



# Solitary Peutz-Jeghers type hamartomatous polyp in duodenum with gastric foveolar epithelium: a case report

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Peutz-Jeghers type hamartomatous polyp is known to be associated with Peutz-Jeghers syndrome, which shows characteristic multiple hamartomatous polyp involvement in the gastrointestinal tract, combined with mucocutaneous symptom, familial history of Peutz-Jeghers syndrome or *STK11/LTB1* mutation. However, some cases showing histologic appearance of the polyps discovered in Peutz-Jeghers syndrome while lacking other diagnostic criteria of the syndrome have been reported, and these are called solitary Peutz-Jeghers type polyps. Herein, we report a case of solitary Peutz-Jeghers type polyp covered with heterotopic epithelium. The patient was 47-year-old female without any mucocutaneous symptoms nor familial history of Peutz-Jeghers syndrome. Microscopic examination revealed Peutz-Jeghers type hamartomatous polyp in duodenum covered with gastric type foveolar epithelium. Considering the definition of hamartomatous polyp, which is, the abnormal overgrowth of the indigenous epithelial component, the histological feature of current case is noteworthy in a point that it shows proliferation of heterotopic component, rather than the indigenous component.

**Key Words:** Hamartoma; Intestinal polyps; Duodenum; Gastric mucosa; Peutz-Jeghers syndrome

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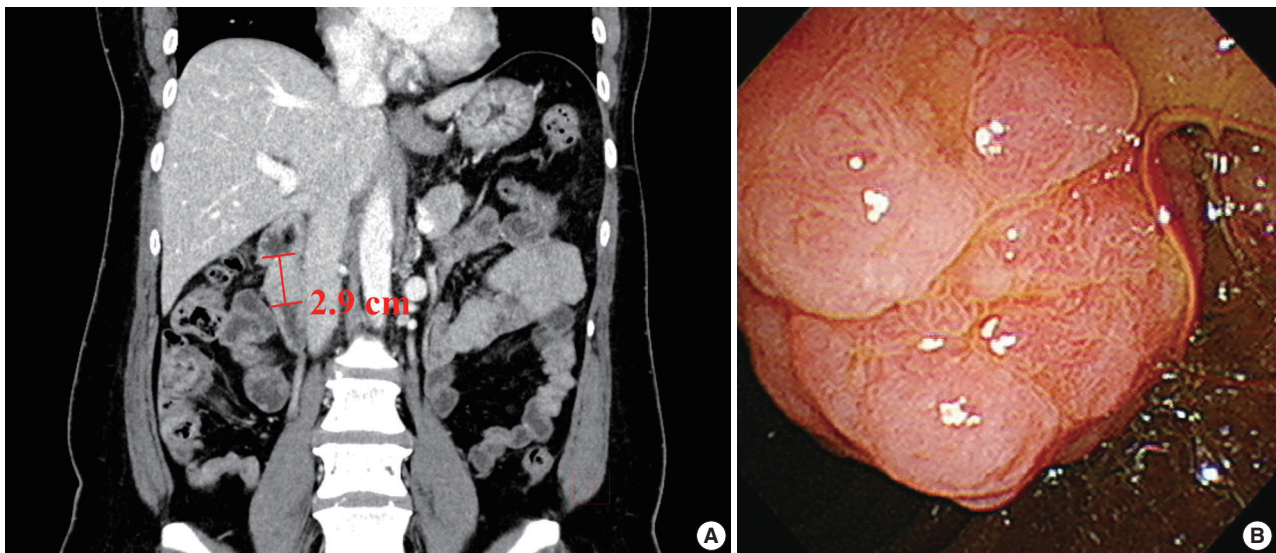
Hamartomatous polyp is defined as a polyp with disorganized growth of normal cells indigenous to the organ. Especially, Peutz-Jeghers type hamartomatous polyp represents as the arborization of smooth muscle within the lamina propria beneath pre-existing normal epithelium of the organ it involves. This polyp is known to be associated with Peutz-Jeghers syndrome, which shows characteristic multiple hamartomatous polyp involvement in the gastrointestinal (GI) tract, combined with mucocutaneous symptom, familial history of Peutz-Jeghers syndrome or *STK11/LTB1* mutation [1]. However, there have been a few cases which are histologically identical to those polyps associated with Peutz-Jeghers syndrome, without other clinical evidence diagnostic of Peutz-Jeghers syndrome. These polyps are called solitary Peutz-Jeghers type polyps [2]. We experienced a case of solitary Peutz-Jeghers type hamartomatous polyp in duodenum covered with gastric type foveolar epithelium, rather than the native intestinal columnar epithelium.

