International Journal of Medical Science and Health Research

Vol. 3, No. 06; 2019

ISSN: 2581-3366

Multiple Long Segment Small Bowel Intussusceptions Due to Peutz-jeghers Syndrome

*Murat Sanal, MD *Paul Hechenleitner, MD

*Medical University Innsbruck Department of Pediatric Surgery-Austria

Abstract

Peutz-Jeghers syndrome (PJS) is a rare, autosomal dominant disease. It is characterised by gastrointestinal polyps, mucocutaneous pigmentation and the increased risk of developing cancer. Radiological investigations such a contrast studies, CT, MRI, ultrasound and endoscopy are useful in diagnosis and surveillance. The purpose of this report was to analyse the clinical characteristics, diagnosis and surgical treatment of a 12-year-old boy, who suffered from long segment small bowel intussusceptions caused by PJS.

Keywords: Peutz-Jeghers syndrome, intussusception, childhood, polyp

I. Introduction

Peutz-Jeghers syndrome (PJS) is a well-defined rare, inherited, autosomal dominant disease, leads enclosed gastrointestinal polyposis and hyperpigmentation on mucocutaneous regions of the body. PJS frequently causes rectal bleeding, abdominal pain, intussusceptions, and over time, the development of a malignancy.

Intussusception is a well-known consequence of PJS and may require repeated laparotomies and bowel resections.

Our 12-year-old patient was first diagnosed four years ago, and underwent a laparotomy and resection of polyps, after being admitted with colicky abdominal pain, recurrent vomiting and a worsened general condition.

II. Material and methods

At the age of eight years old, our patient was referred from the primary physician to the paediatric gastroenterology department of our hospital, to clarify an underlying illness that resulted in lip hyperpigmentation and iron deficiency anaemia.

He had Peutz-Jeghers syndrome with no family history and the diagnosis was confirmed through upper and lower gastrointestinal endoscopy investigations, with a histological examination of multiple polyps removed from the stomach, jejunum, terminal ileum and rectum.

During further investigations, MRI imaging showed a typical “target sign” in the upper abdominal region, which is a characteristic feature for invagination, which did not cause serious clinical symptoms (Fig. 1).

Endoscopy examinations revealed at the proximal jejunum, right distal to the ligament of Treitz, one polyp, with a size of 5x6 cm. However, it did not cause a mechanical obstruction, there were no signs of bleeding and the patient had no feeding problems. Endoscopic removal was not

Vol. 3, No. 06; 2019

ISSN: 2581-3366

possible on one hand because of the broad basis of the polyp and on the other hand because of a difficult-to-access localisation.

During a symptom free, 4-year period, the patient remained under paediatric surveillance with yearly endoscopy controls, wherein further small gastro-duodenal polyps and rectum polyps were removed endoscopically. However, the big proximal jejunal polyp remained.

At the age of 12 years old, the patient was admitted to our department of paediatric surgery because of colicky, abdominal pain and recurrent vomiting at. Over three days, he could not eat normally, only fluid intake was possible and recurrent vomiting worsened his general condition.

Clinical investigations showed slight tenderness on the upper abdomen but no signs for acute abdomen. After admission we started conservative treatment with fluid electrolyte infusion.

Abdominal X-ray investigation, after contrast uptake, showed no typical signs of ileus. Sonography revealed typical signs of small intestine intussusception, which was confirmed with an MRI. An endoscopy showed the known polyp at the proximal jejunum, which had grown considerably up to 5–6 cm in length and filled out nearly the entire circumference of the intestinal lumen (Fig. 2). There was still no prospect of success with the endoscopic therapy approach and indication of explorative laparotomy was made.

During laparotomy, we saw that practically all the small bowel was invaginated into three parts, with long segments. The first one was on the proximal jejunum, induced by the known big 5x6cm polyp. The second intussusception was on the jejunoileal region caused by another polyp that measured 3x4 cm, the last invagination, on the terminal ileum, was the result of a similar polyp. All the invaginated segments showed no circulation issues and they were desinvaginated without any damage.

The biggest proximal jejunum polyp was excised with a segment resection (Fig. 3) and the polyp on the terminal ileum was also removed with segment resection.

For the jejunoileal polyp, an enterotomy was performed and a complete excision was achieved without any problems (Fig. 4).

The postoperative period was uneventful, and the patient was discharged eight days after the procedures. Histopathological investigations confirmed three Peutz-Jeghers polyps with hyperplasia of the mucosal glands. No malignancy was observed.

III. Results and discussion

Peutz-Jeghers syndrome is an autosomal dominant condition and clinical manifestations generally occur in adolescence or adulthood[1].

PJS is characterised by gastrointestinal polyps, mucocutaneous pigmentation and the increased risk of developing cancer. A physical examination is an important element for diagnosing PJS. The mucocutaneous pigmentations caused by melanin aggregation can be seen in 93% of

Vol. 3, No. 06; 2019

ISSN: 2581-3366

patients, even in infancy, and they are generally located around the lips, buccal, hand, foot and sometimes the perianal and genital areas [1-3].

