Duodenal Gastrointestinal Stromal Tumour Imitating as Pancreatic Head Tumour: A Typical Case of Whodunit

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Abstract

Duodenal gastrointestinal stromal tumours (D-GISTs) are rare disease. It may arise commonly from second or third part of the duodenum and can be erroneously diagnosed as a pancreatic head tumour due to proximity and morphology on imaging studies.

We present a case of a sixty-year-old woman who presented with abdominal pain and was diagnosed as a case of pancreatic neuroendocrine tumour on radiologic imaging and granulomatous lesion on aspiration cytology. On laparotomy, a ~5x3 cm mass was noted in the pancreatic head and pancreatoduodenectomy was performed. Histopathology reported an exophytic GIST arising from the second part of the duodenum.

Hence, D-GIST can invade into the pancreas and mimic pancreatic head tumour; therefore, these tumours should be kept in the differential diagnosis of an atypical pancreatic head mass.

Introduction

Gastrointestinal stromal tumours (GISTs) are uncommon mesenchymal tumours of the gastrointestinal tract. It may arise from the duodenum, but the incidence is less than 5% among all GISTs [1]. Such stromal tumours usually have an extraluminal extension and exophytic growth from the medial wall of the second part of the duodenum and may grow into the head of the pancreas and mimic a pancreatic tumour. Endoscopic ultrasound (EUS) assessment with fine-needle aspiration cytology (FNAC) has been reported to be useful to establish the diagnosis, but has low sensitivity and specificity [2]. Hence, it may mislead the pre-operative diagnosis frequently and an upfront surgical intervention may be the only way out.

We present a classic case of whodunit where the patient was diagnosed as having pancreatic pathology on both imaging and EUS-FNA, but came out as duodenal GIST at the end, along with a review of literature of similar such cases.

Case Presentation

We encountered a 60-year-old lady who presented with pain in the upper abdomen for the last 4 months which was dull aching and gradually progressive. She also noticed anorexia and undocumented weight loss over the past month. However, there was no history of vomiting, hematemesis, jaundice, awareness of lump, or altered bowel habits. Abdominal examination and biochemical investigations were within normal limits.

She was evaluated with an ultrasound abdomen which showed dilated common bile duct (CBD) with intrahepatic biliary radical dilatation (IHBRD) due to a mass in the pancreatic head region. Contrast enhanced computed tomography (CECT) of the abdomen showed a well-defined mass lesion (~5x4.2cm) in the pancreatic head. Mass showed peripheral enhancement in the pancreatic phase, progressively increasing in the venous phase with few areas of central necrosis and foci of peripheral calcification. The lesion was causing mass effect on the distal CBD with upstream dilatation and a provisional diagnosis of pancreatic neuroendocrine tumour (PNET) was evoked (figure 1a-c). Subsequently, contrast enhanced magnetic resonance imaging (CE-MRI) was done which corroborated the findings of PNET as on CECT.

EUS revealed a heterogenous mass in the head of pancreas with increased vascularity and multiple specks of calcifications. EUS guided aspiration cytology from the lesion showed singly scattered and clusters of spindle shaped cells resembling non-necrotizing epithelioid cell granulomas, which adds the diagnostic dilemma (figure 2a-b). A provisional diagnosis of PNET with differential diagnosis of pancreatic tuberculosis, sarcoidosis, or perivascular epithelioid cell tumour (PEComa) was kept and the patient was planned for surgery.

Intra-operatively, a hard cystic mass was found in the head of the pancreas and pylorus resecting pancreatoduodenectomy was performed with pancreatico-jejunostomomy by modified Heidelberg's technique (Figure 3a-b). Her post-operative course was smooth and allowed orally on the 2^{nd} post-operative day (POD) and subsequently discharged on the 7^{th} POD.

Cut section of the gross specimen revealed a firm growth involving the 2^{nd} part of duodenum close to the ampulla of Vater and occupying the head of the pancreas with solid grey white areas (figure 3c-d). Histopathology showed a circumscribed spindle cell tumour arising from the submucosa of the duodenum. Nests of tumour cells were separated by fibro-collagenous septa which was infiltrated by lymphocytes and plasma cells admixed with neutrophils and eosinophils. Along with it, areas of necrosis and giant cell reaction were also noted. The tumour was abutting the pancreas and CBD however; no definite invasion was noted in the pancreatic parenchyma (figure 4a-c). All surgical resection margins were free of the tumour with no lymphovascular or perineural invasion.

