The patient is now very well and leading a normal, active life, though the subdermal tissues in his arms and legs remain bound down and tight. Skin biopsy shows no skin atrophy or loss of appendages but some subdermal fibrosis.

Comment

Since the initial description several authors have emphasised the distinctive features of eosinophilic fasciitis.^{2,3} While the appearances may be confused with scleroderma, the pattern of skin affection is unusual in that the hands, feet, and face are spared. Furthermore, the thickening and induration affect the subcutaneous tissues but not the skin itself. Raynaud's phenomenon does not occur. The blood eosinophilia is a striking accompaniment, with counts as high as $6 \times 10^9/l$ ($6000/mm^3$). Nevertheless, eosinophils are not often noted on histological examination of the skin, muscle, or subcutaneous tissues. Hypergammaglobulinaemia is prominent, though IgE levels are normal. Antinuclear antibodies are absent.

The histology of the skin and appendages is normal, without atrophy. There is a prominent increase in subcutaneous collagen with an associated inflammatory infiltrate composed largely of lymphocytes and plasma cells. The underlying muscles may be infiltrated but there is no evidence of muscle fibre necrosis. There seems to be no associated visceral involvement and the response to corticosteroid therapy is usually prompt and sustained. For these reasons the condition merits recognition as a separate entity from scleroderma.

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Department of Medicine, Royal Postgraduate Medical School, Hammersmith Hospital, London W12 0HS

SUSAN L STUBBS, MB, senior house officer

GRAHAM R V HUGHES, MD, MRCP, consultant physician and senior lecturer in medicine

Involution of mucocutaneous pigmentation of the Peutz-Jeghers syndrome

The Peutz-Jeghers syndrome consists of mucocutaneous melanin pigmentation with gastrointestinal polyposis.¹ There may also be polyps in the respiratory and genitourinary tracts. There is a risk of gastrointestinal malignancy of 12.6% and an association with ovarian neoplasia.² As originally described, pigmentation develops during childhood and gradually fades with age. The growth rate of those polyps is variable, with a tendency for more rapid growth in younger patients.³ Large polyps have been described in a patient in whom there had been no radiologically detectable polyps seven months previously. At the other extreme, a solitary polyp appeared radiographically unchanged in size over 11 years. Polyps may slough off, diminish in size, or even disappear.³ Small bowel hamartomata have been described in the absence of mucocutaneous pigmentation.

Case report

A 43-year-old Jamaican woman presented in May 1975 with a year's history of recurrent abdominal colic and a six-hour history of severe abdominal pain. She was obese with pigmentation of the oral mucosa and on the volar aspect of the fingers. She had left hypochondrial tenderness, guarding, and rebound tenderness with high-pitched bowel sounds. Findings on rectal examination were normal. Plain x-ray examination of the abdomen showed dilated loops of small bowel with multiple fluid levels. Laparotomy confirmed ileocaecal intussusception with a pedunculated fleshy polyp at its apex. There were also three pedunculated polyps proximal to the intussusception with multiple ileal polyps of about 1 cm in height. The four polyps were excised through separate ileostomies. Histological examination

of the polyps showed hamartomatous lesions consisting of branching strands of smooth muscle and connective tissue compatible with the Peutz-Jeghers syndrome.^{1,4} At review of the patient 11 months later there was no evidence of mucocutaneous pigmentation. A double contrast barium enema, a barium meal, and a small bowel enema failed to show any evidence of gastrointestinal polyps. The patient's daughter also presented to us with similar pigmentation and iron deficiency and was found to have a single large bowel pedunculated polyp, which was resected via colonoscopy.

The mother and daughter have Peutz-Jeghers syndrome as confirmed by clinical history, physical examination, and histological appearances of resected tissue. Nevertheless, in the mother's case surgical resection of the large polyps resulted not only in the disappearance of the mucosal and digital pigmentation, but also in regression of the unresected small intestinal hamartomata.

