

# Giant solitary Peutz-Jeghers-Type Hamartomatous Polyp in the Duodenum Presenting as Gastric Outlet Obstruction

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## Abstract

Solitary Peutz-Jeghers type hamartomatous polyp is a variant of PJS characterized by the presence of hamartomatous polyps in the absence of other manifestations of PJS. It is quite rare with only 19 cases reported in English literature. These cases present in older individuals are usually asymptomatic and carry increased risk of malignant transformation. Hamartomatous polyps are characterized histologically by tree-like branching of smooth muscle fibres covered by mucosal tissue of near normal appearance. Endoscopically, they are characterized by a lobular or nodular surface and whitish colour. We are presenting a case of giant Solitary Peutz-Jeghers type hamartomatous polyp of the duodenum who presented with symptoms of gastric outlet obstruction that was treated with pylorus preserving pancreaticoduodenectomy.

**Keywords:** Peutz-jeghers; Hamartomatous polyp; Giant polyp; Gastric outlet obstruction

## Introduction

Peutz-Jeghers syndrome (PJS), first described by Peutz in 1921 is a rare, autosomal dominant disorder characterized by multiple gastrointestinal hamartomatous polyps and muco cutaneous pigmentation around the lips and oral cavity [1-3]. A germline mutation of the STK11 gene located on chromosome 19p13.3 is responsible for development of PJS [4]. A solitary Peutz-Jeghers type hamartomatous polyp is a variant of PJS characterized by the presence of hamartomatous polyps in the absence of other manifestations of PJS [5]. These patients present with nonspecific symptoms with most diagnosed incidentally during endoscopy. We are reporting a case of giant solitary Peutz-Jeghers-type hamartomatous polyp in the duodenum in a young girl presenting as gastric outlet obstruction.

## Case Report

An 18 year old girl presented with history of recurrent non bilious vomiting of 4 month duration associated with vague upper abdominal pain. On abdominal examination, she had a mass in right hypochondrium with features suggestive of mass arising from the pancreas. She didn't have any peri oral muco cutaneous pigmentation. Oesophago gastro duodenoscopy (OGD scopy) revealed a polypoidal mass of about 10 × 6 cm arising from the medial wall of second part of duodenum close to ampulla of vater (Figure 1A). Biopsy from the mass was reported as villous adenoma with severe dysplasia. Contrast Enhanced Computed Tomography (CECT) of the abdomen revealed mass of size arising from the second part of duodenum with loss of fat planes with pancreatic head (Figure 1B). The mass was abutting superior mesenteric vein and right renal vein (Figure 1C). Patient underwent pylorus preserving pancreaticoduodenectomy (Figure 1D). Post operatively she developed post-operative pancreatic fistula (POPF) grade B which was managed conservatively. Histopathological examination of the specimen revealed solitary Peutz-Jeghers-type hamartomatous polyp arising from the 2<sup>nd</sup> part of duodenum involving the ampulla of vater (Figure 1E and F). There was mild dysplasia, however no evidence of invasive malignancy. All the resection margins were free and lymph nodes were reactive. After recovery she underwent colonoscopy which was within normal limits. On retrospective enquiry, patient's parents had 2<sup>nd</sup> degree consanguineous marriage though there

was no family history of any hereditary disorders. At 8 months follow up, patient is disease free and in good health. At last follow up, patient underwent colonoscopy to rule out any other polyps.

## Discussion

A solitary Peutz-Jeghers type hamartomatous polyp in the duodenum was first described in 1986 by Bott et al. [6]. This disorder differs from Peutz-Jeghers Syndrome (PJS) in following characteristics: Diagnosis at a more advanced age, absence of mutation of the STK11/LKB-1 gene and family history as well as lack of muco-cutaneous pigmentation [5]. However in many cases it is difficult to determine whether these polyps are a separate variant or initial clinical manifestation of PJS. Present patient had no significant family history except for 2 degree consanguineous marriage among her parents and no muco cutaneous pigmentation around the oral cavity.

Literature review by Suzuki et al revealed only 19 cases of solitary Peutz-Jeghers type hamartomatous polyp of the duodenum reported in English literature [7]. Age at diagnosis ranged from 23-89 years. Most common site affected was second portion of the duodenum followed by the duodenal bulb. Size of the polyp ranged from 5-70 mm. Present patient had a solitary hamartomatous polyp of size 80×30×30 mm [7]. The present case represents the largest polyp of this subtype reported in English literature. In addition, unlike previous reported cases she presented at the age of 18 years.

