superficial epithelium. It consists of regular gland elements which are larger than normal but not hyperplastic, separated by a stroma which is vascular and in places oedematous but does not differ essentially from the normal lamina propria. The individual epithelial cells are tall columnar cells with basal nuclei and do not differ essentially from normal epithelial cells. These polyps are essentially hamartomas or malformations and not benign neoplastic polyps like the single or multiple sessile or pedunculated adenomas of large intestine."

Biopsy of one of the pigmented spots showed melanin pigment in the basal cell layer of the buccal mucosa. Sigmoidoscopy and cystoscopy before discharge, a month after his operation, did not reveal any polyps in the large bowel or bladder. There was no evidence of nasal polyps. Apart from some short attacks of abdominal colic and constipation, he has remained well since his last admission.

COMMENT

Few cases of the Peutz-Jeghers syndrome have been reported in childhood. There were four children from three families in Dormandy's series. Fisher (1951) reported one of the youngest, in a boy aged 7 years, with multiple polyposis of the stomach, duodenum, and jejunum. The radiological findings in the patient were unusual and remarkable in that such appearances—demonstration of an intussusception and polyposis—are rarely seen or recorded in the Peutz-Jeghers syndrome. Behrer (1951) recorded the history of a 14-year-old boy with the Peutz-Jeghers syndrome who was initially thought to have a bleeding peptic ulcer and in whom barium-meal examination showed a jejunal intussusception. The radiological appearances were strikingly similar to the films taken in our case. Colquhoun (1959) demonstrated a jejunal intussusception by barium-meal in a 33-year-old woman who was later found to have an isolated jejunal polyp as a cause of her intussusception. Buckstein (1953) mentioned the case of a 6-year-old child with melaena and anaemia, in whom a barium study showed a dilated segment of small bowel, with an irregular longitudinal line running through it, caused by an intussusception, led by an adenomatous polyp. Although intestinal polyps may appear as filling defects on ordinary barium studies, their detection demands a high degree of suspicion on the part of the radiologist. Special methods such as the double-contrast technique, whereby air is injected after a barium enema, are particularly valuable in visualizing these lesions (Buckstein, 1953; Dormandy, 1957).

I am indebted to Dr. H. M. T. Coles, under whose care the patient was admitted, and to Dr. Basil Strickland, Mr. Duncan Forrest, and Dr. Ian Dawson for their advice on the radiological, surgical, and pathological findings. Dr. Peter Hansell kindly took the photographs.

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Peutz-Jeghers Syndrome: A Further Family History

The association of small-intestinal polyposis and buccal pigmentation, often presenting as a familial condition, has aroused great interest in recent years. In 1896 Jonathan Hutchinson described twin girls who had a peculiar spotty pigmentation around their mouths. In 1919 Parkes Weber recorded that one of the twins had died 11 years later after an operation for intussusception. Peutz (1921) noted the familial association and Jeghers *et al.* (1949) reviewed the literature and described 10 further cases. This condition is consequently referred to as the Peutz-Jeghers syndrome. Tanner (1951) described the first case in this country, and the recent world literature from time to time records further cases. We present another family in which mother and daughter were affected by the disease.

CASE 1

A girl aged 9 was admitted to Southampton Children's Hospital on June 3, 1961, as an acute abdominal emergency. She complained of severe abdominal pain of 36 hours' duration, associated with vomiting. The pain was central and subumbilical, constant in nature, but with occasional colicky spasms. The patient had vomited eight times on the previous day and six times on the day of admission. She gave a history of a number of similar but milder attacks over the previous six months and was said to go pale during these episodes. These were associated with very marked borborygmi, which her mother said could be heard across the room. On account of these attacks she had been referred to a paediatrician who concluded that they were hysterical in nature and attributed them to her change of school—which she did not like. There was no other relevant history.

