

Small Intestine Intussusception in Adults: A Case Report

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Abstract:

Introduction: Small intestine intussusception in adults is a rare clinical event and should arouse a suspicion of genetic syndromes like Peutz Jeghers syndrome.

Case Report: We present a 28 year old man with recurrent episodes of pain abdomen with oral hyper pigmented patches. He was diagnosed to have multiple small intestine polyps with intussusception. He underwent an explorative laparotomy and resection of the involved jejunal segment due to impaired vascularity.

Conclusion: Peutz Jeghers syndrome is a rare cause of small intestine polyps and intussusception and obstruction especially in young adults.

Keywords: Peutz Jeghers syndrome, Intussusception.

I. Introduction

Adult intussusception is a rare entity and accounts for 1-5% of bowel obstruction. They are almost always associated with a pathological lead point. Small intestinal polyps are not as common as the large intestinal polyps. Such polyps in small intestine are associated rarely with congenital syndromes such as Peutz Jeghers syndrome.

II. Case Report

A 28-year-old male presented with periodic episodes of colicky abdominal pain and bilious vomiting since 1 week. On examination, he had pigmented macules over the lips and buccal mucosa (Figure 1). Abdominal examination during one of the episode of pain revealed palpable mass in the umbilical area. Ultrasound abdomen showed "target sign" and "hay fork sign" (Figure 2) involving the distal jejunum suggestive of intussusception. As Peutz-Jeghers syndrome was suspected, an Upper and lower gastro-intestinal endoscopy were done to look for polyps which were negative. In view of recurrent episodes of intestinal obstruction secondary to intussusception he underwent emergency laparotomy. During the surgery a long segment distal jejuno-jejunal intussusception was noted (Figure 3). On reduction of the intussusception an intraluminal polyp was identified as the lead point of the intussusception. (Figure 3). Multiple such polyps were noted along the entire length of jejunum. Segmental resection of jejunum (30cm) with an end to end jejunostomy was done due to doubtful viability of the involved segment. Resected specimen was cut open which revealed multiple polypoidal lesions (Figure 4). Histopathological examination of the polyps revealed smooth muscle hyperplasia, with an arborized, elongated pattern of polyp formation towards the epithelial layer this findings are consistent with Peutz-Jeghers polyps. Post-operative recovery of the patient was uneventful. He is on regular follow-up and is symptom free for the past one year. He and his family members were counselled about the nature of the disease and need for further evaluation for which they were not interested.



Figure 1: hyper pigmented patches along the mucocutaneous junction of lips

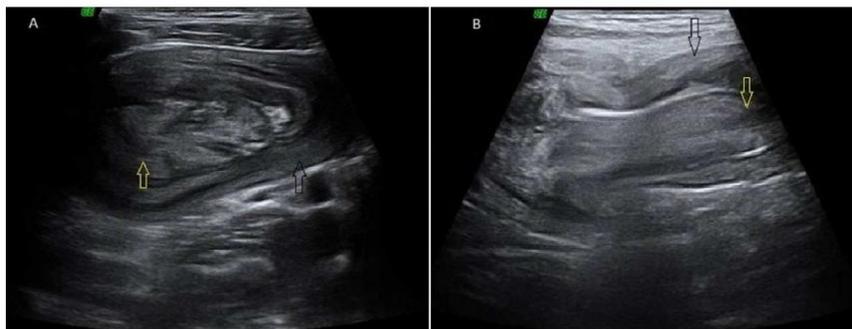


Figure 2: Ultrasound images showing target sign (A) and hay fork sign (B) (yellow arrow indicates intussusceptum and black arrow indicates intussusciptum).



Figure 3: Intraoperative image shows distal jejuno-jejunal intussusception (picture A) (yellow arrow indicates intussusceptum and black arrow indicates intussusciptum) and lead point shown by artery forceps (picture B) indicates intussusciptum.



Figure 4: Image showing multiple polyps in the resected segment of jejunum.

III. Discussion

Peutz Jeghers syndrome is an autosomal dominant genetic disease characterized by numerous hamartomatous polyps in the gastrointestinal tract and hyperpigmented lesions in the oral cavity [1]. In 1921 Jan Peutz, a Dutch physician described the relationship between mucocutaneous hyperpigmentation and gastrointestinal polyps in a Dutch family. In 1949, an American physician Herold Jeghers described in detail about the syndrome [2]. The molecular basis for the disease is found to be a germline mutation in the *STK11/LKB1* gene on chromosome 19p13.3 [3].

The Peutz Jeghers syndrome can be diagnosed based on few diagnostic criteria [4]:

- a) Three or more histologically confirmed Peutz-Jegher's polyps **or**
- b) Any number of Peutz-Jegher's polyps with a family history **or**
- c) Characteristic prominent mucocutaneous pigmentation with a family history **or**

d) Any number of Peutz-Jegher's polyps with characteristic prominent mucocutaneous pigmentation.

The hamartomatous polyps of Peutz-Jeghers syndrome are located most in the jejunum and ileum, but they can extend anywhere between stomach and rectum [3, 5]. These polyps can also be found in non-gastrointestinal sites such as nasal passages, bronchial tree, gallbladder, kidney and ureter [5]. These polyps can give rise to complications such as intestinal obstruction and intussusception, abdominal pain, haemorrhage and rectal prolapse [3]. They can also ulcerate causing an acute blood loss or chronic anaemia [5]. Apart from having hamartomatous polyps in GIT, patients with Peutz-Jeghers syndrome are more susceptible to develop gastrointestinal as well as various non-gastrointestinal malignancies such as breast, pancreas, lung, uterus, cervix, ovary thyroid and testes [6, 7].

Although Peutz-Jegher's polyps can be picked up by Ultrasound or CT scan, they are used usually to demonstrate complications such as intussusception. Ultrasound demonstrates a target sign in the presence of an intussusception. Upper and lower gastrointestinal endoscopy may show numerous polyps.

Standard therapy for patients with symptomatic polyps with obstruction or intussusception would be an explorative laparotomy with resection of the involved segment [5]. With the advent of technology laparotomy can be combined with intra-operative enteroscopy or double balloon enteroscopy for resection of the polyp so that multiple enterotomies are avoided and possible short bowel syndrome may be prevented [3, 6, 7]. Patient should be periodically assessed for recurrence of symptoms and also for screening for cancers.

In the present case, a diagnosis of Peutz-Jeghers syndrome was made based on the clinical findings of hyperpigmented lesions around the oral cavity and multiple polyps noted in the duodenum on gastroscopy. As patient had recurrent episodes of obstructive symptoms and ultrasound demonstrated an evidence of intussusception, laparotomy was planned. During the surgery, as bowel viability was doubtful, resection of the segment was done.

IV. Conclusion

Adult intussusception is a rare clinical event and it is almost always associated with a lead point. Intussusception in young adults should always arouse a suspicion of congenital abnormalities like Peutz-Jeghers syndrome. A detailed clinical examination and investigations can guide proper treatment and avoid unnecessary surgeries.

References

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