Case Report

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A Rare Cause of Intestinal Obstruction in Paediatric Patients

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Abstract

Peutz-Jeghers syndrome is an autosomal dominant hamartomatous polyposis involving gastrointestinal tract associated with mucocutaneous hyperpigmentation involving lips, buccal mucosa ,hand and feet and predisposing the patient to various extraintestinal and intestinal cancers.This article includes case report of a 12 year female diagnosed with intussusception due to Peutz-Jeghers-Syndrome. This patient underwent emergency exploratory laparotomy followed by resection of about 30 cm necrosed , gangrenous ileum about 50cm proximal to ileocecal junction and then ileoileal anastomosis performed . This patient had pigmented hypermelanotic macules over lower lip ,buccal mucosa and face. This case was thoroughly examined and found to be a rare case of intestinal obstruction in paediatric patients.

Key words: Peutz Jeghers syndrome; intussusception; hamartomatous polyposis.

Introduction

Peutz-Jeghers Syndrome is an autosomal dominant disease with multiple hamartomatous intestinal polyps associated with melanosis of oral mucosa, lower lip and occasionally digits(not in tongue). It is associated with germline defect in STK-11 GENE/LKB-1 GENE present in chromosome-19. Most common site of involvement : small intestine(jejunum)> colon> stomach. There are hypermelanotic macules over lips and buccal mucosa. Recurrent intussusceptions is seen which is responsible for recurrent abdominal pain. There is increased risk of malignancies of small intestine(duodenal adenocarcinoma), pancreas (maximum risk), breast,thyroid ,endometrium and sertoli–cell tumour. Histologically polyps shows arborising pattern . Patient most commonly presents with signs and symptoms of intestinal obstruction.

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Case History

A 12 year old female patient presented to paediatric-opd with complaint of pain in epigastric region region since 2 days associated with vomiting since 2 days and giddiness and patient is known case of epilepsy with 1st episode of epilepsy at 1.5 years of age and 2nd episode at 8years of age and then patient was admitted in pediatric ward on 17/11/2020 with blood investigations having Hb-3.6gm%, WBC-5200/mm3, Platelet- 4.98 lacs /mm3 with normal renal function test and liver function test. Patient was reffered to surgical unit for the same on 23/11/2020. Radiological investigations were done and CECT-Abdomen suggestive of multiple intussusceptions with a long-segment intussusception at mid-ileal site with segment length of 17cm with a proximal jejunal intussusception

in left hypochondriac region with segment length of 7cm with a mid-jejunal intussusception in left hypochondriac region with segment length of 3 cm with polypoidal lesions in body and pylorus of stomach, duodenum, and small bowel loops with largest polyp of size(2.1 x 2.2)cm in mid jejunal loop and then emergency exploratory laparotomy was done through a midline vertical incision followed by identification of ileoileal intussusception where lead point of intussusceptions was found to be a (2x1) cm polyp followed by resection of approximately 30cm length of necrosed, edematous and gangrenous ileum about 50cm proximal to ileocecal junction followed by ileoileal anastomosis and identification of multiple polyps from stomach to ileocecal junction.

Intussusception part of bowel(ileum) could not be reduced manually and hence necrosed part was resected and ileoileal anastomosis done followed by Morrison and pelvic drain placement and fixation followed by mass closure.

Patient was then stabilized in Recovery room and then shifted to PICU for further post-operative care and management and daily rounds were taken from surgical side.On postoperative day 5,7 and 9 wound seen under all aseptic precautions and on post-operative day 10 wound seen and alternate SR was done and pelvic drain removed. On day 13 All SR was done and Morrison drain removed and patient was discharged on post-operative day 15.

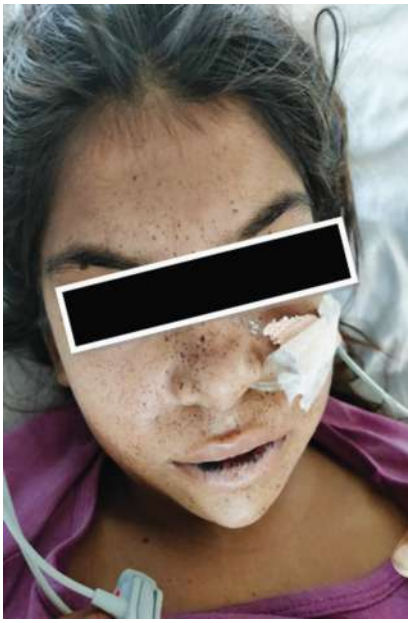

Fig. 1: Hypermelanoticmacules over face and lower lip.

