

## “Intussusception Due To Solitary Hamartomatous Peutz-Jeghers Type Polyp –A Rare Case Report”

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**Abstract:-** Intussusception is a rare cause of the intestinal obstruction in adults and it represents a diagnostic challenge for the surgeon. In majority of cases, presenting symptoms are not specific, making preoperative diagnosis very difficult. We present a case of a 17-year-old male patient with intestinal obstruction as a result of intussusception due to ileal (hamartomatous) polyp. For which diagnostic laparoscopy followed by exploratory laparotomy in which resection of that solitary polyp containing gut loop was done and side-to-side anastomosis was done. Postoperatively patient went into ileus but managed conservatively and then discharge satisfactory. This is an extremely rare case in which the first manifestation of the intestinal polyp was ileo-ileal intussusception. Prophylactic surveillance i.e. timely gastrointestinal tract investigations, screening of at-risk individuals and their family members, genetic testing, and lifelong patient follow-up, are recommended.

**Keywords:-** Intussusception – Peutz-Jeghers syndrome – Intestinal obstruction – Polyp

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### I. Introduction:-

Intussusception is defined as the condition in which part of the intestine slides with its mesenteric fold in the lumen of the adjacent distal bowel. This process impairs peristalsis, obstructs the free passage of the intestinal contents, compromises the mesenteric vascular flow of this part of the bowel and finally results in intestinal obstruction and inflammation of the bowel wall. Intussusception represents an uncommon cause of intestinal obstruction in adults, with a reported incidence of 1–5%. The ratio of adults to children is 1 : 20. (1) Unlike in children, in adults only 10% of intestinal intussusceptions are idiopathic, whereas in the remaining 90% a demonstrable cause is revealed. (2) In such cases, intussusception is associated with a lead point lesion, which may be a benign (adhesions, adenomatous polyp, Meckel’s diverticulum, hamartomatous polyp, lipoma, endometriosis, gastrointestinal stromal tumor, haemangioma, neurofibroma, tuberculosis) or a malignant mass (adenocarcinoma, carcinoid tumor, leiomyosarcoma, lymphoma, metastatic carcinoma). (1) Malignant lesions are responsible for up to 30% of cases of intussusception in the small bowel, whereas in the colon malignant etiology is found in up to 66% of cases. (2) The most seen types of intussusception are jejunojejunal, jejunoileal, ileoileal and ileocolic. Peutz-Jeghers syndrome is an autosomal dominant disorder, with an incidence of 1 in 150 000 people. It is characterized by a family history of PJS, mucocutaneous pigmentation and hamartomatous polyposis of the gastrointestinal tract (3).

### II. Case Report:-

A 17 year old male present to our surgery department with c/o pain in abdomen and vomiting since 6 days. Pain was colicky in nature was located in left upper quadrant mainly in left hypochondrium and left lumbar region, mild in intensity, occur on and off, non-radiating in nature, pain became stronger after eating or drinking anything, and vomiting was found to relieve the pain for a while, vomiting occur as soon as he eats or drinks something, patient also c/o not passing flatus and motion since 6 days. On per abdomen examination - abdomen was soft, mild distension was present, not tenderness, no guarding, and no rigidity. On per-rectal examination - anal tone normal, no ballooning, no fecolith was present. On local examination mucocutaneous pigmentation was noted on her lips and oral mucosa. Patient admitted to surgery department for further evaluation and investigation