On Immunohistochemistry, spindle cells revealed strong immuno-reactivity for 'discovered on GIST-1' (DOG-1) marker, faint reactivity for (cluster of differentiation) CD34 and were non-reactive for S-100P with Ki-67 index of 1% (figure 4d). Hence, the final diagnosis was Duodenal-Gastrointestinal stromal tumour (D-GIST), spindle cell type with a mitotic rate of 2/50 high power field. In view of large size of the tumour (>5cm), adjuvant imatinib therapy (400mg OD) was started and she is doing well at 6 months of follow-up.

Table 1: Various cases of Duodenal GIST masqueraded as pancreatic head mass

No.	Authors	Age / Gender	Symptoms	Pre-operative CT findings	Pre-operative diagnosis	Surgery performed
1	Futo Y et al [5]	79/M	Tarry stool with transient loss of consciousness	Well-defined ~5cm tumor in the pancreatic uncus which showed enhancement from arterial to venous phase	Pancreatic NET	Pylorus-preserving PD
2	Dhakal D et al [6]	58/F	Post-prandial vomiting, loss of weight and appetite	Homogeneous soft tissue tumor mass in the uncinate process of the pancreas	Pancreatic mass	Wedge resection with cholecystectomy
3	Vasile D et al [7]	59/F	Abdominal pain, melena and faintness	Tumor mass ~6cm in the head of the pancreas which was hypodense, hypocaptive with ill-defined borders	Duodenal GIST	Partial resection of DII and total resection of DI, antrum, distal CBD, and enucleation of the mass from the pancreatic head with RYHJ and GJ
4	Bormann F et al [8]	64/F	Pain in right upper abdomen	Heterogeneous mass of 5×3 cm in the pancreas with both enhancing and non-enhancing areas	Pancreatic mass	PD
5	Slavik T et al [9]	49/F	Chronic abdominal pain	~2.8cm well-defined and circumscribed pancreas head mass lesion	Non-functioning Pancreas NET	PD
6	Mouaqit O et al [10]	37/F	Recurrent pain in right upper abdomen associated with sweating and nausea	Tumor of 7×5 cm with an area of central necrosis and intense contrast enhancement (arterial phase) in the pancreas head	NET or an adenocarcinoma of the pancreas	PD
7	Singh S et al [11]	30/M	Awareness of lump and pain in the right upper abdomen	Well defined mass ~15x10cm with heterogeneous	Pancreatic mass	PD

				density extending from pancreas to pelvic brim; Enhancing peripheral component and non-enhancing (necrotic) central component		
8	Frampton AE et al [12]	37/F	Recurrent right upper abdomen pain associated with sweating and nausea	~2cm hypervascular lesion lying between the head of pancreas and the second part of the duodenum	NET or a solid pseudopapillary tumor of the pancreas	Pancreatic head resection with duodenal segmentectomy
9	Ohtake S et al [13]	60/F	Asymptomatic; Discovered on screening ultrasound	A hypervascular tumor ~14 mm in the pancreas head	Pancreas NET	Tumor resection with a duodenal wedge resection
10	Kwon SH et al [14]	49/M	Incidentally detected in follow up	Well-defined tumor of ~2.3cm in the uncinate process which showed strong enhancement	Non-functioning islet cell tumor	Segmental resection of the duodenum and duodeno-jejunostomy
11	Uchida H et al [15]	53/F	Asymptomatic	Well-defined tumor in the pancreatic head ~ 3.0cm in size; enhancement in the arterial phase which lasted until the delayed phase	Non-functioning islet cell tumor	Pylorus preserving PD

PD- Pancreatoduodenectomy; NET- Neuroendocrine tumor; GIST- Gastrointestinal stromal tumor

Discussion

GISTs are common mesenchymal tumours and are encountered predominantly in the stomach (60%–70%), small intestine (25%–35%), and colo-rectum (10%) [3]. Only 1-5% of GISTs occur in the duodenum with the most common location being its second part [4]. Duodenal GIST with purely medial extra-luminal extension, especially from the second or third portion of the duodenum, may be difficult to diagnose pre-operatively. It may mimic non-functioning P-NET, islet cell tumour, retroperitoneal mass and PEComa which is reported in

the literature, akin to our case. On reviewing the literature, we could find only 11 such cases where D-GIST masquerades as pancreatic head mass (Table 1) and the surgeon must be aware of such a unique conundrum [5–15].

