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Case Report

Acute intestinal obstruction in Peutz Jeghers syndrome: a case report

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ABSTRACT

Peutz Jeghers syndrome is an autosomal dominant hereditary disorder affecting male and female equally. It is characterised by mucocutaneous hyperpigmentation and hamartomatous polyp in gastrointestinal tract with increased risk of malignancy. We report here a case of 52-year-old patient with traits of Peutz jeghers syndrome presented with acute intestinal obstruction following colocolic intussusception. Peutz jeghers syndrome is an autosomal dominant inherited disorder. Individual may present in rare case with acute intestinal obstruction associated with intussusception due to polyps.

Keywords: Peutz Jeghers syndrome, Autosomal dominant, Malignancy, Intussusception

INTRODUCTION

Peutz Jeghers syndrome (PJS) is an autosomal dominant disease caused by germline mutation of the serine threonine kinase 11 and characterized by hamartomatous polyps in the gastrointestinal tract and mucocutaneous pigmentation. Patients with PJS have an increasing risk of developing cancer or transformation to malignant polyps in the gastrointestinal tract and other organs.

Colonic intussusception is an uncommon entity. Intussusception account for 1% of bowel obstruction and symptoms of complete obstruction occur in fewer than 20% of cases, and adult patient rarely present with signs of acute obstruction.

CASE REPORT

52-year-old gentlemen, with no family history of malignancy presented to emergency department of our hospital in January 2020.

He presented with history of colicky abdominal pain and gradual abdominal distension for last 20 days and obstipation for the same duration. Pain was aggravating after having meal and associated with on and off bilious vomiting. He had history of Melena for 5 days, weight loss, loss of appetite, but, no history of fever, jaundice, no previous history of abdominal surgery, and had no comorbidity. On general examination, he had multiple hyperpigmented macules in oral mucosa, palms and soles (Figure 1 and 2).

On per abdominal examination, abdomen was distended with visible intestinal peristalsis. Diffuse tenderness was found and bowel sounds were exaggerated. On digital rectal examination multiple rectal polyps felt, and all hernial orifices were normal. X-ray abdomen showed dilated small bowel loop with air fluid levels. Computed tomography (CT) abdomen showed dilated small bowel and large bowel, with descending colon pulled into sigmoid colon along with its mesocolon and vessels suggestive of colocolic intussusception (Figure 3).

In view of intestinal obstruction, he underwent emergency laparotomy. Intraoperative findings showed a colocolic intussusception of descending and sigmoid colon causing intestinal obstruction, and multiple large polyps all over the colon (Figure 4). In emergency subtotal colectomy done and histopathology report showed Peutz jeghers polyp and moderately differentiated adenocarcinoma. Post-operative period was uneventful. His gastroduodenoscopy showed multiple sessile and pedunculated gastric and duodenal polyps. Further, he underwent adjuvant chemotherapy and completion proctocolectomy.

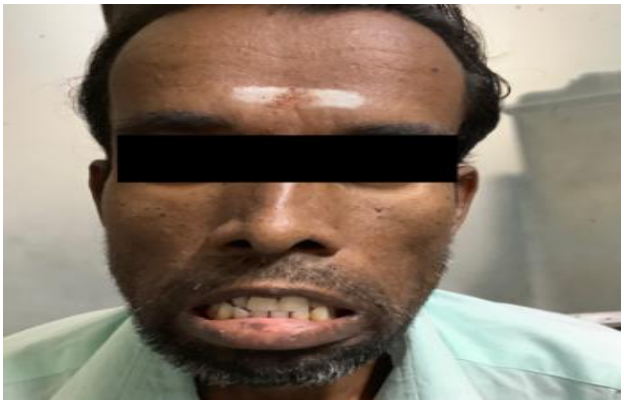

Figure 1: Mucocutaneous pigmentation.

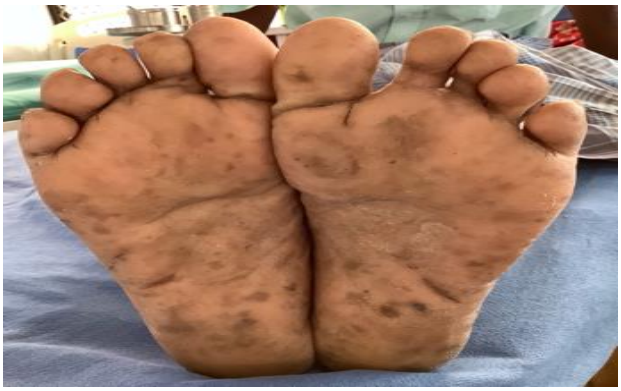

Figure 2: Pigmentation over soles.