## CASE REPORT

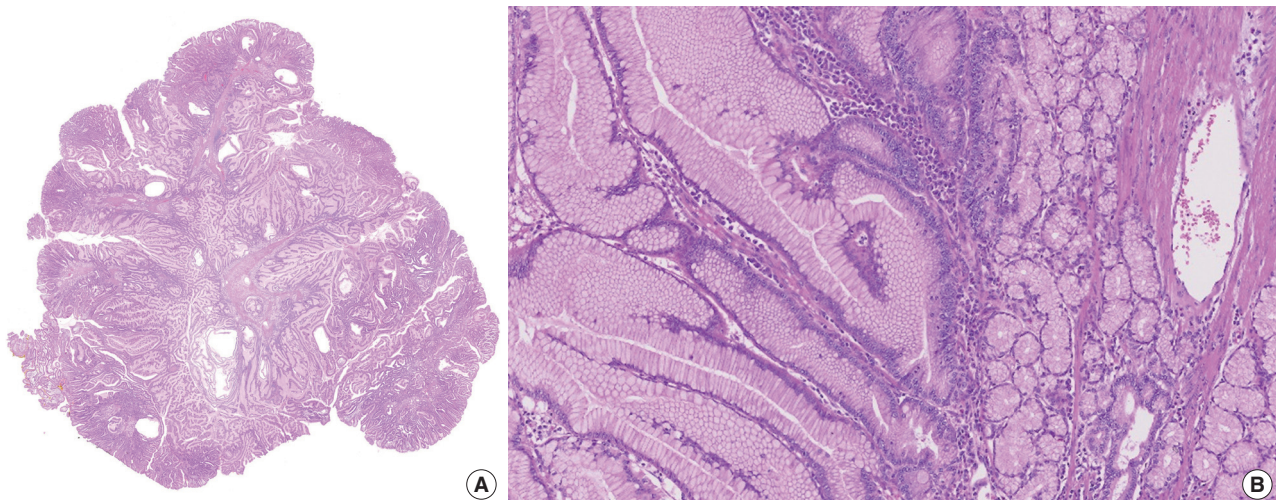
A 47-year-old woman visited outpatient clinic for incidentally discovered duodenal polyp during regular medical check-up. She did not experience any associated symptoms. She had a history of invasive ductal carcinoma of both breasts in her early to mid-30s and been treated for papillary thyroid carcinoma in her late 30s.

On initial esophagogastroduodenoscopy, a pedunculated polyp occupying half of the luminal space was found in the 2nd portion of duodenum. Histological examination of a biopsy specimen mainly showed hyperplastic gastric foveolar epithelium. Following computed tomography scan revealed a 2.9-cm-sized polypoid lesion in duodenum without any evidence of metastasis (Fig. 1A). The polyp was entirely resected by endoscopic submucosal dissection (Fig. 1B). The patient was discharged without any post-procedural complications.

Histological examination of the resected polyp showed disorganized overgrowth of epithelial cells supported by thick, branching



**Fig. 1.** Radiologic and endoscopic findings of the duodenal polyp. (A) Abdominal computed tomography image showing 2.9-cm-sized mass in duodenal 2nd portion. (B) A pedunculated polyp occupying half of the duodenal lumen discovered in endoscopic examination.



**Fig. 2.** Histological examination of the resected polyp. (A) Hematoxylin and eosin staining of the resected polyp showing arborizing smooth muscle bundles with overgrowth of superficial foveolar epithelium. (B) Foveolar type epithelium lying over pyloric type-like mucinous glands without dysplasia.

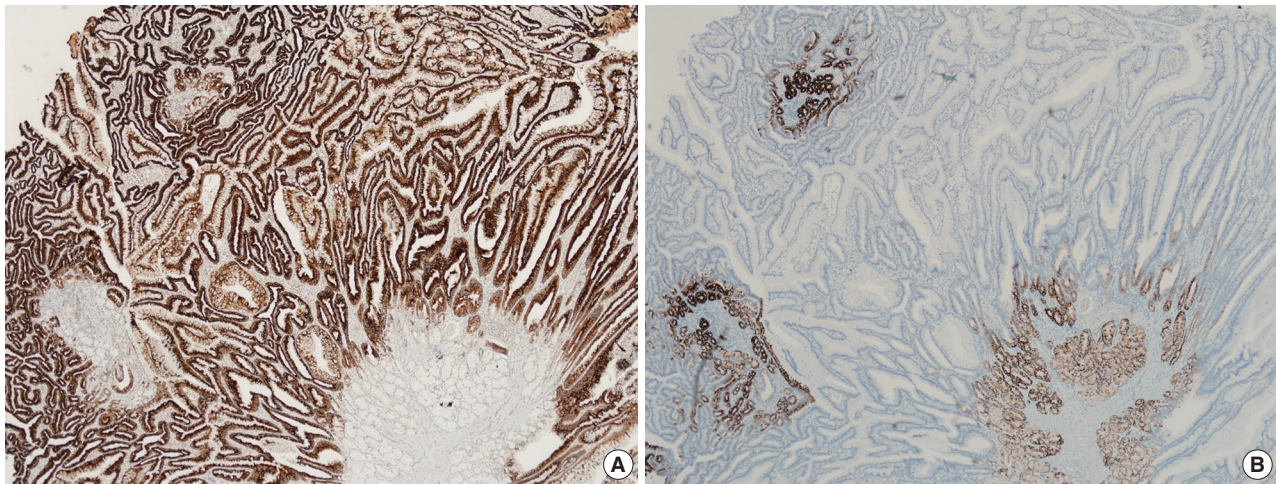
bundles of smooth muscles. The epithelium mainly consisted of foveolar type cells lying over mucinous glands resembling pyloric glands without dysplastic change (Fig. 2A, B). A small amount of surrounding duodenal tissue was identified at the margin of the specimen and it showed focal foveolar type epithelium among normal duodenal villous structures, suggestive of gastric metaplasia. Immunohistochemical staining for MUC5AC and MUC6 confirmed the covering hyperplastic epithelium to be gastric foveolar type with pyloric glands underneath (Fig. 3A, B).

