CASE REPORT

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Giant Solitary Gastric Peutz-Jeghers Polyp Mimicking a Malignant Gastric Tumor: the Largest Described in Literature

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ABSTRACT

A solitary Peutz-Jeghers polyp is defined as a unique polyp occurring without associated mucocutaneous pigmentation or a family history of Peutz-Jeghers syndrome. Gastric solitary localization is a rare event, with only eight reported cases to date. We report herein the case of a 43-year old woman who presented with upper gastrointestinal bleeding, severe anemia, weight loss and asthenia. Endoscopy revealed a giant polypoid tumor with signs of neoplastic invasion of the cardia, with pathological aspect suggesting a Peutz-Jeghers hamartomatous polyp. Computed tomography suggested a malignant gastric tumor and a total gastrectomy was performed. The pathological specimen showed a giant 150/70/50 mm polypoid tumor and immunochemistry established the final diagnostic of a Peutz-Jegers type polyp. This is the largest solitary Peutz-Jeghers gastric polyp reported until now, and the second one mimicking a gastric malignancy with lymph node metastasis.

Key words: Peutz-Jeghers polyp — gastric polyp — solitary polyp — giant polyp.

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INTRODUCTION

A solitary or sporadic Peutz Jeghers polyp is defined as a unique polyp occurring without associated mucocutaneous pigmentation or a family history of Peutz-Jeghers syndrome [1]. The solitary Peutz-Jeghers polyps are extremely rare, and are more often encountered in the small bowel, but they may also occur in the large bowel and stomach [2, 3]. Gastric location is among the rarest, with only eight well documented cases to date [4-11]. We present herein the rare case of a woman with a giant solitary gastric Peutz-Jeghers polyp, mimicking a gastric malignancy, which required a total gastrectomy, and a review of the literature on solitary gastric Peutz Jeghers polyps.

CASE REPORT

A 43 year old female patient with no relevant past medical history was admitted for upper gastrointestinal bleeding, abdominal discomfort, weight loss of 6 kg in 3 months, loss of appetite and marked fatigability. Clinical evaluation showed only skin pallor. Laboratory data confirmed a severe microcytic hypochromic anemia (hemoglobin 6 g/dL). The carcinoembryonic antigen had normal value. During the workup for anemia, a gastroscopy showed a giant polypoid tumor arising immediately below the eso-cardial junction, occupying the entire stomach down to the pylorus. The cardia appeared surrounded by the tumor, with infiltration of the inferior aspect of the eso-gastric junction. The pathology report revealed arborizing smooth muscle bundles arising from muscularis mucosae and extending in the lamina propria, confirming the diagnosis of Peutz Jeghers hamartomatous polyp. The thoraco-abdominal computed tomography (CT) scan confirmed a large gastric tumor that stretched over a length of 14 cm from the eso-gastric junction to the pylorus, with mild enhancement after i.v. contrast administration (Fig. 1). There were multiple perigastric and celiac enlarged lymph nodes (16/11/15mm the largest) with significant contrast enhancement. The CT scan suggested a gastric malignant tumor with possible pancreatic involvement. A complete colonoscopy was also performed but no other polyps were found. Due to the large size of the tumor, an endoscopic excision was excluded, and a surgical approach was decided. Intraoperatively the stomach was slightly enlarged but fully mobile on the adjacent structures and its entire lumen was occupied by the tumor. Enlarged perigastric and celiac lymph nodes were noted, but intraoperative pathology with frozen

