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Autoimmune Polyglandular Syndrome Type II Associated with Jejunal Gastrointestinal Stromal Tumor: Diagnosis by Capsule Endoscopy



Figure 1 Capsule endoscopy revealed a mass in the proximal jejunum.

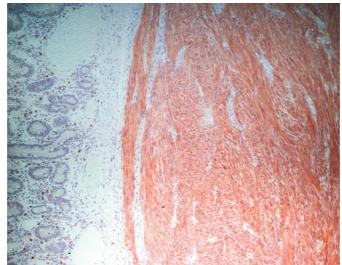


Figure 3 Histopathological examination revealed a highly cellular spindle-cell gastrointestinal stromal tumor (GIST) expressing Kit (CD117 antigen).

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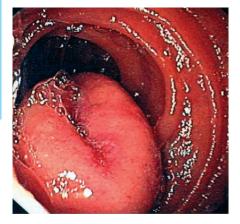


Figure **2** Push enteroscopy confirmed an ovoidal umbilicate protruding jejunal mass.

A 49-year-old woman with autoimmune polyglandular syndrome type II (Addison's disease and hypothyroidism) was referred to our department, as she was suffering from persistent iron-deficiency anemia and had positive results from fecal occult blood test. The history of bleeding was approximately 3 years during which period she had been treated with iron supplementation. Mean Hb concentration was 8.4 g/100 ml (range 6.1–11.7). Previous diagnostic investigations had included two esophagogastroduodenoscopies, two total colonoscopies, a 99Tc-labeled red cells scan, a 99Tc-pertechnetate scan, a small-bowel follow-through investigation, and a computed tomography (CT) scan. All these tests had failed to identify the source of bleeding.

Capsule endoscopy was performed and identified a small-bowel mass. Rapid® location showed the lesion to be in the upper left midline, and it was estimated to be in the proximal jejunum area (Figure 1). Push enteroscopy was subsequently carried out and confirmed an ovoidal umbilicate protruding mass of the proximal jejunum (size 4 cm) (Figure 2). The lesion was resected laparoscopically. The gross specimen of resected small bowel showed that the tumor had a large extraluminal component. No pathological lymph nodes or liver metastases were found. Histopathological investigation demonstrated a highly cellular spindle-cell tumor which turned out to be a gastrointestinal stromal tumor (GIST) expressing Kit (CD117 antigen) (Figure 3).

Tumors of the small bowel comprise 5% to 7% of all gastrointestinal tumors. The most important symptom in cases of small-bowel neoplasia is undoubtedly obscure bleeding with secondary iron-deficiency anemia. Indeed, small-bowel tumors are the second most common cause of obscure gastrointestinal bleeding, accounting for 5% to 10% of all cases of

chronic blood loss. Among patients with obscure gastrointestinal bleeding, small-bowel tumors are the single most common lesion in patients younger than 50 years [1]. The median time to diagnosis for patients with obscure bleeding has been estimated as 2 years [2]. Delays in diagnosis may alter the outcome, and should be minimized whenever possible. Careful utilization of diagnostic examinations may lead to early identification of a potential small-bowel bleeding source and may help improve the diagnostic outcome while decreasing the cost of hospitalization.

Our patient was ultimately diagnosed by means of capsule endoscopy to have a small-bowel tumor and underwent curative surgery. If capsule endoscopy is carried out early in the course of the work-up of these patients (i.e. immediately after negative esophagogastroduodenoscopy and colonoscopy), it could shorten considerably the time necessary to reach a diagnosis and allow the early institution of definitive treatment in a significant proportion of patients [3].



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