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Case Report

# Leiomyoma at the Ampulla of Vater: A Case Report and Literature Review

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## ABSTRACT

We describe a case in which transduodenal ampullectomy was performed for leiomyoma. A 19 years old female was found to have a tumor at the distal end of the common bile duct (CBD). Excision of the ampulla showed a well defined mass 2x1cm size and histopathological study revealed a benign smooth muscle tumor, leiomyoma.

**INTRODUCTION**

Stromal tumors of the GI tract are a large family constituting the majority of non epithelial neoplasm of gastrointestinal tract (GIT). They may also involve the omentum, mesentry, and retroperitoneum (1). Gastrointestinal stromal tumor rarely develops in the duodenal ampullary region (2).

**Key word:** Ampula of Vater, leiomyoma.

## CASE REPORT

A nineteen years old housewife female was admitted to our hospital complaining of jaundice lasting more than six weeks associated with low grade fever without rigor, anorexia, and vomiting. On examination, the patient was jaundiced and had hepatomegaly, four fingers below costal margin, not tender, with soft abdomen. The patient was thoroughly investigated; liver function test showed high total serum bilrubin level, 180 mmol/L (N 5-17), the direct bilrubin serum level was 157 mmol/L while the indirect was 23 mmol/L. The serum SGOT, SGPT transaminases enzymes levels were 99U/L (N40) and 95U/L (N45) respectively. On the other hand serum alkaline phosphatase level was highly elevated, 750 U/L (N30-85). No other significant lab investigation test was noted.

Abdominal Ultrasound showed enlarged liver, normal texture, with no space occupying lesion, dilated intra and extra hepatic biliary duct, CBD was dilated (9.4mm) till distal end. Gallbladder was distended measure 129x31x41 mm, thin wall with multiple stones. Other organs were normal. No ascitis. OGD endoscopy was performed and there was pangastropathy with prominent ampulla. The patient was also submitted for ERCP which revealed a large prominent ampulla, dilated intra and exrtahepatic biliary systems with distal CBD tapering, Fig (1).

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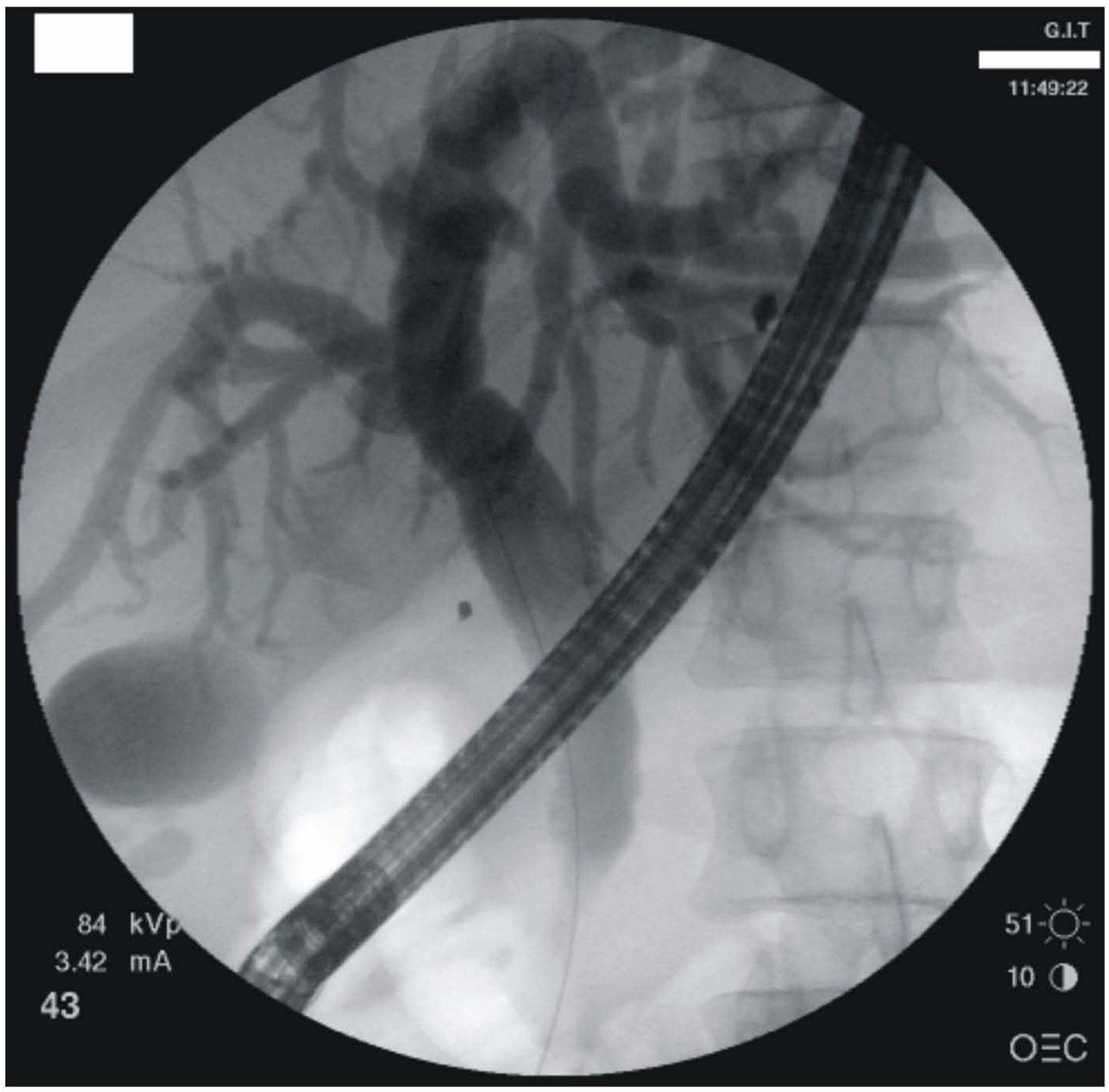