Discussion

The disappearance of the mother's pigmentation occurred over 11 months. It is unlikely that this change was due to the regression that is associated with aging. All these findings therefore pose certain questions regarding the natural history of the syndrome. Firstly, is there a relation between fading of pigmentation and regression of polyps, and could this be explained by certain hamartomata secreting biologically active amines? Secondly, does the removal of the large polyps influence the growth rate of the small sessile hamartomas, as it appears to have done in this case? Finally, if disappearance of mucocutaneous pigmentation indicates regression of polyps, is the risk of malignancy therefore reduced in such patients?

In view of these suggestions has the daughter's colonoscopic resection had an unexpected therapeutic benefit? Should the daughter's pigmentation fail to regress over the same period of time, will further investigation disclose a large pedunculated polyp?

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St Thomas's Hospital, London SE1 7EH

P W KEELING, MRCP, MRCP, medical registrar

N ASTON, MB, BCHIR, casualty officer

H J ANDERSON, FRCP, consultant physician

Perforated jejunal diverticulum due to local iron toxicity

Jejunal diverticulosis is a rare acquired condition, developing progressively with age. The diverticula occur at the mesenteric border of the bowel, mainly where the vasa brevia pierce the muscularis mucosae. They are false diverticula, being lined with mucosa and muscularis mucosae only. The condition is usually asymptomatic. A straight x-ray film of the abdomen often shows fluid levels in the diverticula, which may be misdiagnosed as intestinal obstruction. Nevertheless, abdominal discomfort from gut dysfunction, malabsorption, intestinal obstruction, diverticulitis, haemorrhage, or perforation may occur. Perforation of a jejunal diverticulum is rare and fewer than 30 cases have been reported.¹ In most of these cases perforation was secondary to diverticulitis, though foreign body perforation has been recorded.^{2,3} The present case of jejunal diverticular perforation, however, appears unique.

Case report

A 74-year-old woman presented as an emergency with a 24-hour history of severe generalised abdominal pain of insidious onset but rapidly increasing severity. There was no history of indigestion or analgesic ingestion. Five

months previously the patient had begun a course of Ferrogradumet (slow-release iron), after being found to have a haemoglobin of 11.2 g/dl.

On admission her abdomen was rigid and a plain x-ray film showed free air under the right diaphragm but no other fluid levels. Intestinal perforation was diagnosed. At operation severe generalised peritonitis was found, though with little free fluid. The upper jejunum contained several diverticula along the mesenteric border. The largest of these (1.7 cm in diameter) was perforated, with a tablet extruding through the hole. The affected bowel was resected and an end-to-end anastomosis performed. Initially after the operation the patient was unable to maintain normal blood gas tensions without mechanical ventilation and she did not regain full consciousness. For a short period she could breathe without assistance but after a day her condition deteriorated and, despite antibiotic treatment and further mechanical ventilation, she died 72 hours after the operation. At necropsy the cause of death was found to be severe bronchopneumonia. Histological examination of the resected jejunum showed the perforated diverticulum to be only minimally inflamed, but at the site of the perforation the tissues were heavily stained with iron, with increasing density up to the actual perforation. The perforation appeared to be due to focal necrosis from local iron toxicity. The tablet lodged in the hole was shown by section and staining to be a slow-release matrix containing some unreleased iron. The tablet had the appearance of the Ferrogradumet previously prescribed for the patient.

Discussion

Slow-release tablets may consist of either multiple small particles of differing rates of dissolution or single entities in which the active principle leaches out from the matrix which remains unabsorbed. Local toxicity has not been reported with the first type, but has been reported when tablets of the second type have been retained in contact with the intestinal wall for prolonged periods. This has been most frequently described with potassium preparations. One case involving Ferrogradumet has been reported,⁴ where a tablet lodged in a Meckel's diverticulum caused local gangrene.

Since it is not normally possible to predict which patients are likely to have gut abnormalities it would seem wise to restrict this type of tablet formulation to patients unable to tolerate oral medication in any other form.

I would like to thank Mr R T Burkitt for allowing me to report this case.