The common symptomatology of all such differentials makes a pre-operative diagnosis difficult. The common presentation of D-GIST is recurrent pain in the epigastrium followed by a history of gastrointestinal bleeding due to the highly vascular duodenal submucosa. The other presentation may be palpable mass, features of gastric outlet obstruction, or infrequently diagnosed on routine screening. On the other hand, P-NETs are infrequent with an incidence of 1–2% among all pancreatic neoplasms and more than one-third of cases are non-functional. Hence, the presentation of the disease usually doesn't provide a convincing clue towards etiology. The mean tumour size of D-GIST reported across literature was 6 cm (range 1.5–31 cm) similar to the presented case [4,16].

P-NETs, as well as D-GISTs show intense arterial enhancement on CT scan, hence may be challenging to diagnose based only on imaging. In 10-20% of cases, D-GISTs have been misdiagnosed as pancreatic head mass [16,17]. Therefore, it is suggested that in patients with large pancreatic head mass without jaundice, duodenal GIST should be kept as a differential.

Usually, the EUS is helpful in confirming the origin of D-GIST which is either from 2^{nd} (muscularis mucosae) or 4^{th} (muscularis propria) layer of the duodenum. However, in larger and predominantly extraluminal tumours, this may not be possible due to its compression effect, as happened in our case. Further, EUS-FNA has been reported to be effective in diagnosing such lesions with lower risk of complications. However, in context to differentiate D-GIST from pancreatic tumour, there are concerns about the post-procedure bleeding as these lesions are vascular. Further, the sensitivity of EUS-FNA cytology is better for the gastric GISTs as compared to the duodenal origin, as the reported yield is suboptimal in nearly $2/3^{rd}$ of cases [2]. In our case, pancreatic tuberculosis/sarcoidosis were kept as differentials as tumour stromal cells were misinterpreted as epithelioid granulomas. The limitation in our case was that we could not perform EUS guided biopsy.

The optimal surgical procedure for D- GISTs, even with a pre-operative diagnosis, is not distinct due to the complex anatomy of the pancreaticoduodenal region. The wide local excision or segmental duodenectomy has been reported to be oncologically and technically safe, but Pancreatoduodenectomy (PD) is warranted in cases where tumour size is large, involving second or third part of the duodenum, close to the ampulla of Vater or other vital structures and in cases with a diagnostic dilemma as in our case. It is reported that more than $2/3^{rd}$ cases of duodenal GISTs underwent PD due to diagnostic uncertainty and the large size of the tumour [5,11,16].

GIST of the small intestine is more aggressive and has more chances of recurrence than its gastric counterpart. Gold and colleagues have developed a nomogram based on tumour site, size, mitotic rate to assess recurrence free survival and selection of a patient for adjuvant therapy [18]. In context to our case with tumour size >5cm and mitotic rate $\leq$5 per 50 high power field, the risk of metastasis is noted in nearly one-fourth of the patient [3]. Hence, imatinib therapy for 3 years is beneficial to prevent future recurrence.

In conclusion, GIST located in the second part of the duodenum is seldom diagnosed correctly in the pre-operative phase especially with a lesion in the pancreatic head or mesenteric side of the duodenum. The attempt to establish preoperative diagnosis should be done with EUS guided biopsy, whenever feasible. Pancreatoduodenectomy is a safe and viable option with low morbidity for such lesions.

Declarations

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Ethics approval: This is a case report and Institute Ethics Committee has confirmed that no ethical approval is required.

Consent to participate: Verbal and written consent was obtained from the patient to be included in the study.

Consent for publication: The participant has consented to the submission of the case report to the journal.