Fig (1): Choangiography of the patient showed dilation both extra and intrahepatic biliary tree with distal common bile duct narrowing due to ampullary tumor (leiomyoma).

Then scanning of the ampullary region by using endoscopic ultrasound (EUS) showed a hypoechoic homogenous mass like lesion 2x1.3cm well localized to the ampullary area without evidence of invasion of duodenal wall muscularis propria or invasion into the pancreas, the mass was completely occluding the CBD (1.4cm) but not the pancreatic duct (2.4mm), Fig2 A&B. Thus the provisional EUS diagnosis was an ampullary tumor, stage T1.

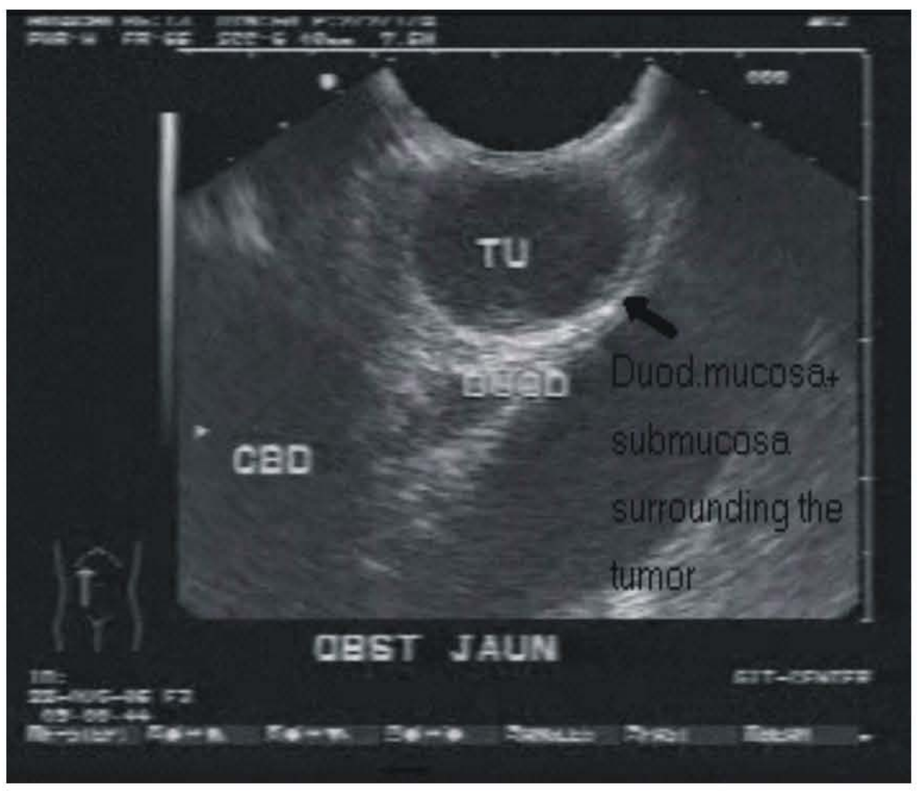



Fig 2: A, Both the mucosa (inner thin hypoechoic layer) and submucosa (outer thick hyperechoic layer) surrounding the lesion which gives a hint that the lesion arising from more deeper layer (muscularis properia) of duodenal wall.

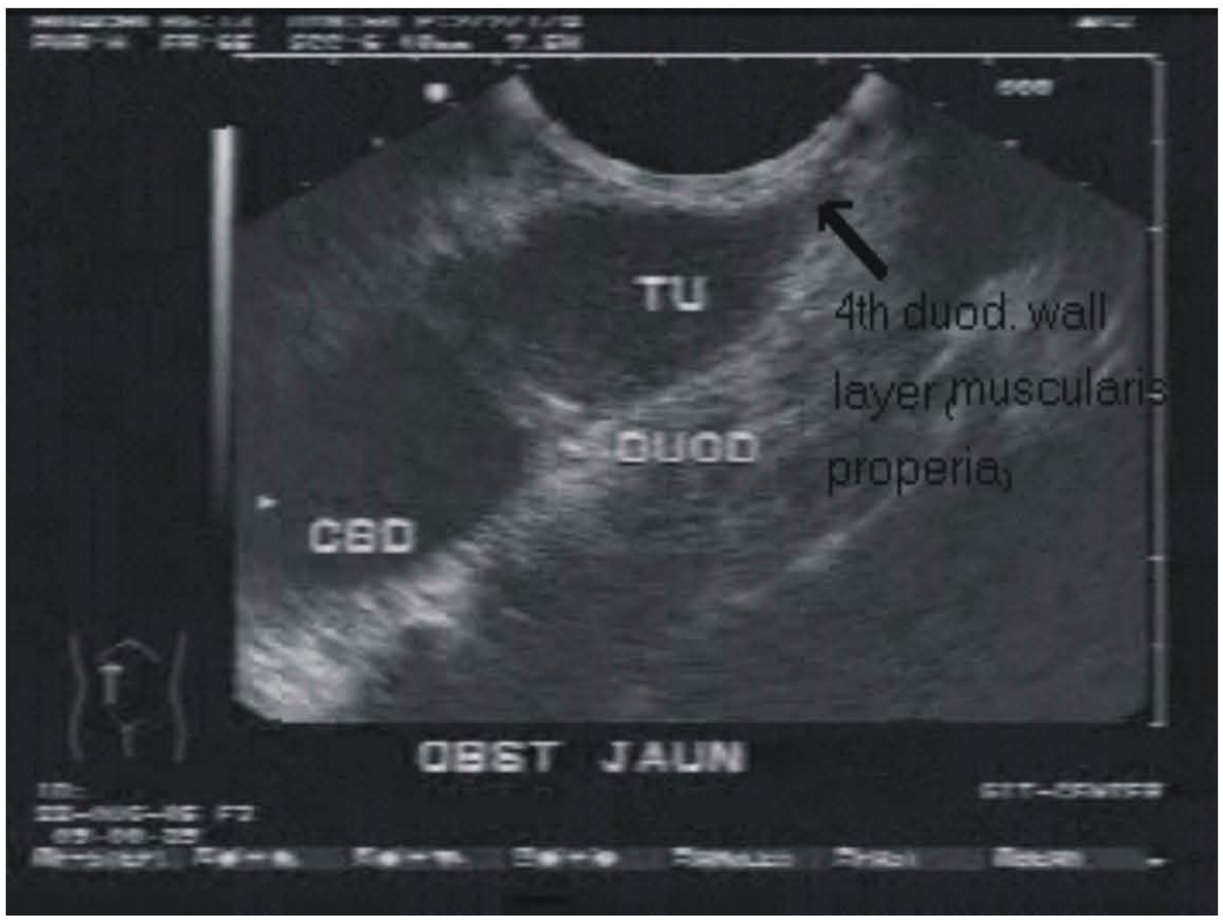



Fig 2: B, Hypoechoic lesion located at the region of ampulla of vater arising from the 4th layer of duodenal wall (musculars properia).