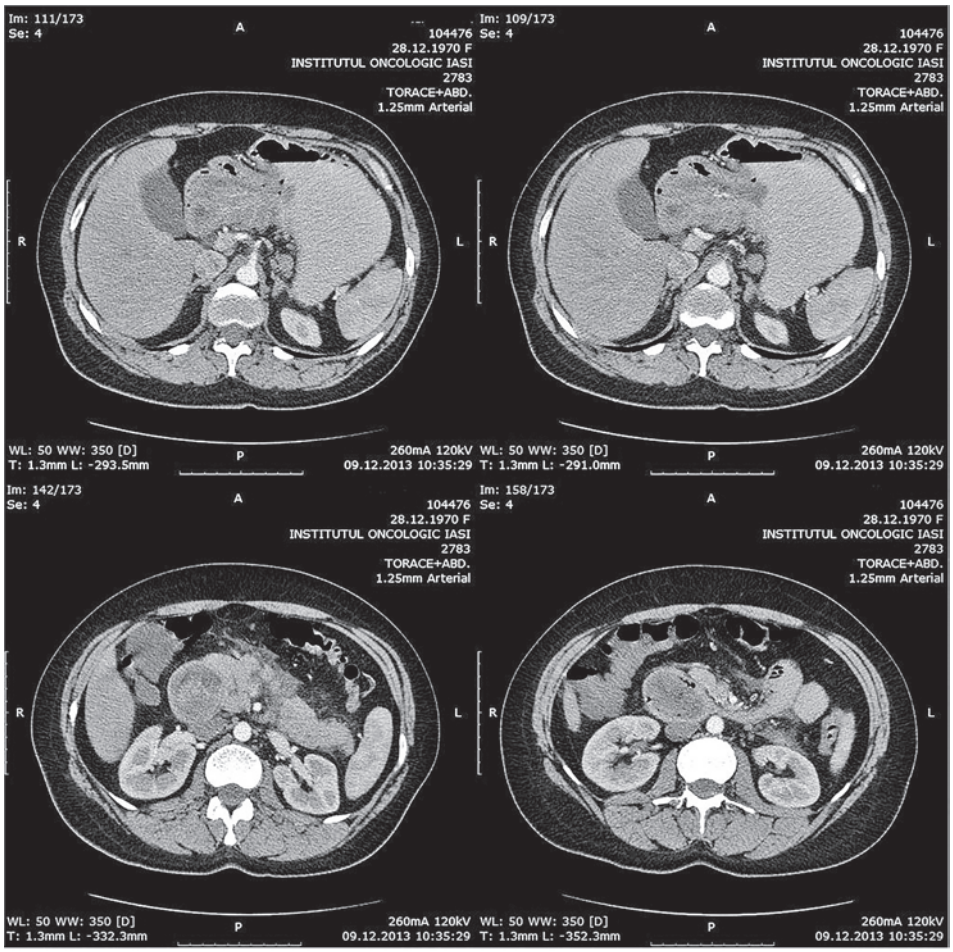

Fig. 1. CT scan showing the tumor with mild enhancement after i.v. contrast, and multiple perigastric and celiac trunk enlarged lymph nodes with significant contrast enhancement

sections excluded tumoral invasion. Total gastrectomy with esojejunal anastomosis Roux en Y loop was performed. Postoperative course was uneventful and the patient was discharged after 9 days.

The gastrectomy specimen revealed a 150/70/50 mm polypoid tumor with a 6.5/5.5 cm base around the cardia, developed around a central axis and covered with granular mucosa (Fig. 2). On cross-section the tumor showed multiple cysts (0.3 to 1.5 cm) with mucinous content. On microscopic examination, the crypts showed marked hyperplasia and columnar gastric superficial epithelium, as well as multiple gastric pit cystic ectasias up to 1.5 cm. The conjunctive axis of the tumor showed the muscularis mucosae dissociated by cardial type glands, with arborizing smooth muscle bundles extending up to the superficial lamina propria (Fig. 3). The polyp origin at the eso-gastric junction was covered with esophageal type mucosa. The 17 lymph nodes showed nonspecific aspect, with no cellular atypia. Immunochemistry analysis was highly positive for smooth muscle actin in the lamina propria (Fig. 3). All these features supported the diagnosis of a gastric Peutz Jeghers hamartomatous polyp.