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Figures

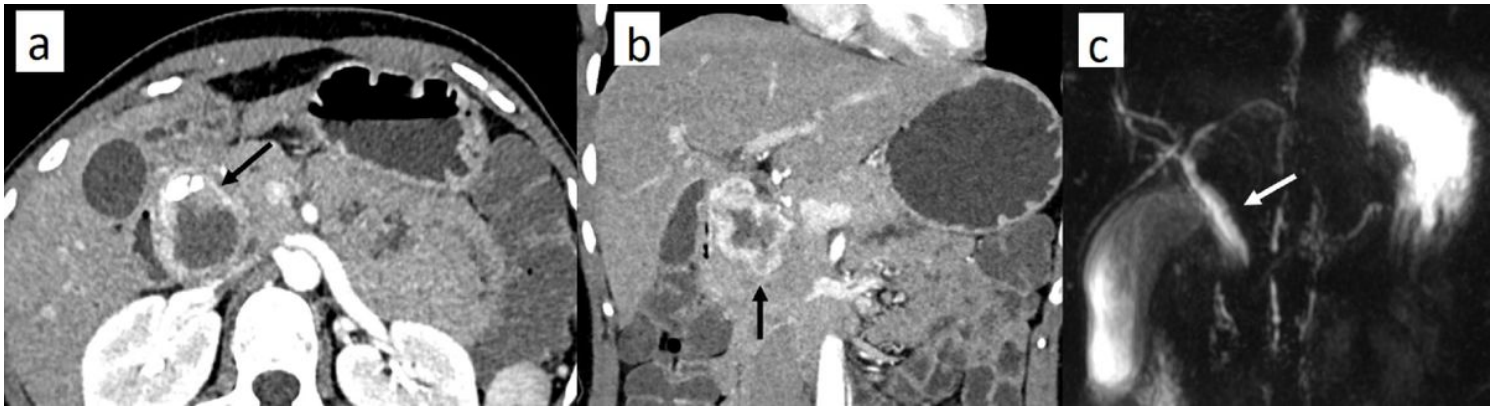

Figure 1

Radiology images: (a) Axial, (b) Coronal images of Contrast enhanced CT in pancreatic phase show a heterogeneously enhancing mass (black arrows) with foci of calcifications and central non-enhancing areas. (c) 3D MRCP image shows dilated common bile duct (white arrow). Pancreatic duct is normal in caliber.

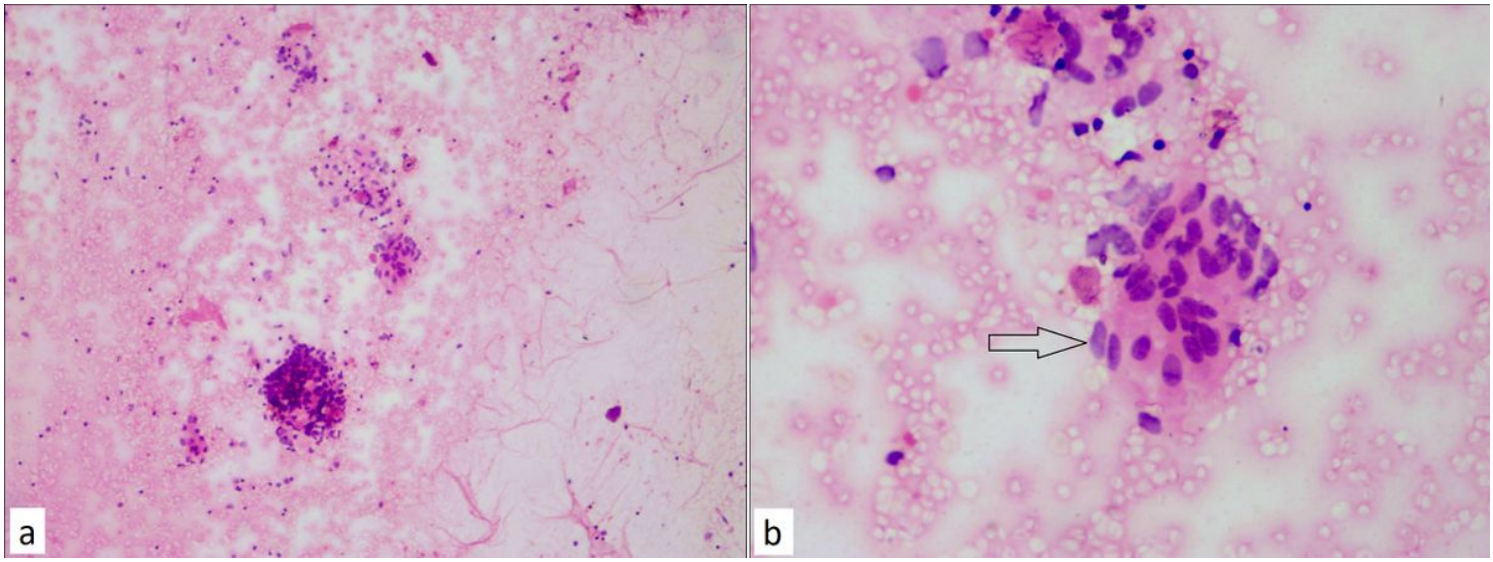

Figure 2

Fine needle aspiration cytology: a) Hematoxylin and Eosin (H&E), 10X and b) H&E 40X showing spindle shaped cells with epithelioid morphology (arrow)