Transduodenal ampullectomy was performed and the Histopathological study showed well circumscribed tumor nodule with pushing borders composed of uniform spindle cells arranged in whorls and fascicles with eosinophilic cytoplasm and cigar shaped elongated nuclei. The mitotic figure was negligible and there was no tumor necrosis. (Fig3 A&B). Immunohistochemical markers were applied to the tissue sections, the tumor cells were positive for caldesmin (Fig4), and negative for c-kit (CD117), CD34, and desmin, indicating that the tumor is of smooth muscle origin rather than a gastrointestinal stromal tumor.
Considering the tumor size, histopathological features and the results of immunohistochemical stains, the resultant diagnosis was a benign leiomyoma.

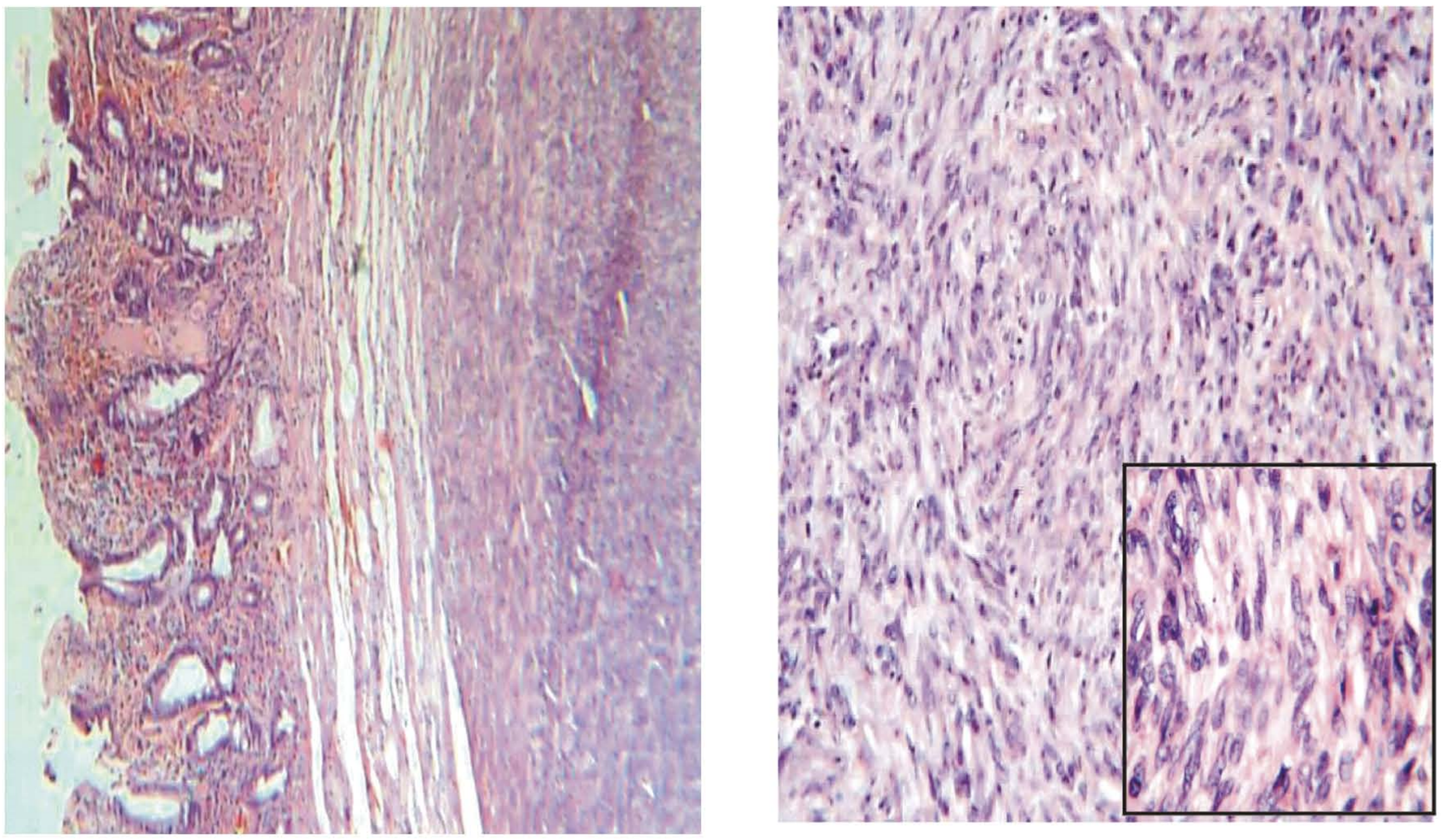


