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Case Report

Recurrent Intussusception Secondary to Multiple Peutz-jegher polyps: An Unusual Case with Review of Literature

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ABSTRACT
Peutz-Jegher polyps are hamartomas of the small bowel which occur as part of the Peutz-Jegher syndrome, a rare autosomal dominant condition characterized by gastrointestinal polyps and mucocutaneous melanotic pigmentation. Here we report the case of a young male suffering from this syndrome who presented with acute intestinal obstruction due to intussception. Exploratory laparotomy was done which revealed jejuno-jejunal and jejuno-ileal intussusceptions for which resection and anastomosis was done. There were multiple intraluminal polyps in the jejunum which on histopathological examination, showed features of Peutz-Jegher Polyp. Patient behaved well in the post-operative period and is still on follow up with us.

Key words: Hamartoma, Intussusception, Intestinal obstruction, Polyps, Peutz-jegher syndrome.

INTRODUCTION

Peutz-Jegher Syndrome is an autosomal dominant condition characterized by the combination of hamartomatous polyps of the intestinal tract and hyperpigmentation of the buccal mucosa, lips and digits. The polyps may cause bleeding or intestinal obstruction (from intussusception). While the hamartomatous polyps themselves only have a small malignant potential, patients with this syndrome are at greater risk of developing carcinomas of the pancreas, liver, lungs, breast, ovaries, uterus, testicles and other organs.[1] Here we report a case of multiple Peutz-Jegher polyps leading to intussusception and presenting with small bowel obstruction as an emergency. Such a presentation is rare in the Indian subcontinent and due to scarcity of literature on Peutz-Jegher polyps/syndrome in India we felt reporting this case may be a noteworthy contribution.

CASE REPORT

A 15 years old, male presented to the emergency department with complains of colicky abdominal pain and bilious vomiting along with history of abdominal distention and non-passage of flatus and stools for two days. There was a past history of surgery for similar complaints at the age of nine years. Records of the previous illness were not available but the parents were well aware that the child had intussusception which was surgically corrected. Since then the child had been asymptomatic. On physical examination, there were pigmented lesions on the lips and tongue (Figure 1). Abdomen was distended with tenderness in the lower abdomen. There was a mobile, tender mass palpable in the hypogastrium measuring 7 × 4 cm with smooth surface and well-defined margins. The mass was resonant on percussion. Per rectal examination was normal. Ultrasonography of the abdomen showed telescoping of gut at multiple levels suggestive of multiple intussusceptions. The patient underwent an emergency exploratory laparotomy after resuscitation with intravenous fluids and antibiotics. Intra operative findings revealed two small bowel intussusceptions (jejuno-jejunal and jejuno-ileal) with gangrene of the gut loops in the jejuno-ileal intussusception (Figure 2). We reduced the jejuno-jejunal instussusception and resected the gangrenous distal segment with end to end jejuno-ileal anastamosis. On careful examination of the proximal bowel, there were multiple palpable polypoidal lesions present intraluminally in a segment of jejunum which was also resected and after enterotomy of this segment, multiple polyps were noticed (Figure 3). Post-operatively the patient recovered well and was discharged on 6th post-operative day. Histopathological examination of the resected bowel segment confirmed presence of multiple hamartomatous jejunal polyps (Figure 4).

DISCUSSION

Peutz-Jeghers Syndrome (PJS) is rare autosomal dominant disease characterized by hamartomatous polyps in gastrointestinal tracts and mucocutaneous

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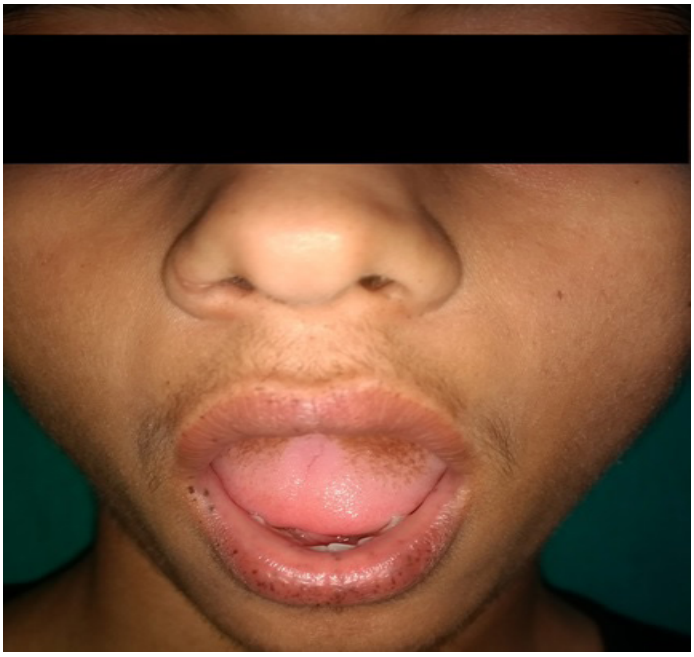

Figure 1: Clinical photograph showing pigmented lesions on the lips and tongue.