Different radiological modalities are used to diagnose PJS, such as sonography, CT, MRI or contrast studies. Using these imaging techniques, the polyps can be visualised as well as the characteristic "target sign", with a nodular soft tissue mass in the centre thought to be a polyp and caused intussusceptions [1, 2].

Peutz-Jeghers patients should be under medical care and receive periodic follow-up examinations such as gastroduodenoscopy, colonoscopy and blood test to avoid complications of the disease [1, 3, 4].

In surgical procedures, combined endoscopic and surgical treatment are advocated. Classic symptoms of the Peutz-Jeghers syndrome are abdominal pain, mass and jam like stools [1-5].

The cause of these abdominal complaints is generally acute intussusception and the intussusceptions occurred mostly as a result polyps $\geq$ 15 mm in diameter in PJS patients [1].

The polyps, which induced intussusception and produced mechanical problems, were removed by laparotomy and resection. The procedure selected was based on the location and size of the polyps. Bowel viability is another important factor. Abdominal exploration, with conventional resection and end-to-end anastomosis of the involved segment, is usually recommended. Occasionally, a stoma creation may be required due to poor conditions in emergency situations.

In our patient, the proximal jejunal polyp induced the first episode of intussusception at the time of diagnosis, four years ago. During this time, he did not present with acute "surgical abdomen" and had no feeding problems. We can now speculate that recurrent intussusceptions, caused by the jejunal polyp, resolved spontaneously and did not cause ileus. Finally, because of worsening symptoms and signs of mechanical obstruction, the indication for surgery was made.

IV. Conclusion

In conclusion, Peutz-Jeghers syndrome is characterised by the association of gastrointestinal polyps and mucocutaneous pigmentation. Patients with PJS should be under close follow-up care via radiological investigations and endoscopy. The increased risk of invaginations and malignant changes should be considered.

The indication for the operation must be performed in a timely manner to avoid critical situations like perforation with peritonitis and disturbance of circulation in the intestine, which would result in long segment resections possibly leading to short bowel syndrome.

If a laparotomy is required, asymptomatic polyps should be sought and removed, in addition to the polyp responsible for obstruction.

Vol. 3, No. 06; 2019

ISSN: 2581-3366

References

Wang H, Luo T,Liu WQ, et al. Clinical Presentations and Surgical Approach of Acute Intussusception Caused by Peutz–Jeghers Syndrome in Adults. J Gastrointest Surg (2011) 15:2218–25.
DOI 10.1007/s11605-011-1724-2

Kalavan AB, Menon P, Mitra S, et al.Solitary Peutz Jeghers polyp of jejunum: A rare cause of childhood intussusception. JIAPS 2017;22(4): 245-7.

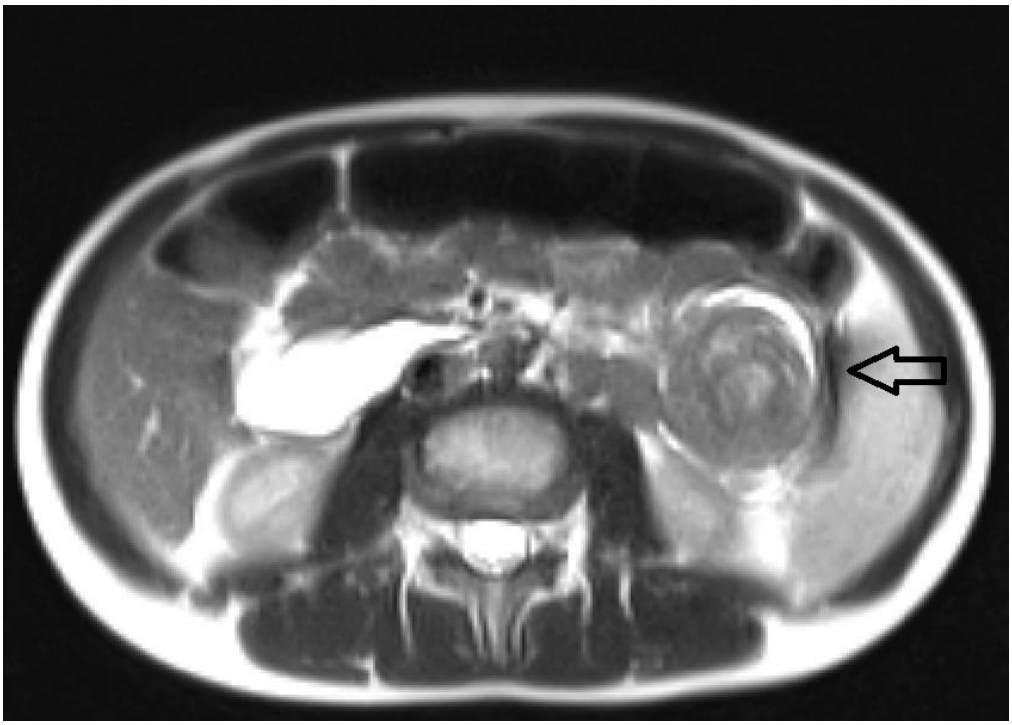
Kilic S, Atici A, Soyköse- Acikalin Ö. Peutz Jeghers syndrome: an unusual cause of recurrent intussusception in a 7 y/o boy. The Turkish Journal of Pediatrics 2016; 58: 535-7.