On examination her temperature was 98° F. (36.7° C.), pulse 120, respirations 24. The most arresting feature was the considerable freckling of the face and circumoral

FIG 1.—Photographs of Case 1.

pigmentation (Fig. 1). This pigmentation extended on to the mucous and buccal surface of both lips. The pigmented spots were 2-3 mm. in size, quite clearly circumscribed, and did not fade on pressure. There was also one pigmented spot of similar size on the palm of the left hand. Examination of the abdomen showed central abdominal distension and there was a mass to the right of the umbilicus. Bowel sounds were absent. A diagnosis was made of intussusception consequent on small-intestinal polyposis.

Operation.—Exploration of the abdomen through a right paramedian incision disclosed a gangrenous intussusception; this was resected and continuity of the bowel was restored by side-to-side anastomosis. The intussusception, when reduced, was shown to consist of almost 5 ft. (1.5 m.) of gangrenous small bowel at the apex of which was a polyp the size of a small chestnut. The histological report on the specimen was as follows: "*Small-gut intussusception with polyp—Peutz-Jeghers syndrome.*—A markedly distended spherical loop of small bowel measuring 15 cm. in diameter. On releasing the intussusception approximately 40 cm. of bowel formed the intussusciptions. At the proximal end of the bowel a dimple was noted measuring 1 cm. on the external surface, and a 3-cm. raised dark red polypoid lesion was present on the inner aspect of the bowel at this point. The mesenteric lymph nodes were slightly enlarged and congested but appeared otherwise normal. **Histology.**—The polypoid mucosal lesion is composed of hyperplastic papillary mucous epithelium typical of Peutz's syndrome. Detailed cytological study is not possible owing to infarction of most of the polypus, a result of the intussusception."

The patient made a satisfactory recovery from this operation and was discharged home on June 20.

CASE 2

An interview was arranged with the mother of Case 1. She was aged 35, and when she attended her own circumoral pigmentation and buccal pigmentation were very obvious (Fig. 2).

At the age of 19 she suffered from recurrent attacks of severe abdominal colic associated with vomiting and sometimes fainting. These attacks became more frequent over a period of one year. Borborygmi were very prominent

FIG. 2.—Photographs of Case 2.

during the attacks. She attended her doctor, but no abnormality was found and she was regarded as hysterical.

On December 28, 1946, she presented with a 24-hour history of severe abdominal pain which came on suddenly. Since the onset vomiting was almost continual, the pain, however, coming and going. On examination her condition was described as fair. The abdomen was tender on palpation and a mobile mass was present subumbilically. Bowel sounds were heard. Operation was performed by Mr. S. N. Lytle, and a gangrenous intussusception consisting of 5 ft. (1.5 m.) of small bowel was resected and end-to-end continuity was restored. Examination of the specimen showed several small polyps on the mucous surface. The patient made a satisfactory recovery from this operation and was discharged home three weeks later.

When seen with her daughter 15 years later she said that she had had no further trouble and on examination there was no evidence of anaemia or ill-health. She had a pigmented spot on the palm of her left hand identical with that on her daughter's hand.

Other members of the family, including the grandparents and the brother of the younger patient, were interviewed. None of these had characteristic pigmentation or had suffered abdominal symptoms.

COMMENT

These patients presented several features of interest. In both cases the characteristic symptoms of colicky abdominal pain associated with loud borborygmi had been present for some time preceding the acute episode, and both patients were labelled as hysterical. Although gangrenous intussusception is said to be rare (Dormandy, 1958), both mother and daughter suffered this calamity. Both patients had a pigmented spot on the palm of the left hand. Neither exhibited other manifestations of the disease, such as clubbing of the fingers or skeletal or cutaneous defects. The mother has suffered no other further attacks. That she is probably free of polyps is indicated by the lack of anaemia. She is very fortunate because this is often a progressive condition, further episodes of colic associated with chronic anaemia being all too common a sequel to an apparently successful operation. It remains to be seen whether the daughter likewise remains free of symptoms.

Although the disease is said to be inherited through a Mendelian dominant gene of high penetration, it is surprising that neither parent of the affected mother nor any other near member of the family seems to be affected. However, either pigmentation or polypi may occur alone and the polypi if present may themselves give rise to no symptoms. Other members may therefore be affected unless this condition happens to be a recent mutation within the family.

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