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Abstract

Peutz–Jeghers syndrome (PJS) is a rare autosomal-dominant inherited disorder characterized by gastrointestinal hamartomas, mucocutaneous pigmentation, and an elevated cancer risk. Moreover, intussusception risk may be as high as 50% at the age of 20 years and is caused by large polyps. There is some evidence that endoscopic surveillance of PJS patients with removal of small intestinal polyps with a diameter of more than 15 mm efficiently prevents intussusceptions. In recent years, capsule endoscopy (CE) has largely replaced small-bowel radiography techniques to screen for small-bowel polyps. Magnetic resonance imaging may be equally efficient as CE for screening of large polyps. Balloon enteroscopy may be used for endoscopic snare resection of polyps. This article is part of an expert video encyclopedia.

Keywords

Balloon enteroscopy; Capsule endoscopy; Enteroscopy; Peutz–Jeghers syndrome; Polyposis syndrome; Video.

Video Related to this Article

Video available to view or download at doi:10.1016/S2212-0971(13)70115-8

Technique

- Capsule endoscopy (CE).
- Small-bowel endoscopy.

Materials

- Capsule endoscope: PillCam SB2, Given Imaging EMEA, Hamburg, Germany.
- Double-balloon enteroscopy: DBE EN-450T5, Fujinon/FujiFilm, Willich, Germany; working channel: 2.8 mm, working length: 230 cm.
- Snare for polypectomy: Diameter 30 mm, length 260 mm, for 1.8 mm or 2.2 mm working channel, medwork, Höchststadt/Aisch, Germany.

Background and Endoscopic Procedure

Peutz–Jeghers syndrome (PJS) is a rare autosomal-dominant inherited disorder characterized by gastrointestinal hamartomas, mucocutaneous pigmentation, and an elevated cancer risk. Moreover, cumulative intussusception risk may be as high as 50% at the age of 20 years and is caused by polyps with an average diameter of 35 mm. Study results support the approach of endoscopic surveillance of PJS patients,^{1–5} with removal of small intestinal polyps with a diameter of more than 15 mm to prevent intussusceptions. CE has been suggested to replace

former small-bowel radiography techniques to screen for small-bowel polyps, but magnetic resonance imaging may be comparable for screening for significant polyps. Balloon enteroscopy may be used for endoscopic snare resection of polyps.

In the present case, several small-bowel polyps with typical features of PJS were detected by CE. Afterward, the polyps were resected by double-balloon enteroscopy. Histological workup showed the typical features of a PJ polyp with extensive smooth muscle arborization throughout the polyp with branching at the surface.

Key Learning Points/Tips and Tricks

- PJS polyps may have easily recognizable features, such as long stalks, arborizing structure, and intact surface. Ulceration is not a typical feature.
- Intussusception risk may be very high in young patients and preliminary data argue strongly for endoscopic resection of large (> 15 mm) Peutz–Jeghers polyps in the small bowel to prevent intussusception.

Complications and Risk Factors

Complication rates of endoscopic polypectomy in the small bowel might be as high as 10–15% and comprise perforation and bleeding.

Scripted Voiceover

Time (min:sec)	Voiceover text
0:00	In this young male patient with known Peutz–Jeghers syndrome capsule endoscopy is performed to screen for small bowel polyps. The first polyps are detected in the duodenum.

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- 0:26 A big Peutz-Jeghers polyp is visualized now occupying most of the display window. It takes some time for the capsule to pass the polyp. Some debris sticks to the polyp and suggests a stenotic effect of the lesion.
- 0:46 A large polyp on the right side of the picture may easily be overlooked. Its structure and configuration are similar to the small bowel itself; only the variant surface structure helps to disclose the hamartoma.
- 1:21 Small Peutz-Jeghers polyps feature the typical arborizing surface of these hamartomas.
- 1:43 Moreover, the long stalk is a characteristic attribute of these polyps.
- 1:56 The dimensions and the multitude of the small-bowel polyps in this case are best observed using double-balloon enteroscopy.
- 2:25 Enteroscopy is also helpful to remove the big polyps by using snare resection technique. Thereby, future intussusception might be avoided.
- The overtube of the enteroscope is kept in place while the endoscope is withdrawn to retrieve the resected polyp.

- 3:12 Histopathology reveals smooth-muscle hyperplasia and an elongated, arborized pattern of polyp formation, resulting in the formation of islands of epithelium within the underlying smooth muscle.

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