Clinical Quiz

Abdul-Wahed N. Meshikhes, MBChB, FRCS, Ehab A. Attia, MS, MD, Nadia N. Al-Eisa, MD, Ali A. Al-Zahir, MB.

Departments of Surgical Specialties and Body Imaging,

King Fahad Specialist Hospital, Dammam 34111, Eastern Province, Kingdom of Saudi Arabia.

Tel. +966 (3) 843 1111 Ext. 4414. Fax. +966 (3) 855 1019. E-mail: meshikhes@doctor.com

Notice: Authors are encouraged to submit quizzes for possible publication in the Journal. These may be in any specialty, and should approximately follow the format used here (maximum of 2 figures). Please address any submissions to: Editor, Saudi Medical Journal, Armed Forces Hospital, PO Box 7897, Riyadh 11159, Kingdom of Saudi Arabia. Tel. +966 (1) 4777714 Ext. 6570. Fax. +966 (1) 4761810 or 4777194.

An unusual cause of intestinal obstruction

Clinical Presentation

A 28-year-old Saudi male presented with symptoms and signs of intestinal obstruction. He had previous laparotomy 10 years earlier for a similar presentation. His toes and fingers showed scattered pigmentation (Figure 1). Computerized tomography (CT) of the abdomen was requested (Figure 2).



Figure 1 - Photographs showing the characteristic skin pigmentation in the toes. Similarly, skin pigmentations were evident in patient's fingers.



Figure 2 - Computerized tomography of the abdomen showing the intussusception with the characteristic doughnut and pseudokidney signs.





Figure 3 - Operative photograph of **a**) intussusception. The polyps usually act as leading points. b) Excised polyps which were of variable sizes. The broad-based polyp which shown in the center of the photograph was excised by a limited bowel resection.

Questions

1. What is the diagnosis?

2. What is the cause of the obstruction?

Clinical Quiz

Answers

- 1. The diagnosis is Peutz-Jeghers syndrome.
- 2. The most likely cause of this intestinal obstruction is intussusception which is clearly seen in the CT scan (Figure 2).

Discussion

Peutz-Jeghers syndrome is a rare autosomal dominant disorder. It is characterized by hamartomatous polyposis of the gastrointestinal tract and melanotic pigmented spots. The pigmented spots are located mainly in the mucous membrane of the oral cavity and lips. They can also be seen in the skin of fingers and toes as shown in Figure 1. The lesions tend to gradually increase in intensity towards, and during, puberty, but slowly start to fade away later in adult life, and may even disappear in some cases. Patients often have a history of multiple laparotomies for various polyp-related complications, for example bleeding and intestinal obstruction. Intestinal obstruction may also occur as a result of intraluminal obstruction by a large polyp. Malignant transformation of polyps occurs in only 2% - 3%. Laparotomy in this patient revealed multiple intussusceptions (Figure 3a) which were reduced and polyps acting as leading points were excised (Figure 3b) Polypectomy was performed via multiple enterotomies, and many polyps (2 were in the large bowel) were removed to minimize the risk of future obstruction or bleeding that may necessitate another laparotomy. Large broad-base polyps may be treated by limited small bowel resection. Preoperatively, once polyps are detected endoscopically, attempts should be made to snare them by upper and lower endoscopies, which should be performed every 2 years, as advised by St. Mark's Polyposis Surveillance.

References

- 1. Spigelman AD, Philips RKS. Peutz-Jeghers syndrome. In: Philips RKS, Spigelman AD, Thomson JPS, editors. Familial adenomatous polyposis and other polyposis syndromes. 1994 London (UK): Edward Arnold; p. 188-202.
- 2. Spigelman AD, Arese P, Philips RKS. Polyposis: the Peutz-Jeghers syndrome. Br J Surg 1995; 82: 1311-1314.
- 3. Meshikhes AW, Al-Saif O, Al-Otaibi M. Duodenal and ampullary obstruction due to a Peutz-Jeghers polyp. *Euro J Gastroenterol Hepatol* 2000; 12: 1239-1241.
- 4. Boardman LA, Thibodeau SN, Schaid DJ, Lindor NM, McDonnell SK, Burgart LJ et al. Increased risk for cancer in patients with the Peutz-Jeghers syndrome. *Ann Intern Med* 1998; 128: 896-899.
- 5. Meshikhes AW. Peutz-Jeghers syndrome: Association with cancer and surgical management. Ann Saudi Med 2000; 20: 413-414.