

analysis using tissue micro-dissection in pulmonary carcinosarcomas. They found identical allelic losses shared by each tumor component, without discordant losses. This was consistent with the hypothesis that the carcinomatous and sarcomatous components were derived from a single pluripotent stem cell or the divergent hypothesis.⁴ Alternatively, ultra-structural studies of carcinosarcoma have revealed the presence of tonofibrils and desmosomes not only in the epithelial component but also in the sarcomatous component, suggesting derivation of the sarcomatous component from the carcinomatous component favouring the metaplastic or conversion theory.⁵

Zhang et al, in their study, reported that patients with tumours that were smaller than 5 cm had a longer survival and that the tumour size should therefore be considered as a major component in the future staging system for CSG, although genetic variations, early detection (stage I or stage II) and a more extensive surgery could also contribute to a better prognosis. They also indicated that the presence of gallstones, epithelial and mesenchymal component types, age and sex were of little prognostic value.⁶ Race (Japanese versus non Japanese) and tumour size were important prognostic factors in carcinosarcoma of gallbladder.⁶

JYOTSNA SINGH¹
SUNAYNA MISRA¹
CHHAGAN BIHARI¹
VINIYENDRA PAMECHA²
ARCHANA RASTOGI¹

¹Department of Pathology, ²Department of HPB Surgery, Institute of Liver & Biliary Sciences, Delhi, India.

Correspondence: Archana Rastogi
Email: drarchanarastogi@gmail.com

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Segmental duodenectomy for gastrointestinal stromal tumor of the duodenum

Gastrointestinal stromal tumours (GISTs) are the most common mesenchymal tumours of the digestive tract, but constitute only 0.1% to 3% of all gastrointestinal tumours.¹ Duodenal GISTs are less frequent and account for 3 to 5% of cases. However they represent approximately 30% of all primary duodenal tumours.² Surgical resection remains the main stay of treatment but the optimal surgical procedure for duodenal GIST remains to be established.³ Case reports and small series describe different surgical procedures such as pancreaticoduodenectomy, pancreas-preserving total, distal or segmental duodenectomy and local wedge resection.⁴ Segmental and local resections in the duodenum are challenging due to the direct proximity to the pancreatic head, the papilla of Vater and the mesenteric root. We report a rare case of GIST located in medial wall of third part of duodenum treated with a challenging pancreas preserving distal duodenectomy procedure.

Case Report

A 51-year-old man presented to our hospital with recurrent episodes of syncope and melenain a span of one and a half years. Investigations showed that his haemoglobin was 5 gm%. Upper gastrointestinal endoscopy,colonoscopy, CT Angiography and RBC nuclear scan could not identify any source of bleeding. In view of the persistent melena, side viewing endoscopy was done. This revealed a deeply ulcerated submucosal lesion of size 1.5x1.5 cm located approximately 3 cm distal to the ampulla on medial wall of duodenum.

Biopsy of the lesion was inconclusive. CECT abdomen revealed a 2x2 cm enhancing lesion just distal to ampulla with no regional lymphadenopathy, possibly a GIST (**Figure 1**). Operative finding revealed a 2x2 cm nodular lesion on the medial wall of third part of duodenum approximately 2 cm beyond the ampulla (**Figure 2**). With careful dissection distal to the ampulla, the duodenum along with the lesion could be taken out from pancreas. The jejunum just beyond DJ flexure was divided and the specimen of pancreas sparing distal duodenectomy was taken out. Cut open specimen revealed a 2.5x1.5 cm submucosal growth with ulceration (**Figure 3**). Reconstruction was done by side to side duodenojejunostomy. Microscopic examination showed a submucosal mesenchymal tumour reaching up to muscularis propria with free cut margins. Mitotic activity

was less than 5/50 high power fields. With IHC showing positivity for CD-117, DOG-1 & CD-34,a diagnosis of GIST was made. The patient is on regular follow up and is doing well.

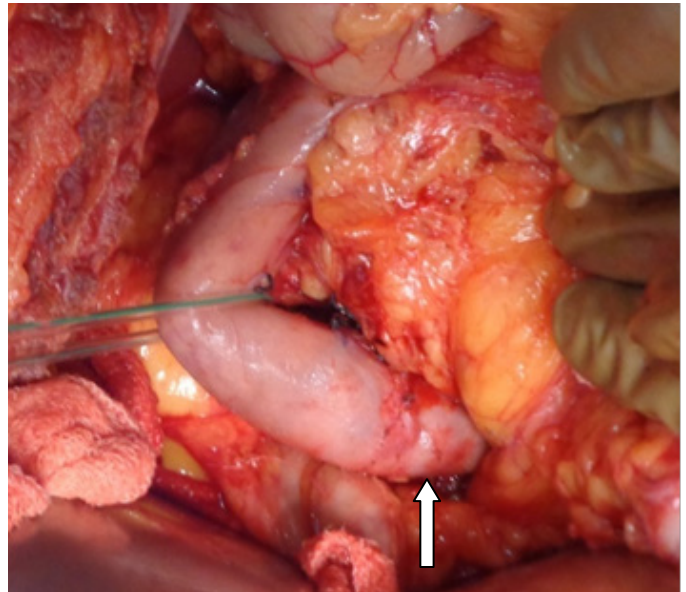


Figure 2: 2x2 cm nodular lesion on the medial wall of third part of duodenum.

