Duodenal GIST Presenting as Large Pancreatic Head Mass: An Uncommon Presentation

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ABSTRACT

Context Duodenal gastrointestinal stromal tumors (GIST) are uncommon. They usually present with gastrointestinal bleed, upper abdominal pain or mass abdomen. Tumor arising from the second part of duodenum can be wrongly diagnosed as pancreatic mass.

Case report We present a case of a thirty-year-old male who came with chief complaint of mass upper abdomen. On laparotomy there was a 15x10 cm mass arising from the whole of anterior surface head of pancreas and was attached with the second part of duodenum for about 1-2 cm only. Patient underwent pancreaticoduodenectomy and histopathology revealed that tumor was arising from the duodenal wall. Conclusion Duodenal gastrointestinal stromal tumors can grow exophytically into large mass and involve the pancreas without infiltrating microscopically and present as pancreatic head mass.

INTRODUCTION

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract with annual incidence of 10-20 per million [1]. Common sites are stomach (60%), small intestine (30%), rectum (5%) and esophagus (<5%). Duodenal GISTs constitute 30% of primary duodenal tumors and less than 5% of gastrointestinal stromal tumors [2, 3]. These tumors mostly occur in second part of duodenum followed by third, fourth and the first part [4].

CASE REPORT

A thirty-year-old man presented with the chief complaint of mass in the right upper abdomen since one and half year. It was gradually increasing in size and was associated with occasional upper abdominal pain. Pain was dull in nature, non-radiating with no specific aggravating and relieving factor. There was history of incomplete bowel evacuation sensation and increased frequency of defecation. There was no history of vomiting, gastrointestinal bleed, jaundice, anorexia and weight loss. On physical examination, there was no pallor, jaundice and lymphadenopathy. A large firm mass about 15x10 cm extending into epigastrum, umbilical and right hypochondrium on per abdomen examination. It had round shape, bosselated surface, smooth margins, and was mobile in the transverse direction. There was no abnormality on digital rectal examination and proctoscopy. Routine laboratory tests were within normal limits. Ultrasound abdomen showed 15x10 cm heterogeneous mass in the umbilical region displacing the adjoining gut loops with no invasion. CECT abdomen showed 15x10 cm size, well defined mass with heterogeneous density in the retroperitoneum extending from pancreas to pelvic brim. It had enhancing peripheral component and non-enhancing (necrotic) central component (Figure 1). Fine needle aspiration cytology of mass smear showed blood only. On exploratory laparotomy there was large hyper vascular mass protruding through the transverse colon mesentery. Mass appeared to be originating from the anterior surface of head of pancreas. It was attached to the whole length of anterior surface of pancreas and macroscopically was not attached with the duodenum except for about one or two cm near the lower end of second part of duodenum (Figure 2). There was no metastasis in liver or peritoneum. Pancreaticoduodenectomy was done. Histopathology showed spindle cell tumor with palisading pattern and foci of necrosis (Figure 3). The mitotic count was up to 15/50 HPF. Tumor was involving duodenal muscularis propria with no infiltration in the duodenal epithelial layer and the pancreas (Figure 4). Immunohistochemical study revealed positive staining for CD117, CD34, vimentin, smooth muscle actin, and negative staining for desmin and CD31 (Figure 5). Based on these findings, the tumor was finally diagnosed as gastrointestinal stromal tumor (GIST) arising from the duodenal wall, growing exophytically and attached with the pancreas without infiltrating the pancreas.
Post-operatively patient had biliary leak which was managed conservatively and discharged in satisfactory condition with the advice to take imatinib 400 mg daily.

**DISCUSSION**

Gastrointestinal stromal tumor (GIST) arises from the interstitial cells of Cajal which are located in the submucosal and myentric plexus of gastrointestinal tract. The main mechanism in the pathogenesis of most GISTs is the mutation in one of two receptor tyrosine kinase genes (KIT and PDGFRA). On immunohistochemical staining, 95% are CD117 positive, 70% are CD34, and 40% stains positive for smooth muscle actin [5, 6]. They are typically negative for desmin and S-100 (<5% positive) [5]. There are three main histological cell types of GIST: spindle cell type (most common), epithelioid cell type, and the mixed spindle-epithelioid type [5]. In our case tumor was of spindle type and stained positive for CD117, CD34 and negative for desmin. The mean age of patients with GIST is 53 years and only about the 5% of GIST patients are younger than 30 years [4]. In our case age of patient was 30 years, which is unusual for GIST. Duodenal GIST majority of times presents with gastrointestinal bleeding, epigastric pain, palpable mass and intestinal obstruction [4, 7]. Microscopic mucosal

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**Figure 1.** Contrast enhanced computed tomography showing: a. heterogeneous mass arising from pancreas; b. A 15x10 cm heterogeneous mass with peripheral enhancement and central necrosis.

**Figure 2.** Macroscopic appearance of resected specimens showing tumor attached mainly along the pancreatic head and with duodenum only near the lower end of second part of duodenum.

**Figure 3.** Histopathology showing spindle shaped tumor arranged in palisading pattern (H&E stain, 200x).

**Figure 4.** Histopathology showing tumor involving the duodenal wall (H&E stain, 40x).
In a study of twenty-five cases of duodenal GISTs, preoperative diagnosis was pancreatic head tumor in five cases [13]. These authors noted that GIST located in second part of duodenum were less frequently correctly diagnosed in the pre-operative phase and suggested that in a large periampullary tumor with no jaundice, duodenal GIST should be ruled out.

Surgical resection with tumor free margin is the mainstay of treatment for the patients with primary GISTs without distant metastasis. Surgery can be wedge resection of duodenum with primary closure of duodenum, segmental duodenectomy with duodenal jejunostomy reconstruction or pancreaticoduodenectomy [3]. Limited resection should be done whenever feasible and pancreaticoduodenectomy should be considered for large tumors of first and second part of duodenum and tumors involving the papilla of Vater or the pancreas [3, 13, 14]. Pancreaticoduodenectomy was done in our case, considering it to be a pancreatic head tumor.

Various parameters are described to predict the malignant potential of GIST, such as tumor size, mitotic activity, tumor location, non-radical resection, tumor rupture, peritoneal dissemination, metastasis, and invasion into adjacent organs. National Institute of Health (NIH) consensus criteria (Fletcher’s criteria) proposed risk stratification of tumor behavior based upon its size and mitotic activity. Tumors larger than 10 cm in size with any mitotic count or of any size with mitotic count more than 10/50 HPF are at high risk of aggressive behavior [15, 16, 17, 18]. Adjuvant therapy with imatinib has been recommended in patients with substantial risk of relapse. Risk of relapse is increased in tumors of large size, increased mitotic activity and resection with positive margins. Adjuvant therapy with imatinib has been shown to increase the relapse-free survival but not overall survival [6, 16, 19]. In our case, adjuvant therapy with imatinib was started considering tumor to be high grade (size >10 cm and mitotic figures 15/50 HPF) and high recurrence rate in patients with duodenal GIST.

To conclude, duodenal GIST can mimic as a pancreatic head mass.

**Conflicts of interests** The authors have no potential conflict of interest.

**References**