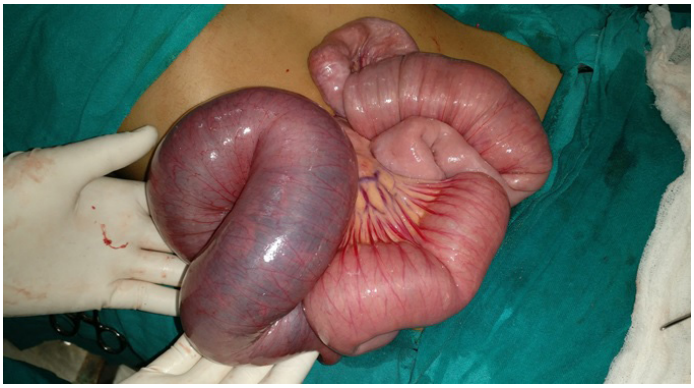

Figure 2: Intra Operative image showing two intussusceptions with gangrenous changes in the distal segment.

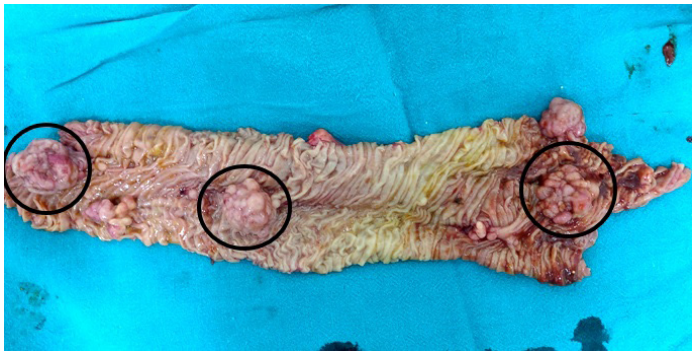

Figure 3: Resected segment of jejunum showing multiple polyps.

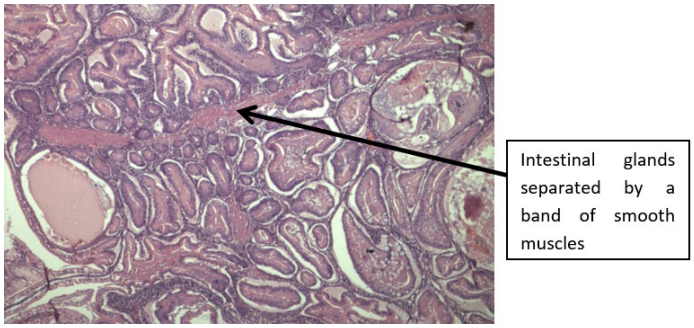

Figure 4: Microscopic view of the Hamartomatous Polyp (H & E, 40x).

pigmentation. It was first reported by Peutz in 1921 and the definitive clinical description of syndrome was written by Jeghers in 1949. The prevalence of PJS differs between studies, estimated incidence has been noted from 1 in 8300 to 1 in 120000 births, and both sexes are almost equally affected.[2] The cause of Peutz-Jeghers syndrome (PJS) in most cases (66-94%) appears to be a germ line mutation of the STK11/LKB1 (Serine/Threonine Kinase 11) tumor suppressor gene, located on band 19p13.33.[1]

Clinical picture of the syndrome includes two major components: mucocutaneous pigmentation and hamartomatous polyposis of the intestinal tract. The pigmented lesions are often seen on the lips, around

Table 1: Surveillance guidelines (American College of Gastroenterology)[9]

Site	Age to begin surveillance (years)	Surveillance interval (years)	Surveillance procedure and comments
Colon	8, 18[a]	3	Colonoscopy[a]
Stomach	8, 18[a]	3	Esophagogastroduodenoscopy[a]
Small Bowel	8, 18[a]	3	Video capsule endosopy[a]
Pancreas	30	1-2	MRCP or Endoscopic ultrasound
Breast	25	1	Annual self-exam starting age 18, annual breast MRI, and/or mammogram starting at age 25
Ovarian	25	1	Pelvic exam and pelvic or transvaginal ultrasound, CA-125 probably not helpful
Endometrial	25	1	Pelvic exam and pelvic or transvaginal ultrasound
Cervix	25	1	Pap smear
Testicular	Birth to teenage years	1	Testicular exam, ultrasound if abnormalities palpated or if feminization occurs; 10 to 20% of benign Sertoli cell tumors become malignant
Lung	-	-	Provide education about symptoms and smoking cessation.

