A CASE OF PEUTZ-JEGHER SYNDROME

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Introduction

An autosomal dominant disorder, Peutz-Jegher syndrome is characterized by hamartomatous polyposis of the gastrointestinal tract. It is distinguished from other polyposis syndromes by the classical mucocutaneous pigmentation [1]. We report here an eighteen year old boy who developed intestinal obstruction twice over a period of 13 years and required multiple laparotomies.

Fig. 1 : Melanotic pigmentation of lips

Case Report

An 18-year-old was admitted with 3 day history of abdomen pain, vomiting, distension and constipation. He was operated in 1985 for ileocecal intussusception resulting from a polyp and was asymptomatic thereafter. Clinical examination revealed a thin boy with distended abdomen having visible peristalsis. He also had melanotic pigmentation of lips and palms. (Fig 1,2) Plain X-ray of the abdomen revealed multiple air fluid levels. Biochemical and haematological parameters were normal. Conservative treatment did not succeed and he was subjected to laparotomy after five days.

Surgery revealed a densely contracted mesentery with interloop adhesions. A dense stricture causing obstruction at the ileocaecal junction was resected and end to end anastomosis performed. Multiple large polyps were palpable in the proximal jejunum and removed via enterotomies. The patient had an uneventful postoperative recovery. Histopathological examination of the polyps revealed hamartomatous features. There were hypertrophic bands of muscularis mucosae enclosing islands of normal intestinal glands containing goblet cells. (Fig 3). HPE of stricture showed gross fibrotic thickening.

Postoperative barium meal follow through and enema of the patient did not reveal any remaining polyp. His two siblings neither had any melanotic pigmentation nor any polyp on radiological investigation.

Discussion

Gastrointestinal polyposis syndromes are rare hereditary disorders. They commonly present with bleeding or intermittent small bowel obstruction [2].

Fig. 2 : Melanotic pigmentation of palms

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Fig. 3 : Hamartomatous polyposis of gastrointestinal tract

Many familial polyposis are transmitted as autosomal dominant traits. The classical melanotic pigmentation of Peutz-Jegher syndrome immediately distinguishes it from other forms of polyposis. Seven cases of Peutz-Jegher syndrome were found over a study period of 3 years among 103 children with gastrointestinal polyposis in Taiwan [3]. The average age of diagnosis is the third decade with no racial predisposition. Our patient did not have pigmentation earlier during initial presentation in 1985 and manifested when he was 7-8 years old. The tendency to have recurrent obstruction and complication requiring repeated laparotomies is less appreciated than the disorder [4]. In addition, operations frequently involve complications and need to be repeated [5]. Radical removal of all polyps must be the aim of treatment.

Though Peutz-Jegher polyps cannot be regarded as premalignant, associated adenomatous polyps with high malignant potential can co-exist [6]. There is an increased risk of developing carcinomas involving

pancreas, breast and lung. Ovarian and testicular tumours may also develop with greater frequency in these patients and need appropriate treatment [7]. Our patient presented with intussusception in 1985 when the syndrome was not evident. The nature of the polyp was reported as being angiomatous. During the present exploration multiple proximal jejunal polyps were found apart from the fibrotic narrowing at the site of previous surgery. It needs to be stressed that radical removal of the polyps must be the aim of surgery. Patients need to be closely followed by examination, endoscopy and radiological investigations to avoid the complications of regrowth and detect malignant degeneration.

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