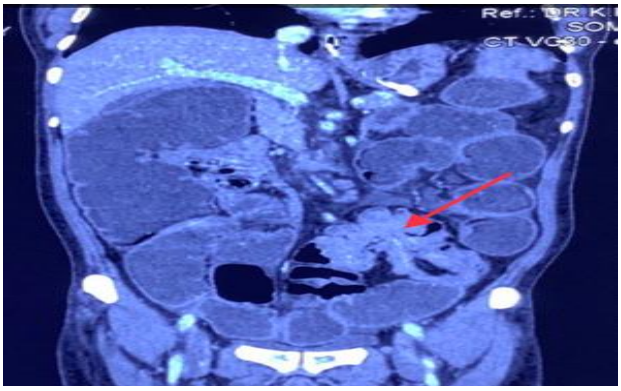

Figure 3: CT scan showing colo-colic intussusception.

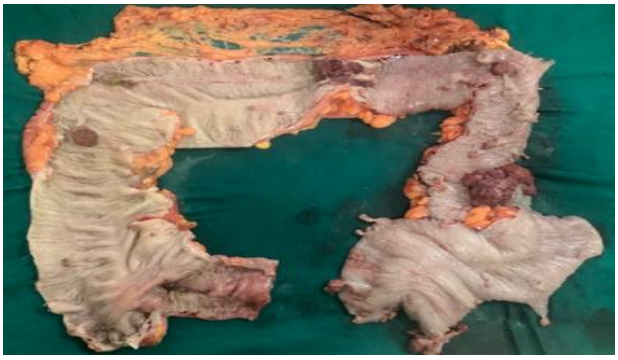

Figure 4: Resected specimen showing multiple polyps.

DISCUSSION

PJS is an autosomal dominant inherited medical condition characterized by hyperpigmented mucocutaneous macules on the lips, mucous membranes of the mouth, hands, and feet, as well as hamartomatous polyps in the digestive tract. PJS was first described by Peutz in 1921 and Jeghers in 1944 and 1949.[1]

It is a rare entity. Its prevalence is around 1 in 100,000 people.[2] This syndrome is seen in both male and female patients with no racial predominance.[3] The World Health Organization has laid the following criteria allowing clinical diagnosis of PJS: three or more histologically confirmed Peutz–Jeghers polyps, any number of Peutz–Jeghers polyps with family history of Peutz–Jeghers syndrome, characteristic mucocutaneous pigmentation with a family history of PJS, any number of Peutz–Jeghers polyps and characteristic mucocutaneous pigmentation.[4] Patients with PJS often present with a history of intermittent abdominal pain due to small bowel intussusception caused by the polyps. The majority that have been reported in the literature are either ileal or jejunal.[5]

Colo-colonic intussusception, such as this one, is very rare and has only been reported only in a few cases.[6] PJS is associated with elevated risk of various malignancies. The most common cancers reported in the literature are gastrointestinal, gynecological, colorectal, pancreatic, and lung cancers.[7] Polyp removal is the standard therapy to prevent complications. Any polyp that is larger than 1.5 cm should be removed if possible. An exploratory laparotomy is required in cases of intestinal obstruction and persistent/intermittent bleeding, if not controlled non-operatively. In our case, an emergency exploration of abdomen was done for acute intestinal obstruction presentation. Periodic surveillance and removal of larger polyps aim to reduce the likelihood of complications in PJS. Surveillance guidelines include upper endoscopy and colonoscopy, starting at 8 years of age. If polyps are detected, the procedure should be performed again every 2-3 years. If no polyps are detected, the procedure should be performed again by 18 years of age, and then every 2-3 years thereafter.[7]

CONCLUSION

PJS is an autosomal dominant inherited disorder. This is rare condition, and associated with mucocutaneous pigmentation and hamartomatous Intestinal polyps. Individual may present in rare case with acute intestinal obstruction associated with intussusception due to polyps. High indexes suspicion is needed in individuals presenting with mucocutaneous pigmentation especially in and around oral cavity. Routine screening with endoscopy in early age group is recommended to prevent polyps related complication. Patient should undergo lifelong follow up with regular clinical examination and genetic counselling.

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