A CASE REPORT OF PEUTZ-JEGHER'S SYNDROME WITH REVIEW OF LITERATURE

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INTRODUCTION

Not infrequently a dermatologist is of vital importance in taking a lead to guide the patient's destiny. Herewith we are reporting a case of Peutz-Jegher's syndrome who was treated for chronic abdominal pain and anaemia for the last eight years. He noticed the pigmentation on his lips (which probably was there for many years) during his convalescence, while he was reading about the clinical manifestation of Intestinal polyposis from a text book of surgery. Lot of incovenience could have been obviated and cost of therapy could have been reduced if the patient had been aware of the pigmentation and tried to find out an explanation for it, at an early date

CASE REPORT

A male medical student aged 23 years attended skin out door for pigmentation on the innerside of his lower lip and for pigmentation on both hands for 6 months (Picture 1 and 2) On enquiry he gave history of consuming chloromycetin 6 months ago, when he was operated for acute intestinal obstruction and was found to be having multiple intestinal polyposis resulting in intussusception at three places in the small intestine. On further enquiry he gave history of chronic abdominal pain and anaemia for the past eight years. Pain was colicky in nature, usually localised in epigastrium or umbilical region, non-radiating and subsiding without any medication. Two years prior to the operation he consulted a surgeon for his chronic abdominal pain, where upon he was given a complete physical check up and investigated. The investigations were as follows: 1. Hb% 10 gms% 2. Stool N. A. D. 3. Gastric Analysis. N. A. D. 4. Plain X'Ray Abdomen. N. A. D. 5. Ba-Meal stomach and Duodenum were normal though barium follow through was not dono. 6. Cholecystography. N. A. D. 7. Sigmoidoscopy N. A. D.

Later date, he developed pain in epigastrium which was burning in character for which he was investigated. Hb% was 11 gms, stool report showed positive occult blood and ba-meal revealed a duodenal cap deformity. He was treated with antacid, milk dies etc. without any relief.

On 15th December 1967, he got an attack of sudden colicky pain, vomited and developed distension of abdomen for which he was admitted. Plain X'Ray was taken which showed gas under diaphragm. So he was operated with pre-operative diagnosis of perforated peptic ulcer. On opening the abdomen, multiple intestinal polyposis with intussusception of small bowel at three places were found. Multiple

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enterotomies were done to remove polypi and reduced the intussusceptions. Biopsy report revealed adenomatous polyposis (Picture–3).

DISCUSSION

In 1921, Peutz described familial gastro-intestinal polyposis with pigmentation of skin and mucus membrane. In 1949, Jegher[1] gave masterly account of 10 patients suffering from the similar condition. Since then more than 100 cases have been reported. As it affects more than one member of family, it is belived to be of heredo-familial in origin. It is inherited through a Mendelin dominent gene of high penetration and it expresses itself by pigmentation and polyposis[1].

Usually patients present themselves for 3 cardinal symptoms, chronic abdominal pain, anaemia and pigmentation. In several cases of intestinal polyposis the characteristic pigmentation of Peutz-Jegher's syndrome may have passed unnoticed and in several cases of muco-cutaneous pigmentation, intestinal polyposis may not have manifested and so many cases have remained undetected[2].

Twin sisters with typical distribution of the pigmentation did not show any symptoms of gastrointestinal polyposis. Follow up of the same cases showed that one sister died of acute intestinal obstruction at the age of 20 and the other died at the age of 32 due to carcinoma of breast[2].

The cutaneous pigmentation may appear either at birth or in infancy. It would be an exception if it appears after a decade or two. Such pigmented spots tend to fade at puberty. The pigmented patches of mucous membrane are the sine-qua-non of the syndrome and they persist throughout the life[2].

Before 1959, curiously, the high incidence of malignant degeneration was reported but contrary to it, in 1959 a low incidence of malignant degeneration of the intestinal polyps was found and published, as a part of the Peutz-Jegher's syndrome[3]. The polyps usually observed in small intestine may cause obstruction of intussuscetion or may show malignant degeneration[4].

A case study of 14 patients on whom follow up information was available, one had died of metastatic carcinoma (associated with polyposis). No other pasient is known to have had malignancy. Seven are free from intestinal symptoms. The reason for this is benign behaviour which is unclear but is said that polyp represents hamartomatous over growth rather than true neoplasm[5].

In our patient, pigmentation in buccal mucosa is of unknown duration, (? 6 months). No other member in his family has any clinical manifestation of Peutz-Jegher's syndrome.

This syndrome might be associated with polyposis in viscera other than intestine viz. urinary, bronchial tree and nose or associated with other abnormalities like clubbing, scoliosis, congenital heart disease, diverticulosis and diverticulitis, retarded growth and diseases of thyroid, suprarenal or ovary.

SUMMARY

A case of Peutz-Jegher's syndrome reported with buccal pigmentation of unknown duration and of palmer pigmentation of six months duration with symptoms of intestinal polyposis.

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