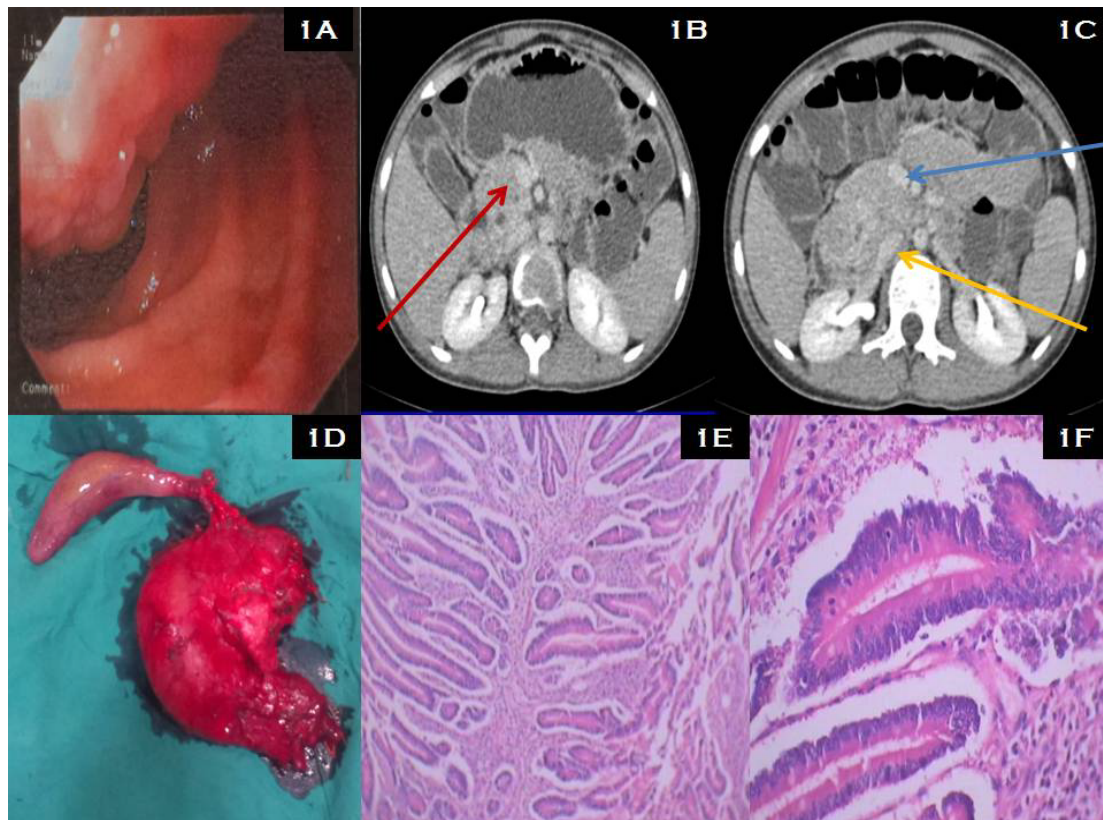
Although most of these patients are asymptomatic with polyp being detected incidentally some may present with features of cholangitis or bleeding [7-9]. Present patient presented with features suggestive

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**Figure 1A:** OGD scopy picture showing a polypoid mass arising from the medial wall of second part of duodenum. **1B.** CECT abdomen showing the duodenal polypoid mass abutting the head of the pancreas (Red arrow). **1C.** CECT abdomen showing the duodenal mass abutting the superior mesenteric vein (Blue arrow) and right renal vein (Yellow arrow). **1D.** Pancreaticoduodenectomy specimen. **1E and F.** Histomicrograph showing the hamartomatous polyp.

of gastric outlet obstruction probably owing to the size of the mass. Hamartomatous polyps are characterized histologically by tree-like branching of smooth muscle fibres covered by mucosal tissue of near normal appearance [10]. Endoscopically, they are characterized by a lobular or nodular surface and whitish colour [11]. Whitish colour is because of the presence of diffusely scattered white spots on the surface of the polyps [12]. However, as diagnosis by morphologic features is difficult, endoscopic biopsies must be performed to establish the diagnosis and hence guide treatment.

There is no consensus on the management of Peutz-Jeghers type hamartomatous polyp of the duodenum. In general, hamartomatous polyps are considered to have very low malignant potential compared to adenomatous polyps [13]. However, malignant transformation has been reported in 3%-6% of the polyps in Peutz-Jeghers syndrome [14,15]. Although, it is a general belief that solitary Peutz-Jeghers type hamartomatous polyp of the duodenum have low malignant potential, 4 of the 19 patients included in the review by Suzuki et al had malignant transformation [7]. In this review, it was observed that higher age of presentation was associated with increased risk of malignancy. In addition, in one of the cases reported by Sekino et al, there were extra duodenal malignancies involving 6 other viscera [16]. In view of these findings, it is reasonable to consider resection of these tumors. The present patient had mild dysplasia in the polyp and hence warranted resection.

## Conclusion

Purpose of reporting this case is because of

Rarity of solitary Peutz-Jeghers type hamartomatous polyp.

Giant size and varied presentation with symptoms of gastric outlet obstruction

## Conflict of Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and publication of this article.

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