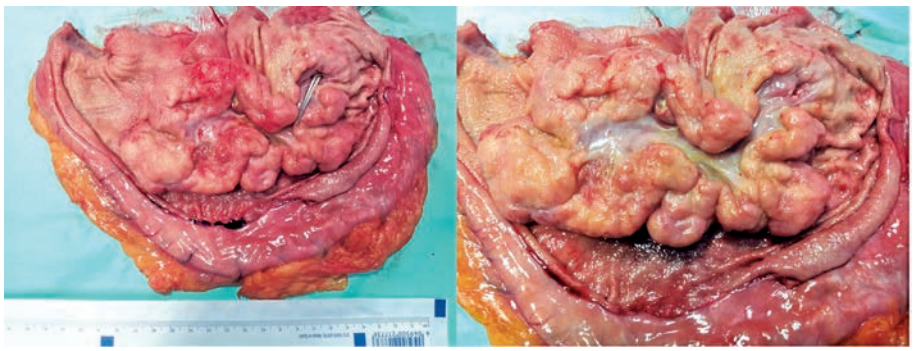

Fig. 2. Gastrectomy specimen revealing a 15/9/5 cm polypoid tumor developed at the level of cardia and encircling it (a forceps was introduce into the eso-gastric junction)

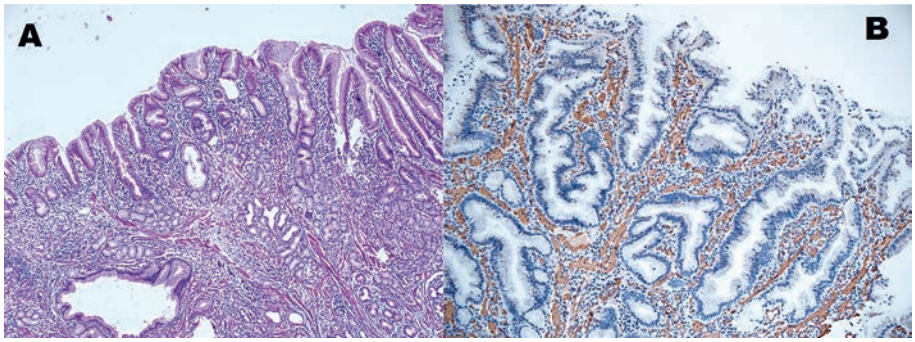

Fig. 3. A. Mucosa with cryptal hyperplasia and smooth muscle bundles into the lamina propria (H&E x5); B. SMA strongly positive in smooth muscle bundles in lamina propria and cryptal hyperplasia (SMA x10).

DISCUSSION

Solitary Peutz-Jeghers polyps are generally reported to have their origin in the small bowel, duodenum, colon, rectum and are extremely rare in the stomach, with only 8 well documented cases being reported to date (Table I). A solitary gastric Peutz-Jeghers polyp was noted for the first time by Kuwano et al in 1989 [4], and since then only 7 more cases have been reported [4-11]. For all reported cases the median age at diagnosis was higher than for patients with Peutz-Jeghers syndrome (48.8 years), with more women reported (62.5%) and larger size polyps ranging from 0.5 to 80 mm (median 40 mm).

In solitary Peutz-Jeghers polyps, the *STK11* gene mutation, which is considered the cause of the Peutz-Jeghers syndrome, was analyzed only in two cases, a gastric polyp [10] and a duodenal polyp [12] and both tests failed to detect the mutation. Histologically, solitary gastric Peutz-Jeghers polyps show the same features as Peutz-Jeghers syndrome polyps, with extensive arborization in the lamina propria of smooth muscle bundles arising from muscularis mucosae. Jin et al reported also a distinct finding of a proliferation of smooth muscle bundles in the submucosal layer [11]. Our case presented with multiple mucinous cysts ranging from 0.3 to 1.5 cm, feature that was reported only by Sakadamis et al and is more characteristic for juvenile hamartomatous polyps [7].