[a]Start at age 8 years; if polyps present, repeat every 3 years; if no polyps, repeat at age 18, then every 3 years, or earlier if symptoms occur.

the mouth, eyes, nostrils, on the buccal mucosa; and sparsely on the fingers, soles of the feet, palms, anal area and intestinal mucosa. Gastrointestinal polyps are most commonly found in small intestine, but can occur anywhere from the stomach to rectum. PJ polyps may lead to intussusception, some of which may spontaneously reduce while others lead to obstruction. Peutz-Jeghers polyps can also ulcerate, leading to acute blood loss or chronic anemia.[3] Fewer than 5% of patients with PJS lack the abnormal mucocutaneous pigmentation, and fewer than 5% of patients with the pigmentation have no PJ polyps.[4] The diagnosis is more likely possible in pediatric patients as pigmented lesions may fade at puberty. In a case series reported by Suda *et al*., solitary Peutz-Jeghers type hamartomatous polyps presented without pigmented spots in 87% cases, involved only one organ in the gastrointestinal tract, and showed a predilection for males in 74% cases.[5]

The diagnostic criteria for Peutz-Jeghers syndrome proposed by the Johns Hopkins Registry include histopathologically verified hamartomatous polyps with at least two of the following: Small-bowel location for polyposis, mucocutaneous melanotic pigmentation, and a family history of Peutz-Jeghers syndrome.[6] Our patient had histopathologically verified hamartomatous polyps with two of the additional criteria i.e. small bowel polyposis and mucocutaneous pigmentation.

The evolution of PJS during life exposes the patient to a predisposition to developing malignant neoplasias, mainly in the small bowel, caecum, rectum, as well as pancreatic adenocarcinomas. Other more rare carcinomas could occur in the breast, uterus, testicles and ovaries. [1] The risk for cancer in these patients is 18 times higher in comparison with the general population, and higher the number of intestinal polyps higher is the chance for malignant transformation in PJS.[7]

As the polyps can develop at any region in the entire GI tract, their recurrence is quite common, making it a challenge for the surgeon to decide regarding the amount of resections that should be done. Short-bowel syndrome is a well-known complication of repeated resections making the patient quite unstable due to malnourishment.[8] The goal of treatment is to clear as much polyp as enough, and to avoid resections. Consequently, minimally invasive approaches are recommended and some of the authors advocate both the surgical and endoscopic techniques at the same session that is called "clean sweep" technique, in which the surgery team examines the whole intestinal tract with gastroduodenoscopy, enteroscopy, push endoscopy and remove/destroy the smaller polyps to achieve a polyp free intestinal mucosa. This technique reduces the further need for abdominal surgery and complications.[1] Patients with Peutz-Jeghers Syndrome should be educated about the symptoms of intestinal obstruction and the need for cancer surveillance.

Table 1 lists the surveillance guidelines as recommended by American College of Gastroenterology.[9]

CONCLUSION

Intussusception as the cause for intestinal obstruction is a frequent finding in pediatric age group but intussusception due to Peutz-Jeghers polyps is a rare presentation. We would like to emphasize that in cases of intussusception, careful examination of the bowel is recommended to look for presence of intestinal polyps and the possibility of Peutz-Jeghers Polyp/Syndrome. A lifelong regular follow-up is advised to these patients once the diagnosis of PJS is made.

REFERENCES

1. Sreeramulu PN, Agrawal VP. A rare case of multiple intussusceptions in Peutz-Jeghers syndrome. J Med Allied Sci. 2013;3(1):26-8.
2. Sekmenli T, Afsarlar CE, Bakir S. An Unusual Presentation of Peutz-Jeghers Syndrome: A Case of Recurrent Jejunal Intussusception. Eur J Gen Med. 2012;9(1):52-5.
3. Parikh SB, Parikh BJ, Prajapati HA, Shah CK, Shah NR. Peutz-Jeghers Syndrome. Guj Med J. 2013;68(2):106-8.
4. Ozer A, Sarkut P, Ozturk E, Yilmazlar T. Jejunoduodenal intussusception caused by a solitary polyp in a woman with Peutz-Jeghers syndrome: a case report. Journal of Medical Case Reports. 2014:8(1):13.
5. Agarwal D, Saluja SK, Parihar D, Kundu P. Solitary Peutz-Jeghers Polyp of Jejunum with Dysplasia Presenting as Intussusception. NJLM. 2015;4(4):47-49.
6. Bingi TC, Teja VP, Ramulu B, Manjula B, Vemula R. Peutz-Jegher's Syndrome Presenting as Multiple Intussusceptions: A Case Report. Indian Journal of Applied Research. 2015;5(3):462-4.
7. Islam MR, Hossain MS, Sheikh MSH, Rahman MR, Lima IJ, Khatun SA *et al*. Peutz-jeghers syndrome: a case report and literature review. J Dhaka Med Coll. 2014;23(1):147-150.
8. Kumar A, Shetty P, Rao AB, Ramachandra. Peutz-Jeghers syndrome presenting with small bowel obstruction as an emergency. The Internet Journal of Surgery. 2007;16(2).
9. Syngal S, Brand RE, Church JM, Giardiello FM, Hampel HL, Burt RW. ACG Clinical Guideline: Genetic Testing and Management of Hereditary Gastrointestinal Cancer Syndromes. Am J Gastroenterol. 2015;110:223-62.

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