Boseto F, Shi E, Mitchell J, Preddy J. Gastroduodenal intussusception due to Peutz-Jeghers syndrome in infancy. Pediatr Surg Int 2002; 18: 178-80.

Tovar JA, Eizaguirre I, Albert A, et al. Peutz-Jeghers syndrome in children: report of two cases and review of the literature. J Pediatr Surg 1983;18(1):1-6.

Figures

Fig. 1: MRI revealed target sign of intussusception by proximal jejunum (arrow).



Vol. 3, No. 06; 2019

ISSN: 2581-3366

Fig.2: Endoscopic view of proximal jejunal polyp.

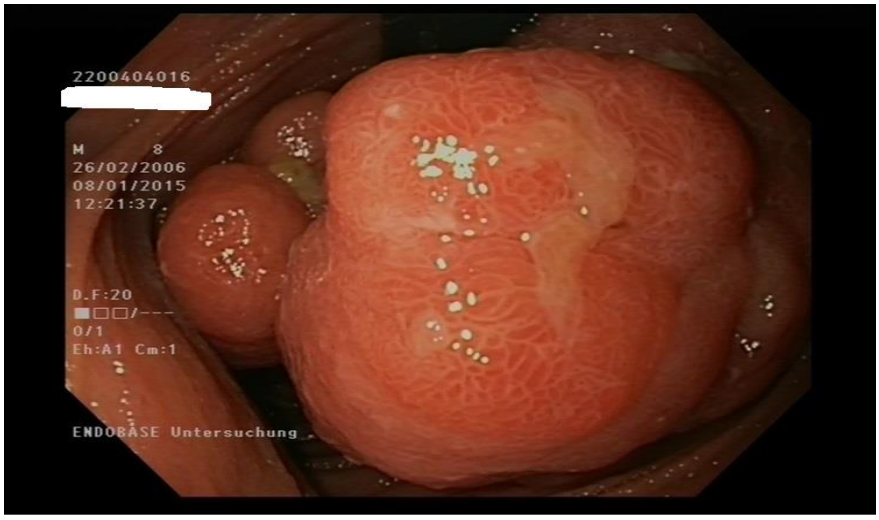


Fig.3: Proximal Jejunum segment resection (5X6cm polyp – arrow; intussusception).

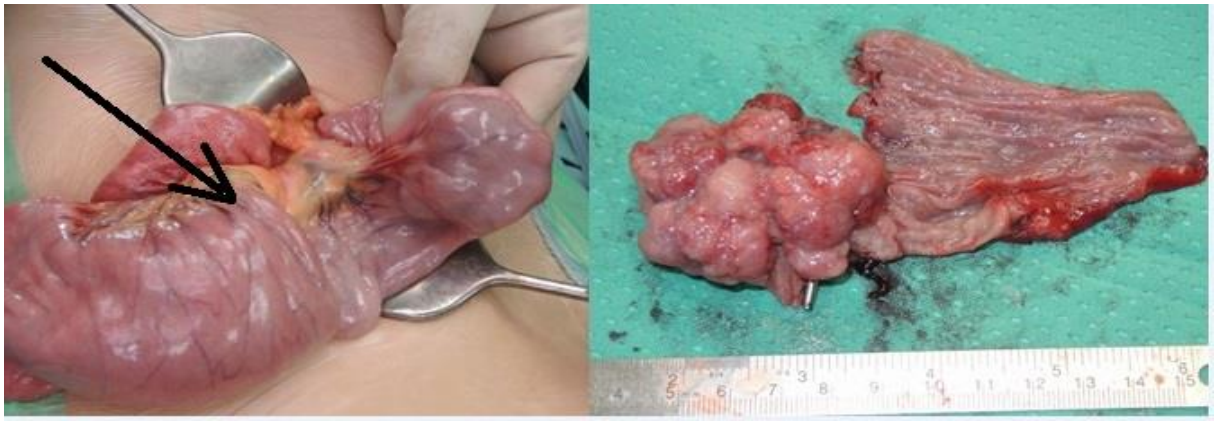


Fig.4: Polypectomy via enterotomy on jejuno-ileal region (arrow; intussusception).