



Case Report

TWO CASE REPORTS OF PEUTZ JEGHER'S SYNDROME

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Summary:

We are reporting two cases in a family, Elder sister was 10 year old & younger one was 8 years old. Both of them were presented to us with the features of gastric outlet obstruction. Further examination revealed that there is pigmentation on their muco-cutaneous junction of lips. Their father also had pigmentation on the muco-cutaneous junction of lips

Introduction:

Association of intestinal polyp with abnormal mucocutaneous pigmentation 1st described by Peutz in 1921. Jeghers & associate defined the syndrome in 1949. Peutz – Jeghers Syndrome (PJS) is a rare condition that tends to run in families¹. About one in 160000 to 1 in 280000 persons develop PJS. Most cases are inherited as an autosomal dominant pattern and some caused by mutation in a gene called STK 11, also known as LKB 1, which is located on chromosome 19².

Its clinical hallmarks are facial mucocutaneous pigmentation and diffuse gastrointestinal polyposis of hamartomatous origin. Other features : Repeated episodes of abdominal pain caused by intussusception, melena, or occult blood loss from the intestine, and anemia³. Peutz Jegher's Syndrome are generally regarded as benign hamartomatous lesion, although in the long term, PJS does carry a slight increased risk of intestinal carcinoma. Currently, it is

believed that patients with PJS have an increased risk for developing cancer both inside and outside the GI tract⁴.

Case-1

A 10 years old girl named LAILA Was admitted in our hospital with the complains of vomiting for 6 months, some thing moving in upper abdomen from left to right for 2 months and epigastric pain for 2 months.

Vomiting occurs usually after taking food and vomitous contains food materials which has taken previously and after vomiting she feels better. For the last 2 months she recognized that something moving in the epigastrium from left to right. She also felt epigastric pain for the same period. The pain was spasmodic and intermittent in nature and was relieved after vomiting.

On examination there is hyper pigmented area on mucocutaneous junction of lower lip (Fig.1), there is visible peristalsis which moves from left to right in the epigastric region, succussion splash was positive, tympanic on percusion ,bowel sound was present

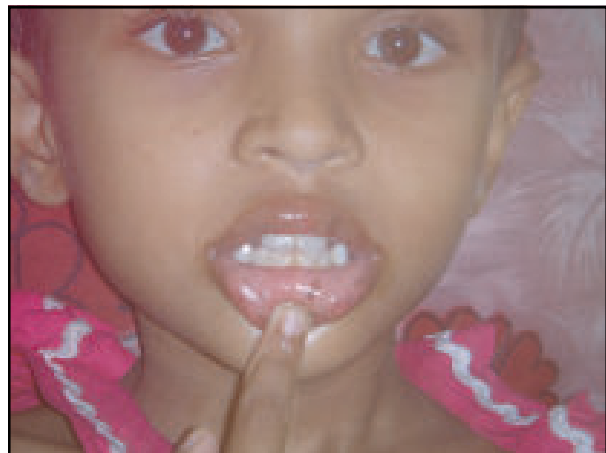


Fig-1: Mucocutaneous pigmentation on lip

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On investigation there was dyselectrolytemia, Ba-meal study of stomach and duodenum reveals that stomach was hugely dilated and delayed emptying.

Per-operatively we found that, there was a couliflower like polypoid growth (Fig. 2) in the proximal jejunum which was resected with part of jejunum and end to end anastomosis was done and send for histopathology. According to histopathology it was a case of a Hamartomayus Polyp (Peutz Jegher's Syndrom). Patient's had uneventful recovery. Upto 14 months followup patient was symptomless.

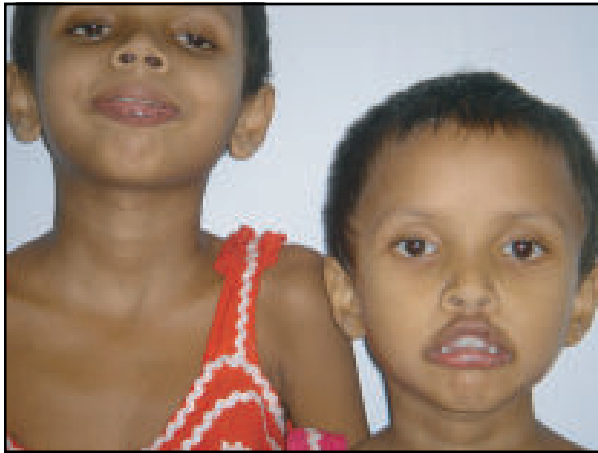


Fig.-2: *Mucocutaneous pigmentation on lip of two sisters in a family (case-1 & case-2)*

Case-2

A 8 years old girl named Laboni, younger sister of the case-1 was accompanied in our hospital with her sister (case-1). After few days she also developed vomiting which usually occurs after taking food and vomitous contains food materials which has taken previously and after vomiting she feels better. She recognized that something moving in the epigastrium from left to right. She also developed epigastric pain and the pain was spasmodic and intermittent in nature and was relieved after vomiting.

On examination there is hyper pigmented area on mucocutaneous junction of lower lip (Fig. 1), there is visible peristalsis which moves from left to right in the epigastric region, succussion splash was positive, tympanic on purcusion, bowel sound was present

On investigation there was dyselectrolytemia, Ba-meal study of stomach and duodenum reveals that stomach was hugely dilated and delayed emptying.

Per-operatively we found that, there was a couliflower like polypoid growth (Fig. 2) in the 2nd part of the duodenum which was resected with surrounding mucosa of the duodenum and was send for histopathology. According to histopathology it was a case of a Hamartomayus Polyp (Peutz Jegher's Syndrom). Patient's had uneventful recovery. Upto 14 months followup patient was symptomless.



Fig.-3: *Excised polyp from jejunum of case-1*



Fig.-4: *Excised polyp from Duodenum of case-2*

Discussion:

Peutz-Jeghers Syndrome is characterized by the presence of multiple hamartomatous polyps in GI tract along with mucocutaneous pigmentation⁵. The polyps are nonneoplastic and may be found in stomach, small intestine and colon, though in the second case the polyp was found in the second part of the duodenum⁶. The complains are often characterized by periodic crampy abdominal pain and associated nausea and vomiting. Both of our cases presented with vomiting after taking food and vomitus contained residual food

particle of previous day, just like the case of gastric outlet obstruction. This obstruction was caused by the duodenal and jejunal polyp.

At the time of exploration the entire GI tract should be examined and palpated. Intestinal resection is not usually required; however, if intussusception is present and not reducible, then a segmental resection with primary anastomosis is required⁷.

Conclusions:

Most patients with Peutz-Jeghers syndrome survive well into adulthood; however, In addition to the slight increase in GI tract cancer, there is an increased incidence of the extraintestinal cancer, such as breast, gonads and pancreaticobiliary tract. The risk of cancer does not require prophylactic removal of these organs, but one should be aware of this possibility as these patients progress into adulthood.

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