Unexpected findings during MR enterography surveillance in a patient with Peutz-Jeghers syndrome

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Section: Abdominal imaging
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Technique: MR
Special Focus: Pathology Case Type: Clinical Cases
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Patient: 22 years, female

Clinical History:

A 22-year-old woman with a known history of Peutz-Jeghers syndrome presented for follow-up, reporting a recent episode of intestinal bleeding. A palpable mass in the right lower quadrant was detected and pigmented macules on the lips were noted. An MR enterography followed.

Imaging Findings:

MR enterography revealed multiple polyps in the stomach and small bowel and an 4 cm polyp in the caecum. Confluent millimetric polyps resulted in a thickened appearance of the small bowel wall. Jejunal luminal dilatation was also noted.

Discussion:

A. Background

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disorder. To make the diagnosis a hamartomatous polyp needs to be detected as well as at least two of the following: mucocutaneous pigmentation, polyposis of the small bowel or known family history of the syndrome [1]. The condition has been attributed to the mutation of the STK 11 gene [2].

B. Clinical perspective

Hamartomatous polyps have normal cellular elements of the gastrointestinal tract with a markedly distorted architecture. The median time to first presentation with polyps is 11-13 years of age. Large polyps in the small bowel present with gastrointestinal bleeding, anaemia, intussusceptions, or bowel obstruction.

MR enterography can detect clinically relevant polyps. Expert consensus suggests that regular surveillance of the small bowel in patients with PJS aiming to enable identification and prophylactic removal of large luminal polyps may help to reduce the frequency of emergency laparotomy and both immediate- and long-term complications associated with recurrent abdominal surgery [3]. In addition, the overall risk of cancer in PJS exceeds 90% [1], making surveillance a priority for these patients.

C. Imaging perspective

Polyps measuring 15 mm or more present the highest risk for intussusception and bowel obstruction, leading to the conclusion that polyps measuring over 10 mm, symptomatic or rapidly growing need to be excised [4-6].

Different ways have been proposed for the imaging surveillance of Peutz-Jeghers disease, aiming to the detection of
small-bowel tumours, such as capsule endoscopy and double-balloon enteroscopy, the second one being more invasive and prolonged and thus not supported [7]. MR enterography has been proven a safe and effective means of surveillance, identifying polyps even in segments of the bowel not examined at endoscopy or surgery [7]. Final diagnosis for a patient with a known history of Peutz-Jeghers syndrome presenting with intestinal bleeding is usually made with either capsule endoscopy, CT enterography or enteroclysis or MR enterography or enteroscopy [7], as preferred in our institution.

D. Outcome
Endoscopic resection of the large colonic polyp was performed, revealing its hamartomatous characteristics at histopathology. The patient recovered fully and was discharged uneventfully. The performance of the MR enterography guaranteed the speedy and effective diagnosis of the cause of the gastrointestinal bleeding.

E. Take home message
MR enterography has been suggested as an effective method of identifying large polyps of the intestine during surveillance in Peutz-Jeghers syndrome. In this case it led to the successful patient care delivery and should be considered in similar cases as a non-invasive and effective imaging method.

**Differential Diagnosis List:** Intestinal bleeding caused by a large polyp in a patient with Peutz-Jeghers disease., Intussusception, Adenocarcinoma

**References:**


Figure 1

Description: Mucocutaneous pigmented lesions in the oral cavity (arrowheads) and lower lip (arrows). Origin: D. Chourmouzi, Interbalkan Medical Center Department of Diagnostic Radiology, Thessaloniki, Greece
**Description:** (a), (c) Coronal Balanced GE: stomach and small bowel polyps and a large polyp in the caecum, (b), (d) Ultrafast GE post contrast: polyp and abnormally thickened small bowel enhancement corresponding to diffuse micropolyposis.

**Origin:** D.Chourmouzi, Interbalkan Medical Center, Department of Diagnostic Radiology, Thessaloniki, Greece.
Figure 3

Description: Coronal Balanced GE: the cecal and stomach polyps as well as abnormally thickened small bowel wall corresponding to diffuse micropolyposis. Origin: Chourmouzi D., Interbalkan Medical Center, Department of Diagnostic Radiology, Thessaloniki, Greece
Description: Coronal Ultra fast GE image: the caecal and stomach polyps as well as abnormally thickened small bowel wall corresponding to diffuse micropolyposis. Origin: D. Chourouzi, Interbalkan Medical Center, Department of Diagnostic Radiology, Thessaloniki, Greece
Description: Coronal Ultra fast GE post contrast image showing enhancement of the cecal (arrows) and stomach (arrowheads) polyps as well as enhancement of abnormally thickened small bowel wall corresponding to diffuse micropolyposis. Origin: D. Chourouzi, Interbalkan Medical Center, Department of Diagnostic Radiology, Thessaloniki, Greece
Figure 5

a

Description: Axial Balanced GE at the level of the caecum, showing a large polyp (arrows).
Origin: D. Chourmouzi, Interbalkan Medical Center, Department of Diagnostic Radiology, Thessaloniki, Greece

b

Description: Axial Ultra fast GE post contrast image showing multiple enhanced polyps of the stomach (arrowheads).
Origin: D. Chourmouzi, Interbalkan Medical Center, Department of Diagnostic Radiology, Thessaloniki, Greece
Description: Axial Ultra fast GE post contrast image showing enhanced polyps of the caecum (arrows). Origin: D. Chourmouzi, Interbalkan Medical Center, Department of Diagnostic Radiology, Thessaloniki, Greece