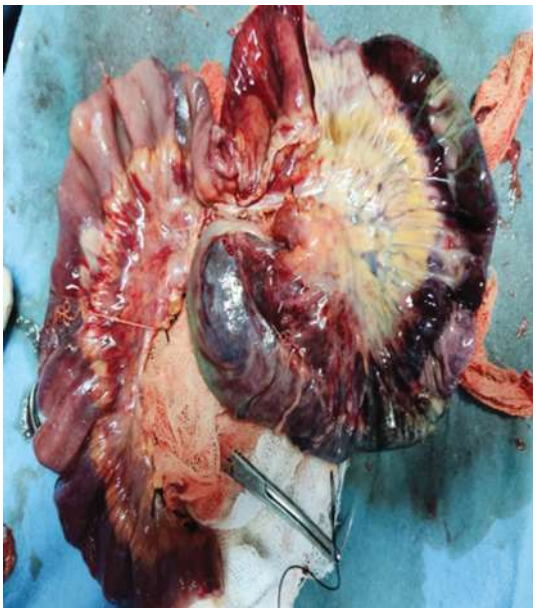

Fig. 2 : Gangrenous part of ileum of approximately 30 cm length with ileoileal intussusception with its polypoidal lead point.

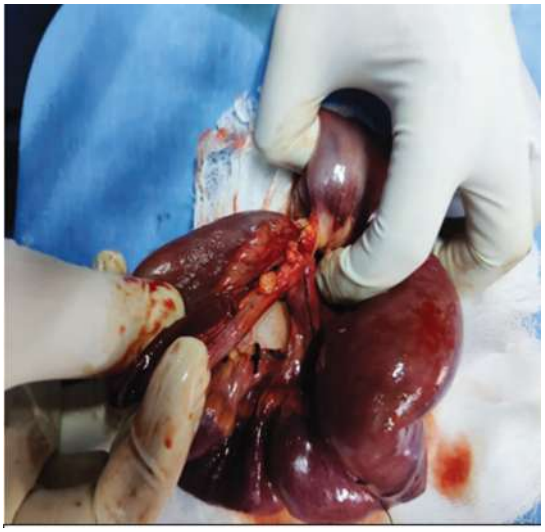

Fig. 3: Resected part of ileum with Necrosis.

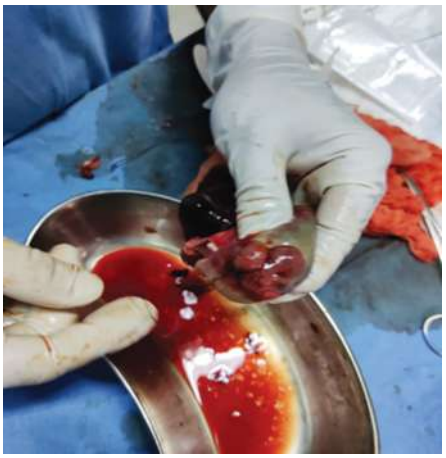

Fig. 4: Lead point of ileoilealintussusception showing polyp.

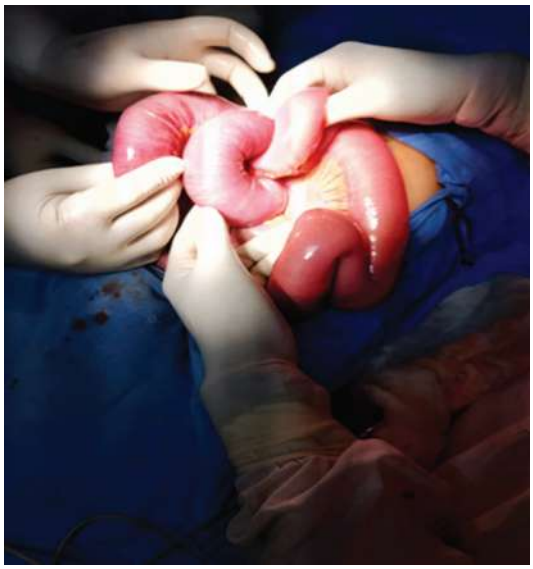

Fig. 5: Intraoperative finding of dilated proximal ileal loop due to distal obstruction due to ileoileal intussusception.

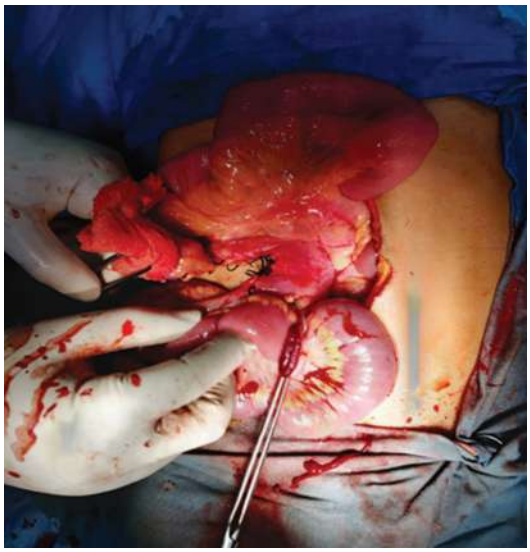

Fig. 6: Showing healthy part of Bowel after resection prepared for ileoileal anastomosis.