**Figure 1 Labial hyperpigmentation**



**Figure 2 Buccal mucosa pigmentation**

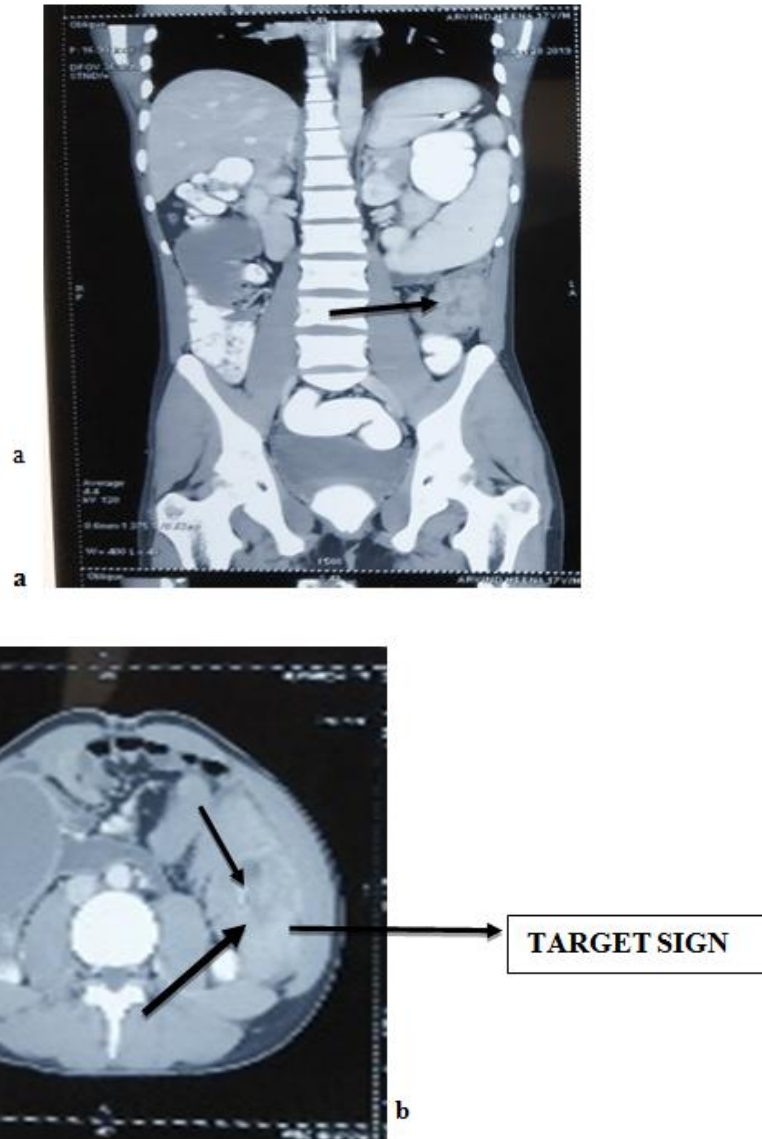


**Figure 3 X-ray FPA shows:- dilated intestinal loops with multiple air fluid levels.**

**USG w/a-**dilated bowel loops with internal bowel diameter 4-5cm

**CT shows:-** An area of bowel within bowel appearance seen in left side of abdomen suggestive of intussusception. There is suggestion of protrusion of proximal ileal loops within the proximal/mid ileal loops. There is moderate dilatation of jejunal and proximal ileal loops with maximum luminal diameter is app..6cm.

**Opinion-** Suggestive of intussusception (ileo-ileal type) with moderate dilatation of jejunal and ileal loops. Mesenteric lymphadenopathy.



