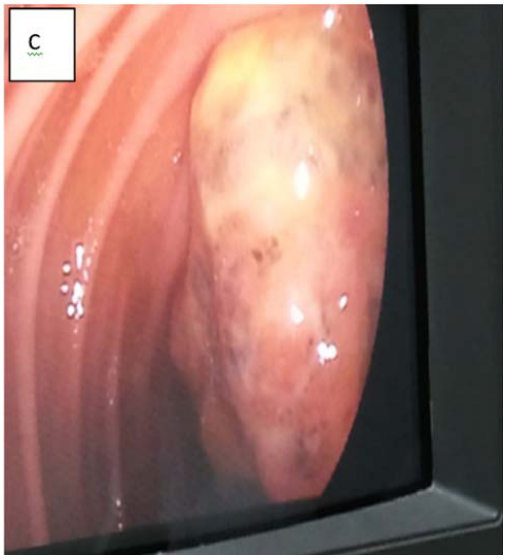

Figure c: Upper Gastrointestinal endoscopy (side view) showing the large periampullary tumor.

Bile mixed with pus was drained on percutaneous transhepatic biliary drainage and its culture was positive for *E. coli*. The patient was treated with antibiotics as per the sensitivity report. Subsequently Whipple's pancreatico-duodenectomy (single loop anastomosis) with feeding jejunostomy was performed.

Histopathology of operated specimen (**Figure d**) revealed a malignant gastrointestinal stromal tumor (GIST) with high mitotic activity (>10/50hpf) with neural differentiation. Immuno-histochemical studies were positive for S-100 and focally positive for CD1, negative for CD34, SMA and c-kit. Patient was started on Imatinib and is stable after 7 months of follow up.

Discussion

The most common site for GIST (gastrointestinal stromal tumor) is the stomach (50-60%) followed by small bowel (20-30%), large bowel (10%), esophagus (5%) and elsewhere in the abdomen 5%. Only 4% of GISTs occur in the duodenum and very rarely in the periampullary region. Mean age of presentation of this tumor is 60 years (40-80 years) with no predilection for either gender. Familial GIST are associated with Neurofibromatosis type I and Carney triad (GIST, paraganglioma, pulmonary chondroma).

Diagnosis of gastrointestinal stromal tumor depends on immuno-histochemical studies. Histologically GIST are classified as neural type, epitheloid type and mixed type.[4] Biological markers used in its diagnosis are c-kit (CD117) with positivity in>95% cases, CD34 positive in 60-70%, PDGFRA (platelet derived growth factor alpha) positive in<5% cases, vimentin and smooth muscle actin positive in 15-60%. Others likedesmin, S-100, keratin are rare. Treatment of primary GIST

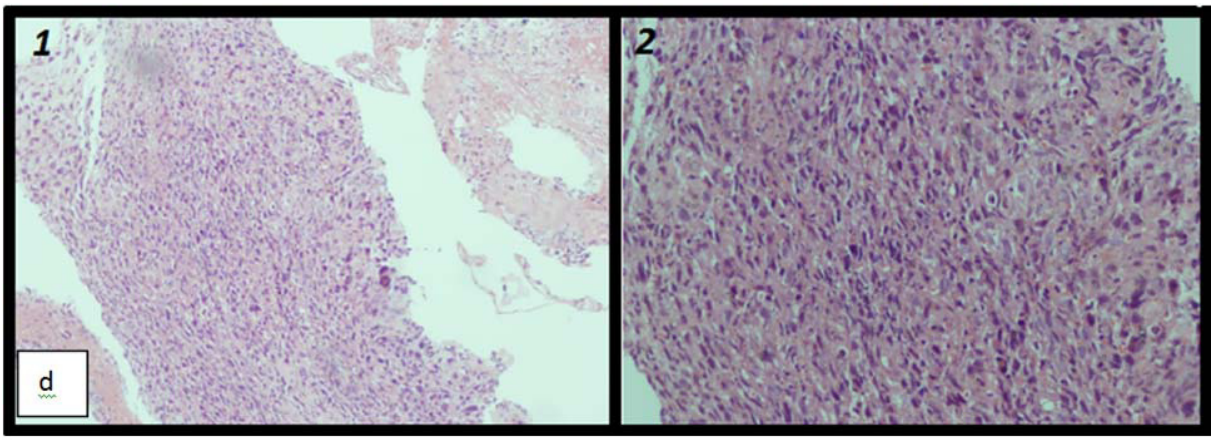

Figure d: H&E staining of the excised GIST. 1-low power. 2-high power. Showing predominantly spindle cells having hyperchromatic nuclei.

is complete surgical resection. Tumor designated as high risk should be treated with adjuvant Imatinib (tyrosine kinase inhibitor). Imatinib is the mainstay of treatment for metastatic or unresectable tumor. Tumor resistant to Imatinib is treated with Sunitinib.

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Primary extraskeletal osteosarcoma of gall bladder

Introduction

Extraskeletal osteosarcoma (ESOS) is an uncommon malignant mesenchymal neoplasm constituting 1% of all soft tissue sarcomas[1] and 2-4 % of all osteosarcomas (OS). Though the primary skeletal OS is a common bone tumor of the first andsecond decades, ESOS has been reported in the elderly (60-70 years).[1] Allan's criteria for diagnosis of ESOSinclude: the tumor, (1) to be ofsoft tissue origin without any attachment to bone/periostium, (2) to have a uniform sarcomatous morphology without carcinomatous area and (3) shouldproduce malignant osteoid, bone or cartilaginous matrix.[2] The present article reports a primary ESOS of gall bladder (GB) in a 72 year old female . To the best of our knowledge this is the second case of the primary ESOS of the GB.[3] A written informed consent was taken in the index case.

Case report

A 72 year old diabeticfemale with coronary artery disease, bronchial asthma, hemiparesis and depression,presentedwith off and on pain on the right hypochondrium, radiating to the ipsilateral sub-scapular region for last 3 years,with loss of appetite and weight (lost10 kgsover 10 months). She had jaundice 2 years back, which resolved spontaneously.Onlocal examination and contrast enhanced computer tomogram (CECT) a distended GB with heterogeneous soft tissue within the lumen anddilated common bile duct (CBD) was noted without calcification or extra-luminal extension (**Figure 1A**). Magnetic resonance cholangiopancreatography (MRCP) also showed similar filling defects in the GB and lower CBD (**Figure 1B**). Radiologically, possibilities considered were: empyema with sludge and calculi or hydatid cyst with rupture. Carcinoma was not considered, as no definite wall thickening or soft tissue extension seen. Ultrasound guided fine needle aspiration cytology (FNAC) showed a few highly atypical cells, suggestive of a malignant tumor. The patient underwent cholecystectomy with partial CBD excisionas it was adherent to the duodenum and on per-operative frozen section examination a positive CBD margin was identified. No lymph node was identified. Grossly a polypoidal friable growth measuring 4.5x4x0.2 cms was identified near fundus of GB, involving the CBD, with attached liver bed measuring 3.2x1.5cms and a free liver margin of 2.1 cms. Microscopically, a pleomorphic malignant tumor infiltrating the wall of GB with overlying normal epithelial lining was seen. Within the tumor, numerous osteoclastic giant cells were noted with significant nuclear pleomorphism and foci of osteoid formation by the malignant cells, as confirmed by Verhoeff-Van Gieson stain (arrows) (**Figure 2A& 2B**). The proximal CBD margin was also involved by the tumor cells. Immunohistochemistry (IHC) showed positivity for