Patients with Peutz-Jeghers syndrome are well known for their increased risk for gastrointestinal and non-gastrointestinal cancers, a risk evaluated to be 18 times greater than in the general population. The relative risk is appreciated at 15.2 and the lifetime risk of any type of cancer is 93%. The lifetime risk for gastric cancer is 29% and the average age of diagnosis is 30 years [13]. Larger hamartomas may contain foci of adenomatous tissue and a malignant development in a hamartomatous polyp has been described in the Peutz-Jeghers syndrome [14-16]. Different reports suggest that malignancy follows a hamartoma-adenoma-carcinoma sequence, comparable to that in familial adenomatous polyposis [17-19]. The rate of malignant transformation of a gastric polyp in Peutz-Jeghers syndrome is around 13% [17, 20]. Oncel et al followed 8 patients with solitary Peutz-Jeghers polyps (5 in the colon, 2 in the duodenum, 1 in the stomach) for a period of 11.5 years. All polyps were endoscopically resected and no recurrences or Peutz-Jeghers syndrome associated neoplasms were found. The authors concluded that solitary Peutz-Jeghers polyps do not carry a risk for gastrointestinal cancer and are not an indication for specific high-risk screening [9]. On the other hand, Burkart et al reviewed the histology of 102 polyps and identified only 3 solitary small bowel Peutz-Jeghers polyps [21]. Although not all these patients met the diagnostic criteria for Peutz-Jeghers syndrome, all of them had clinical features suggesting it: two had pancreatic cancer, one associated also a strong gastrointestinal malignancy history and one had a glomus tympanicum tumor and bilateral ovarian masses. The authors concluded that if solitary Peutz-Jeghers polyps exist, they are extremely rare and may carry the same cumulative lifetime risk of cancer as those with the syndrome, and also suggest that these solitary polyps may be an incomplete form of Peutz-Jeghers syndrome.

None of the nine (including ours) gastric solitary Peutz-Jeghers polyps reviewed in the literature showed associated

Table I. Reported cases with solitary gastric Peutz-Jeghers polyp [4-11]

No.	Author	Year	Polyp size (mm)	Age (years)	Gender	Treatment
1	Kuwano H	1989	20	teenage	male	endoscopic resection
2	Grisendi A	1990	20	53	female	endoscopic resection
3	Hunt J	1996	80	27	female	distal gastrectomy
4	Sakadamis AK	2001	75*50	47	female	partial gastrectomy
5	O´Loughlin CJ	2002	70*40	38	female	endoscopic resection
6	Oncel M	2002	5	78	male	endoscopic resection
7	Harbaum L	2009	10	61	male	endoscopic resection
8	Jin JS	2012	40*30	71	female	subtotal gastrectomy

features to suggest Peutz-Jeghers syndrome. We have to note that in three cases, including ours, the CT scan revealed features of a gastric malignant tumor, two of them presented also with enlarged lymph nodes suggestive of malignancy [7, 11]. While a malignant transformation of a hamartoma in Peutz-Jeghers syndrome and also in solitary Peutz-Jeghers polyps outside the stomach was clearly demonstrated, for gastric solitary Peutz-Jeghers there is no evidence to support that [13, 14, 22]. Only Harbaum et al reported a low-grade intraepithelial neoplasia in their case [10]. However, due to its giant size of 150 mm, there is a major chance for the polyp we encountered to harbor malignant foci.

Treatment for gastric solitary Peutz-Jeghers polyps may vary from endoscopic resection to gastric resection according to their size, location and complications. Most authors recommend resection of polyps greater than 1-1.5 cm in Peutz-Jeghers syndrome, surgery being recommended for larger or complicated polyps [23-25]. Regarding the gastric solitary Peutz-Jeghers polyps, in five cases polyps were resected endoscopically and four patients underwent a gastric resection. We performed the first total gastrectomy for a solitary Peutz-Jeghers polyp, and this operation was justified by the size of the polyp and the theoretical risk for occult malignant foci. Another rationale in favor of radical surgery was also the location around the gastro-esophageal junction. Also malignancy cannot be definitely excluded in such a large tumor, despite histological diagnosis of Peutz-Jeghers hamartoma, even more so when endoscopic features and CT-scan elements suggested an aggressive tumoral behavior.

CONCLUSION

Solitary gastric Peutz-Jeghers polyps are rare events, apparently distinct from Peutz-Jeghers syndrome, with the same pathological characteristics, but with different family history and biological characteristics. Due to the small number of cases reported, no clear conclusion regarding the risk of cancer development or *STK11* germline mutations can be drawn. Our polyp is the largest solitary gastric Peutz-Jeghers polyp reported in the literature and the second one mimicking a gastric malignancy with lymph node metastasis.

Conflicts of interest: None to declare.

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