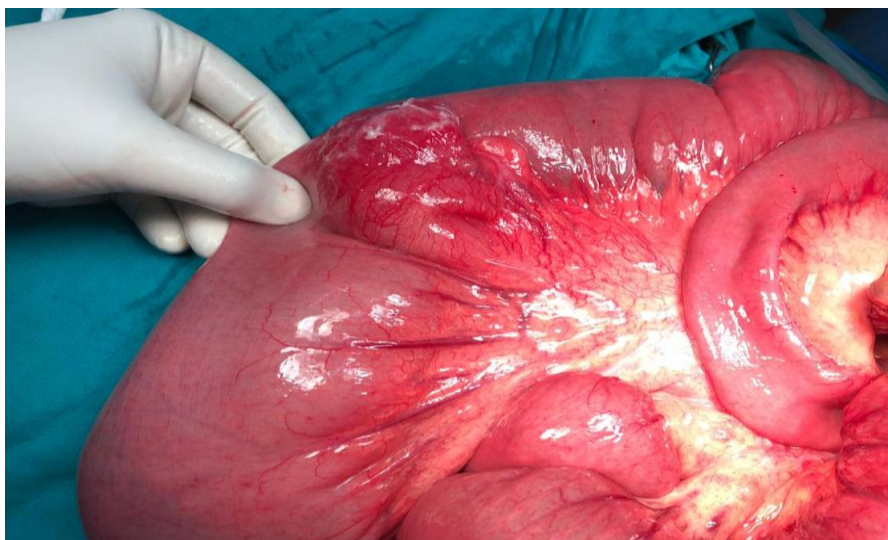
**Figure 4. (a)** Axial CT image oriented parallel to the longitudinal plane of the intussusception demonstrates a heterogeneous “sausage” mass with central low-attenuation fat (arrow), which is being dragged into the intussusciens by the intussusceptum. **(b)** Axial CT image clearly demonstrates the intussusciens (thick black arrow), intussusceptum (thin black arrow), and vessels (black arrows) within the invaginated mesenteric fat. Decision taken to start with diagnostic laparoscopy to find out any other pathology, on laparoscopy we found intussusception at ileo-ileal part of intestine.



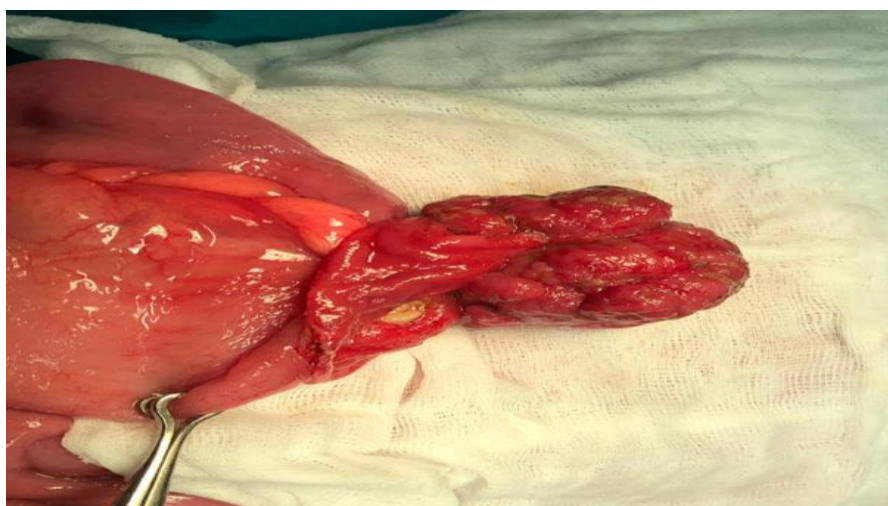
**Figure 5-** laparoscopic view shows ileo-ileal intussusceptions

So decision taken to convert it into open laparotomy, we found intussusception at ileo-ileal level which was reduced manually, we found a growth at 40cm proximal to ileo-caecal junction which leads to formation of intussusception, proximal to this segment ileal and jejunal loops were found to be dilated around 9-10cm and distal to this segment bowel loops were collapsed, so resection of that solitary polyp containing gut loop was done and side-to-side anastomosis was done. We subsequently made a thorough examination of the gastrointestinal tract i.e. from DJ to IC junction and also large bowel, no other masses were palpated in the remaining parts of gastrointestinal tract.

Post operatively patient went into ileus but managed conservatively and discharge satisfactory on post op day 7



**Figure 6.** Intraoperative photograph shows the transition zone of the intussusception, Which was caused by a lead-point Peutz-Jeghers polyp.



**Figure 7-** shows a large polyp of size 5\*4.5 cm.





**Figure 8-** Cut section of polyp

**Histopathology Report:-**Gross pathological findings revealed pedunculated polyp of size 5\*4.5\*4.0 cm. Histomorphology comprising of cores of finely arborizing branches of muscularis mucosa covered with bland columnar intestinal epithelium showing disorganized architecture. The histomorphology favours hamartomatous polyp, highly suspicious of Peutz-Jegher’s syndrome.