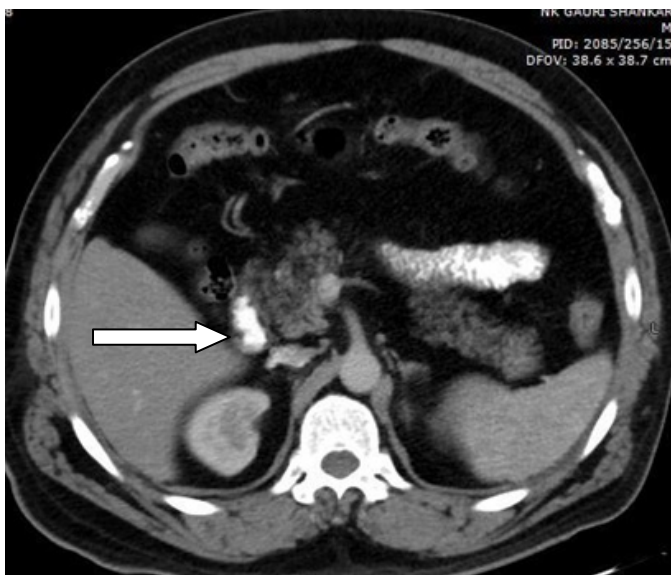


Figure 1: 2x2 cm enhancing lesion just distal to ampulla.

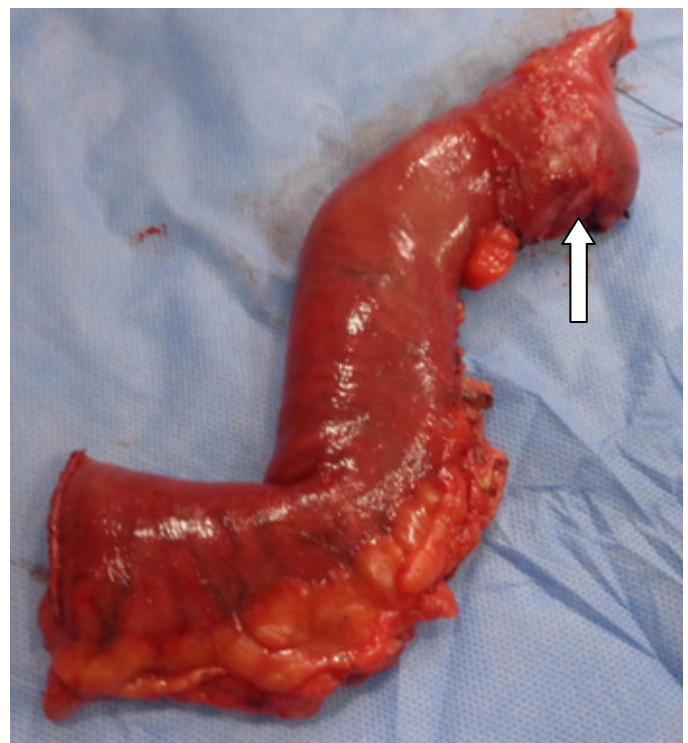


Figure 3: Specimen revealed a 2.5x1.5 cm submucosal growth with ulceration.

Discussion

Gastro-intestinal stromal tumours (GIST) are rare primary mesenchymal tumours arising from the Cajal cells of the gastrointestinal tract. Since longitudinal submucosal spread is very limited and lymph node involvement is rare, margin-negative resection without lymphadenectomy is the commonly accepted surgical treatment. Due to the rarity of duodenal GISTs, there are no large studies to define the optimal surgical procedure. Literature reports a 2% mortality after pancreas sparing total duodenectomy but a high morbidity of around 60% with most of the complications being secondary to anastomotic leaks.⁵ Pancreas sparing distal duodenectomy eliminates the risk of biliary and pancreatic anastomotic leak and thus is a good alternative for infra-ampullary tumours. Since the tumour does not invade the pancreas the procedure is oncologically sound. Our patient was an ideal candidate for this procedure. Positivity for CD117 (protein tyrosine kinase Kit) is the gold standard to establish a diagnosis of GIST. However, around 10% of cases have clinicopathologic features of GISTs but do not express Kit. Recently published reports mention higher sensitivity for DOG-1 than CD-117 for diagnosis of GIST. Our patient's lesion was positive for both CD-117 and DOG-1.

V TREHAN
PANKAJ P RAO
SANJAY SHARMA
A K SINGH
A GAUR
S V KULKARNI
N PATHAK

*Department of G I surgery & Liver Transplant, Army Hospital
(R & R), New Delhi-110010.*

*Correspondence: V Trehan
Email: vtrehan@rediffmail.com*

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