Follow-up endoscopy after 6 months from the procedure revealed no additional polyps. In an in-depth interview after-

wards, the patient insisted she had never been diagnosed with hamartomatous polyp before and denied of any familial history nor any mucocutaneous symptoms. Genetic assessment for *STK11/LTB1* mutation was not done. According to these findings, the polyp was finally diagnosed as solitary Peutz-Jeghers type polyp with gastric foveolar epithelium.

## DISCUSSION

Hamartomatous polyp is characterized by the arborization of smooth muscle bundle up to lamina propria with near-normal



**Fig. 3.** Immunohistochemical stainings of the resected polyp. (A) Immunohistochemistry for MUC5AC confirms the presence of the overlying gastric type foveolar epithelium. (B) Pyloric gland-like structures showing positivity for MUC6 staining.

overlying epithelium. The surface epithelium of hamartomatous polyp is known to be identical with those of adjacent normal mucosa. Multiple hamartomatous polyps are frequently associated with various genetic syndromes, such as juvenile polyposis syndrome, Peutz-Jeghers syndrome and Cowden syndrome [1].

Of note, multiple GI tract polyps, mucocutaneous symptoms, *STK11/LTB1* mutation and familial history of Peutz-Jeghers syndrome are characteristic of Peutz-Jeghers syndrome. As this syndrome represents an increased risk of developing cancers, patient with solitary Peutz-Jeghers type hamartomatous polyp must be meticulously examined to exclude a possibility of syndromic involvement. If the polyp is proved to have no association with Peutz-Jeghers syndrome, it is defined as solitary Peutz-Jeghers type hamartomatous polyp. Although the histological aspect of solitary Peutz-Jeghers type hamartomatous polyp is indistinguishable from the polyps of Peutz-Jeghers syndrome, this solitary polyp seems to be of separated entity, as it does not show *STK11/LTB1* mutation characteristic of the syndrome. Furthermore, the frequently involved site is quite different. The polyp of the syndrome mostly occurs in small intestine, whereas the solitary counterpart develops mostly in sigmoid colon, followed by duodenum, rectum, jejunum, and stomach, according to the previously published literatures [2,3].

Solitary Peutz-Jeghers type hamartomatous polyp itself can provoke GI bleeding and intussusception, which may manifest as acute GI symptoms in some patients [4-6]. Some cases represented with dysplasia and even carcinogenesis [7,8]. Thus, complete resection of the polyp by either endoscopic or surgical procedure should be considered.

In our case, the patient was asymptomatic, and the polyp was

found during routine health check-up. The resected polyp showed hamartomatous proliferation of gastric foveolar epithelium over pyloric gland-like structures in the duodenum. Whether this aspect is related to prior gastric metaplasia or abnormal differentiation of endoderm is unclear at this moment. To date, a case of pyloric metaplasia involving multiple hamartomatous jejunal polyps in a patient with Peutz-Jeghers syndrome and a case of solitary Peutz-Jeghers type polyp with gastric antral and fundic gland mucosa have been reported [6,9]. Considering the definition of hamartomatous polyp, which is, the abnormal overgrowth of the indigenous epithelial component, the histological feature of current case is noteworthy in a point that it shows proliferation of heterotopic component, rather than the indigenous component.

#### Ethics Statement

Formal written informed consent was not required with a waiver by the appropriate IRB (Asan Medical Center IRB No. 2022-1339).

#### Availability of Data and Material

Data sharing not applicable to this article as no datasets were generated or analyzed during the study.

#### Code Availability

Not applicable.

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### Conflicts of Interest

The authors declare that they have no potential conflicts of interest.

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