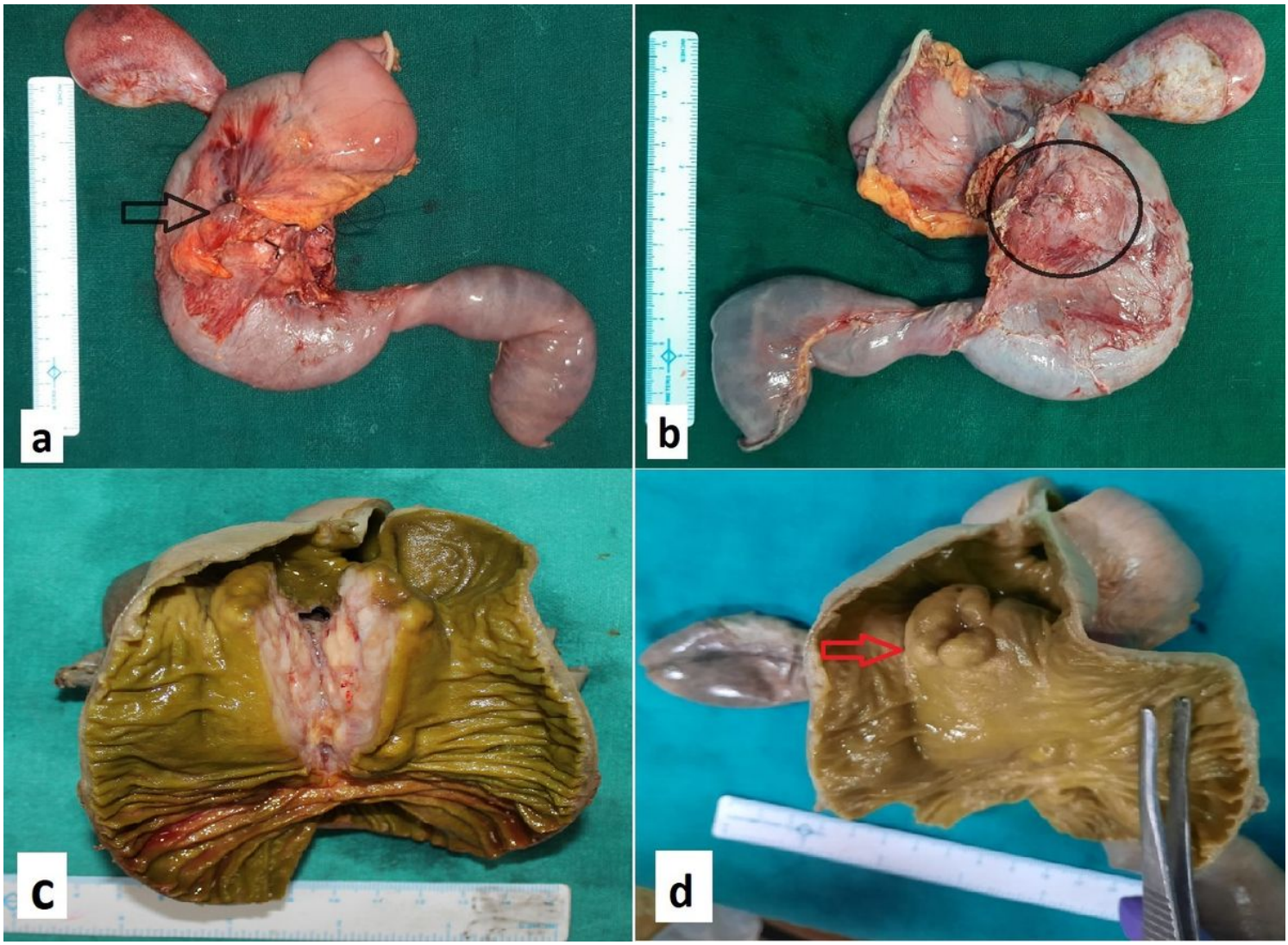

Figure 3

Pancreatoduodenectomy specimen images: a) Anterior and b) Posterior view of resected pancreatoduodenectomy specimen where tumor site marked with arrow and circle; c) Cut section of the specimen showing tumor involving head of the pancreas with solid grey white areas; d) Bulky and projecting ampulla in to the second part of duodenum due to the tumor.