Fig 3: A, a well circumscribed tumor nodule lying below the ampullary surface epithelium. B, the tumor is composed of spindle cell proliferation arranged in whorls and fascicles. Inset, a high power view showing uniform cells with elongated nuclei.

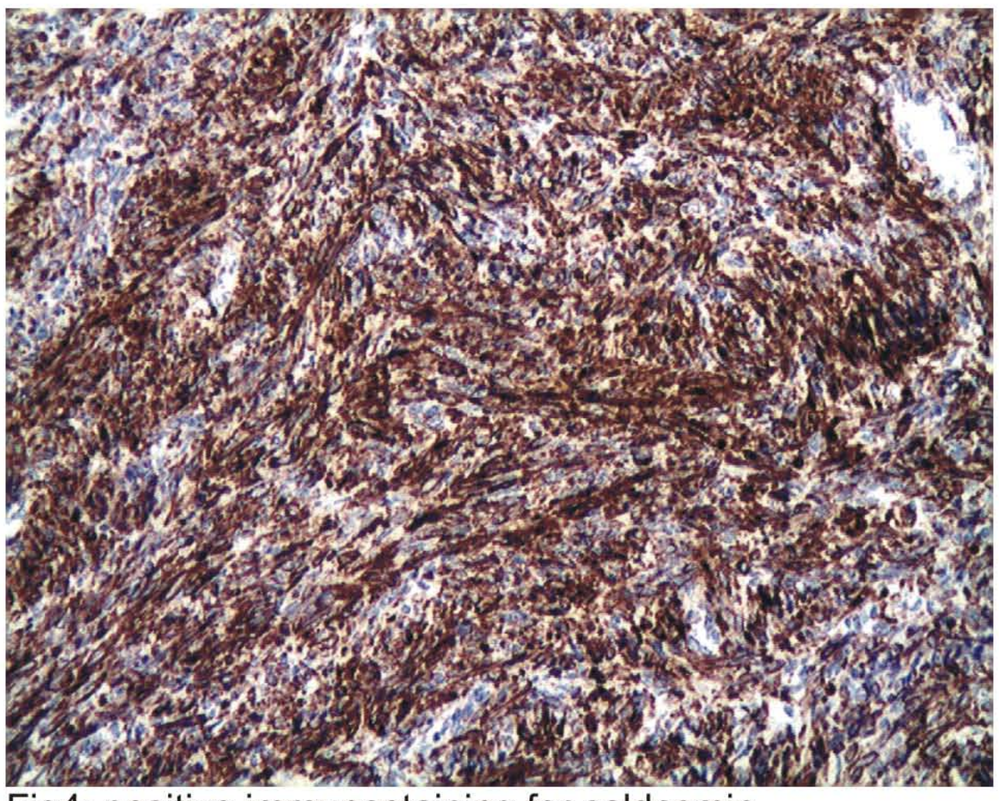
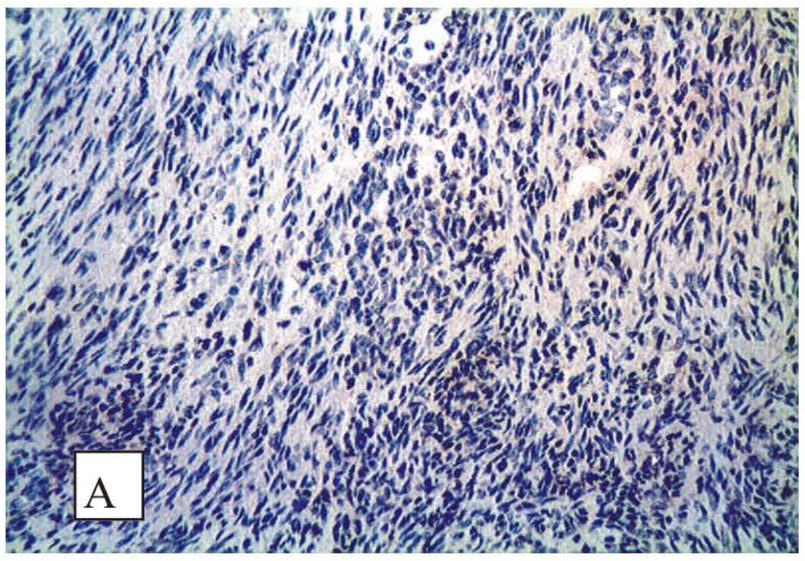
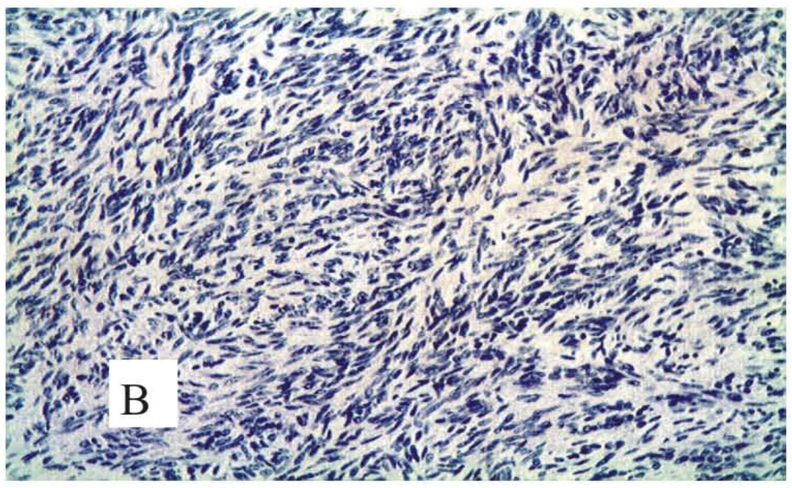