Discussion

Hyperpigmentation is present as mucocutaneousmacules on the lips and around the mouth, eyes, nostrils, and on the buccal mucosa; and sparsely on the fingers, soles of the feet, palms, anal area and intestinal mucosa. Characteristic pigmentations are present in 95% of the patients and are caused by pigment-laden macrophages in the dermis. They are typically flat, blue-gray to brown spots 1-5 mm in size. Malignant degeneration of these lesions is extremely rare. These macules can be distinguished from common freckles as the latter never appear in the oral cavity, are sparse near the lips and nostrils, and absent at birth. Hyperpigmentations can even disappear during adolescence. Diagnosis is defined by the presence of histopathologically confirmed hamartomatous polyps and at least two of the following clinical criteria: family history, hyperpigmentation and polyps in the small bowel. The median time to first presentation with polyps is about 11-13 years of age, and approximately 50% will have experienced symptoms by the age of 20 years. During the first three decades of life, anemia, rectal bleeding, abdominal pain, obstruction and/or intussusception are common complications in patients with PJS. Nearly half of the patients experience an intussusception during their lifetime, most often in the small intestine.Extraintestinal polyps are also reported. Nasal polyposis is thought to be a rare complication.

Conclusion

In summary, timely polypectomy, preferably using the double balloon enteroscopy (DBE)method, are essential for patients with PJS and prevents polyp related intussusceptions and intestinal obstruction. Follow-up of gastrointestinal polyps is necessary. The best combination of methods is capsule enteroscopy and MR enteroclysis. Lifelong screening of malignancies is indispensable on a regular basis. It is necessary to investigate all first-degree relatives of the patient. Although the incidence of PJS is low, it is important for clinicians to recognize these disorders to prevent morbidity and mortality in these patients, and to perform presymptomatic testing in patients at risk.There are two basic modalities in diagnosis and treatment of small bowel hamartomas: intra-operative enteroscopy (IOE) and double balloon enteroscopy (DBE). DBE is a new enteroscopy method that allows examination and treatment of the jejunum and ileum in almost all patients. The system consists of a 200cm enteroscope and a 145cm over-tube which have soft latex balloons at their tips. By using these balloons to grip the intestinal wall, the endoscope can be inserted further without forming redundant loops of intestine. IOE is a combination of laparotomy (or laparoscopy) with endoscopy. It allows manipulation to ensure the entire small bowel is visualized and nearly all polyps are removed in an endoscopic or surgical manner.

References

1. Connor JT. Aesculapian society of London. Lancet. 1895;2:1169.
2. Hutchinson J. Pigmentation of lips and mouth. Arch Surg. 1896;7:290.

3. Jeghers H, McKusick VA, Katz KH. Generalized intestinal polyposis and melanin spots of the oral mucosa, lips and digits; a syndrome of diagnostic significance. N Engl J Med. 1949;241:1031–1036.
4. Weber FP. Patches of deep pigmentation of oral mucous membrane not connected with Adison's disease. Quart J Med. 1919;12:404.
5. Peutz JLA. Over eenzeermerkwaardige, ge combineerdefamiliairepollyposis van de sligmliezen van den tractusintestinalis met die van de neuskeelholte en gepaard met eigenaardigepigmentaties van huid-en slijmvliezen (Very remarkable case of familial polyposis of mucous membrane of intestinal tract and nasopharynx accompanied by peculiar pigmentations of skin and mucous membrane; in Dutch) NederlMaandschr v Geneesk. 1921;10:134–146.
6. Westerman AM, Entius MM, de Baar E, Boor PP, Koole R, van Velthuysen ML, Offerhaus GJ, Lindhout D, de Rooij FW, Wilson JH. Peutz-Jeghers syndrome: 78-year follow-up of the original family. Lancet. 1999;353:1211–1215.
7. Foley TR, McGarrity TJ, Abt AB. Peutz-Jeghers syndrome: a clinicopathologic survey of the "Harrisburg family" with a 49-year follow-up. Gastroenterology. 1988;95:1535–1540.
8. Bruwer A, Bargen JA, Kierland RR. Surface pigmentation and generalized intestinal polyposis; (Peutz-JegherssUse of uninitialized value in scalar chomp at E:\wwwroot\cgi-bin\submit.pl line 49. yndrome) Proc Staff Meet Mayo Clin. 1954;29:168–171.
9. Horrilleno EG, Eckert C, Ackerman LV. Polyps of the rectum and colon in children. Cancer. 1957;10:1210–1220.
10. Calva D, Howe JR. Hamartomatouspolyposis syndromes. SurgClin North Am. 2008;88:779–817, vii.