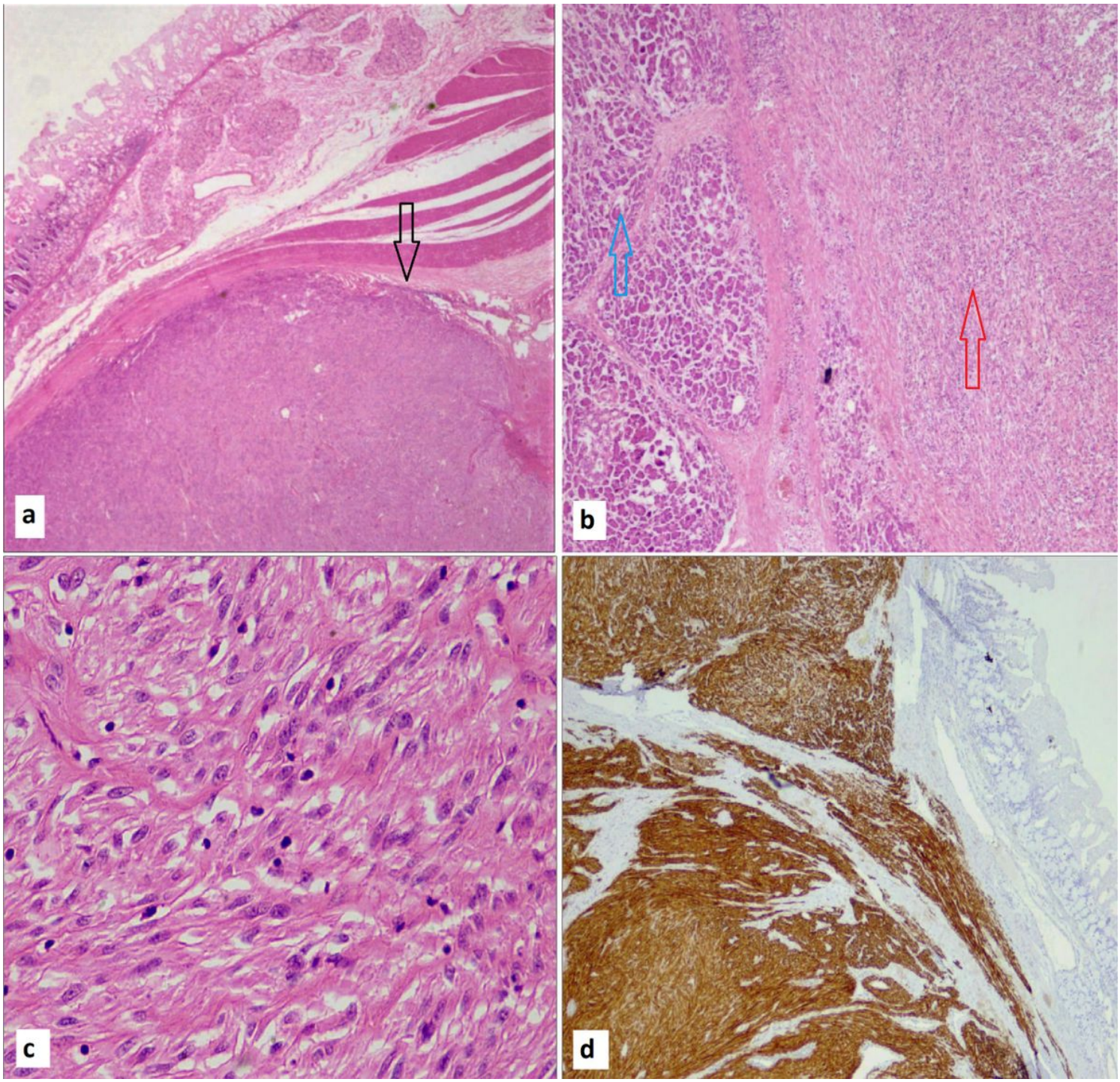

Figure 4

Histopathology: a) Hematoxylin and Eosin (H&E), 4X shows a tumor in the duodenal submucosal area (arrow); b) H&E, 4X shows pancreatic acini (indicated by blue arrow) & tumour (red arrow); c) H&E, 40X spindle cell tumor arranged in fascicles; d) Tumor cells showing diffuse immunoreactivity for DOG-1