Given these findings, a detailed medical history was obtained and a comprehensive clinical examination undertaken, which failed to reveal any relevant family history. There was no family history of cancers in the GI tract or elsewhere. Consequently, the patient is currently under gastrointestinal evaluation and genetic testing.

### **III. Discussion:-**

Peutz-Jeghers syndrome is a rare autosomal dominant inherited diseases, which are also known as hereditary intestinal polyposis syndrome. It is characterized by hamartomatous polyps in the gastrointestinal tract (GIT) with hyper pigmented macules on the lips and oral mucosa. It has an incidence of about 1 in 300,000 births[4].The cause has been proven to be a germline mutation on the serine/threonine kinase 11 (STK11/LKB1) tumor suppressor gene on chromosome 19p13.3 in most cases (70-80%). The World Health Organization has laid down the following diagnostic criteria for Peutz–Jeghers syndrome:

- three or more histologically confirmed Peutz–Jeghers polyps
- any number of Peutz–Jeghers polyps with family history of Peutz–Jeghers syndrome
- have a characteristic mucocutaneous pigmentation with a family history of Peutz–Jeghers syndrome
- any number of Peutz–Jeghers polyps and characteristic mucocutaneous pigmentation.(5)

These patients are more probable to develop various GIT tumors, which includes predominantly small intestine, stomach, pancreas, colon, and esophagus. [6, 7] These patients have typical mucocutaneous pigmentation and melanin spots, which take place predominantly in the oral cavity, lips, nostrils, perianal area and digits. Pigmentation of the oral mucosa is pathognomonic of Peutz-Jeghers syndrome and is not associated with other types of dermatologies pigmented lesions. [8,9] Peutz–Jeghers polyp is an unusual type of hamartomatous polyp, which is microscopically characterized by tree-like branching of smooth muscle fibers, with a core of smooth muscle arising from the muscularis mucosa and extending into the polyp, covered by mucosal tissue with near-normal appearance.

These patients are to be screened regularly for:-[10]

- ❖ Small intestine with small bowel radiography every 2 years
- ❖ Esophagogastroduodenoscopy and colonoscopy every 2 years for colorectal carcinoma
- ❖ Computed tomography scan or magnetic resonance imaging of the pancreas yearly for pancreatic carcinoma
- ❖ Ultrasound of the pelvis (women) and testes (men) every year for genital tumor
- ❖ Mammography (women) from the age of 25 years and continued till life long
- ❖ Papanicolaou test every year.

Although computed tomography of the abdomen is the gold standard in diagnosing intussusception, with an accuracy of 85–100%.

The patients are to be educated about the regular follow up as these are prone to develop carcinomas in later life. [10, 11]

Resection of the intestinal polyps is done only when it complicates in from of serious bleeding or intussusception. [12]

#### **IV. Conclusion:-**

- ❖ Parents must be counseled intimately about the inheritance and the possibility of disease involvement in the newborn.
- ❖ The importance of regular follow-up should be explained in detail to the parents and the patient with the follow-up plan of investigations required should be excused.
- ❖ The genetic analysis of the infant should be exercised as it is a confirmatory diagnostic test and other manifestationstake time for a demonstration.
- ❖ Ileal intussusception is a difficult situation because of its infrequency and nonspecific clinic presentation. Diagnosis is usually missed or delayed because of the patient’s nonspecific, chronic and recurrent symptoms.
- ❖ A thorough review of the patient’s history, physical examination, radiological and endoscopic findings are critical in the case of ileo-ileal intussusception which is a rare form of an uncommon presentation.
- ❖ Surgeons treating patients with acute abdomen should always bear in mind that the presenting symptoms of intussusception are non-specific, thus making preoperative diagnosis difficult and in most cases delayed, leading to ischemia and necrosis.
- ❖ In adult cases,the presence of underlying pathology usually necessitates resection rather than reduction. A solitary PJ polyp though it’s very rare should be kept in mind as a differential diagnosis causing small bowel intussusception.

#### **Conflict of interest disclosure:-**

All the authors state that there are no commercial financial incentives related with publishing current work.

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