Fig4: positive immunostaining for caldesmin



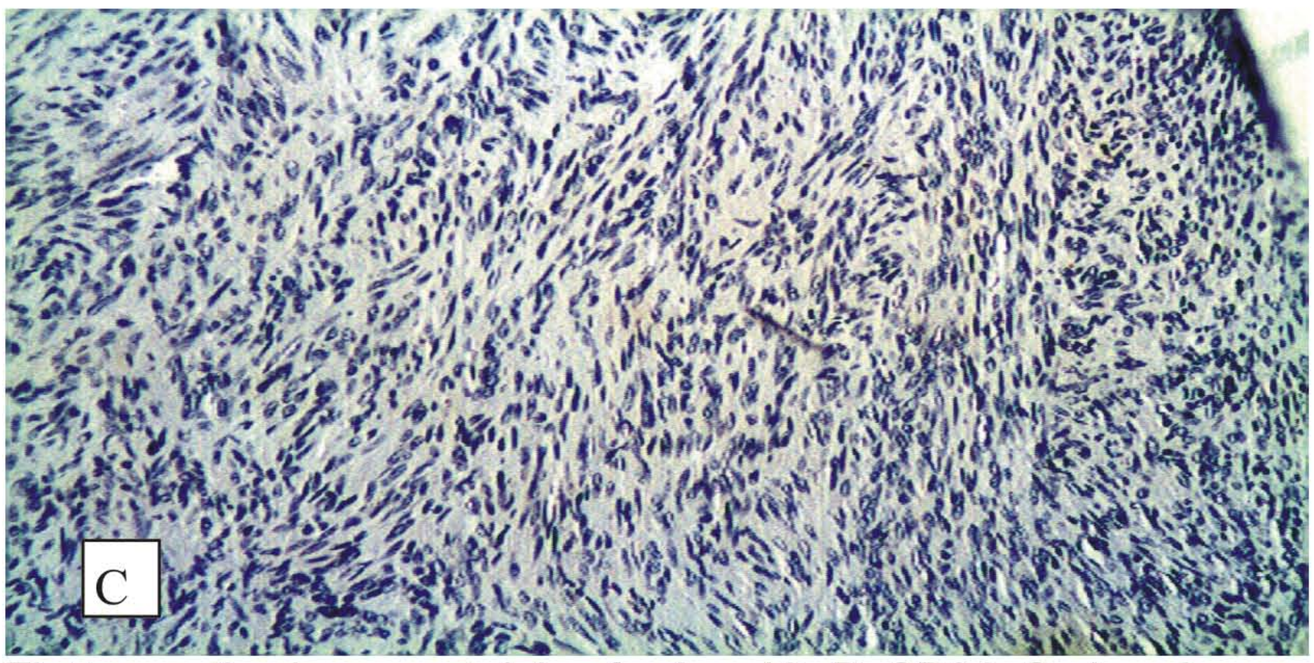




Fig5: negative immunostaining for A: c-kit, B: CD34, C: desmin

## DISCUSSION:

Endoscopic ultrasound (EUS) is the investigative procedure of choice when a submucosal lesion has been visualized endoscopically (3). EUS can accurately show the exact origin of a lesion, whether inside or outside the GIT wall. Inside the GI wall EUS can detect the layer of origin, for instance the 4th layer in leiomyoma or the 3rd layer in lipoma. High frequency ultrasound probe enables proper diagnosis of esophageal leiomyoma derived from muscularis propria (4). Unfortunately, EUS can not reliably show the difference between the benign and malignant submucosal tumors (5). However characteristics such as size, border, homogeneity and presence of necrosis can help to decide whether a lesion should be surgically removed or to be followed up by EUS. Nevertheless, the diagnosis on the basis of EUS is perceptive and can not replace the histopathological diagnosis of such lesions (6).

Stromal tumors of the GI tract show a remarkable variability in their differentiation pathway. This has resulted in a considerable degree of confusion in their interpretation, which if anything has exacerbated recently. For many years they were all regarded as being of basically of smooth muscle nature designated as leiomyoma and leiomyosarcoma when composed of spindle cells and as benign or malignant leiomyoblastoma (or epitheliod leiomyomas leiomyosarcomas, respectively) when composed of epithelioid cells. The many immunohistochemical and ultrastructural studies that have been carried out by numerous authors in recent times have shown a much more complex picture (7). Recently they have been classified as c-kit or CD34 positive mesenchymal tumors based on immunohistochemical and electron microscopic approaches (8). These tumors were designated as gastrointestinal stromal tumors (GISTs) that can be roughly divided into four major categories on the basis of their phenotypic features: 1) tumors showing differentiation toward
smooth muscle cells; 2) tumors with apparent differentiation toward neural element; 3) tumors showing dual differentiation toward smooth muscle and neural elements; 4) tumors lacking differentiation toward either cell type (8, 9). The differential diagnosis of GIST with leiomyoma/ leiomyosarcoma becomes cloudy because of the smooth muscle features that GIST can exhibit. A simple-minded but effective approach to the problem is to designate as leiomyoma/ leiomyosarcoma the tumors having typical morphologic and immunohistochemoical features of respective tumor type (as we know them from other locations) if they lack CD117 immunoreactivity. In our case depending on the morphological and immunohistochemical features, the diagnosis was an ampullary leiomyoma, benign.

After a thorough and comprehensive literatures review, we have found that this is the first time in which benign leiomyoma is described in the ampulla of Vater since the only three previous reported cases were ampullary malignant GISTs (2,10,11) .

Most GI tract stromal tumors are diagnosed histopathologically after resection because of submucosal location (10). They may grow expansively without being invasive and sometimes metastasize to the liver and recur locally (12, 13). Surgery with safe surgical margins and no tumor rupture is necessary and adequate means of treating such tumors. Excessive lymph node dissection is unnecessary, because they rarely metastasize to regional lymph nodes (7).

So despite of its rarity this neoplasm should be included in the differential diagnosis of the tumors appearing in the duodenal ampullary region.

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