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Ashford Hospital, Middlesex

C J H INGOLDBY, FRCS, surgical registrar

Transient hypocalcaemia and T-cell deficiency in an infant

Hypocalcaemia occurs commonly in the first two weeks of life. In the first three days this is a feature particularly affecting sick low-birth-weight infants and later in the first week well infants taking high phosphate feeds.¹ Serum parathyroid hormone concentrations are frequently low.²

Hypocalcaemia in association with T-cell deficiency was first described by Di George. In this syndrome there is abnormal development of structures derived from the third and fourth pharyngeal pouches with absence of the thymus and parathyroids, abnormalities of the great vessels, and characteristic facies.³

The finding of temporary hypocalcaemia and T-cell deficiency has not to our knowledge been reported.

Case report

A 6-week-old Pakistani boy was admitted with a two-day history of convulsions. He had been a full-term baby (birth weight 3000 g), entirely breast-fed, and physical findings were normal. He continued to have intermittent

convulsions for seven days. The results of initial investigations showed a definite hypocalcaemia (serum concentrations of calcium 1.32 mmol/l (5.3 mg/100 ml), phosphate 2.8 mmol/l (8.7 mg/100 ml), and magnesium 0.68 mmol/l (1.66 mg/100 ml)). He was treated with intravenous and oral calcium supplements, together with anticonvulsants, and after the seventh hospital day no more fits occurred. The serum calcium concentration, however, remained below 2 mmol/l (8 mg/100 ml) for almost three weeks.

Immunological values after correction of hypocalcaemia

Age (weeks)	Serum calcium (2.35-2.65) (mmol/l)	T cells (50-70)	B cells (12-25)	Lymphocyte transformation ratio of tritiated thymidine in test/control		
				PHA 2.0 µg/ml (93-918)	PHA 0.2 µg/ml (30-700)	Pokeweed mitogen
10	2.44	25	39	1.0	0.9	161
14	2.58	29	40	17	21	77
22	2.61	26	30	241	45	14
32		51	25	155	63	37

Conversion: SI to traditional units—Calcium: 1 mmol/l $\approx$ 4 mg/100 ml.

Serum parathyroid hormone estimation on the tenth day was 0.28 ng/ml (upper limit in an adult 0.20 ng/ml), when the serum calcium was 1.67 mmol/l (6.7 mg/100 ml). The results of numerous laboratory and radiological investigations were normal, with no evidence of rickets, but there was doubt about the presence of a thymic shadow on the initial chest x-ray film—while the result of the serum parathyroid hormone estimation was not immediately available. No cardiovascular abnormalities were evident, but Di George's syndrome seemed a distinct possibility. Lymphocyte function tests were, therefore, carried out. The lymphocytes showed an impaired response to stimulation with phytohaemagglutinin (PHA) and an exaggerated response to pokeweed mitogen. In addition, lymphocytes giving spontaneous sheep erythrocyte rosettes (T cells) were reduced, while complement rosetting cells (B cells) were increased. These abnormalities were still present at 14 weeks of age, had shown improvement by 22 weeks, and were absent by 32 weeks.

Discussion

Hypocalcaemia presenting at 6 weeks of age is unusual. Our patient had no findings to suggest associated gastrointestinal, renal, or parathyroid disease. The raised serum parathyroid hormone concentration was expected in the presence of hypocalcaemia and excluded parathyroid insufficiency. We found normal concentrations of calcium, phosphate, and alkaline phosphatase in the mother six weeks after delivery. The possibility that vitamin D deficiency in the mother and infant was the cause of the hypocalcaemia is, therefore, unlikely. The initial hypomagnesaemia was corrected with a single dose of magnesium sulphate, and, although the convulsions continued, subsequent serum magnesium concentrations were normal.

Impairment of T-cell function has been shown in hypocalcaemic rats after parathyroidectomy. Lymphocytes in the thymus showed definite reduction in mitotic activity, leading to atrophy of thymic tissue.⁴ B-cell function was not affected and there was no alteration in the number of antibody-producing cells in the spleen.⁵ Partial correction of the hypocalcaemia by increasing dietary calcium reduced significantly the degree of thymic atrophy.

Possibly the hypocalcaemia in our patient, in the presence of normal parathyroid function, may have been responsible for the apparent delay in maturation of immunological function under control of the thymus.

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Departments of Paediatrics and of Immunology, East Birmingham Hospital, Birmingham B9 5ST

C FOSSARD, MSC, MRCP, senior registrar in paediatrics

M J TARLOW, MSC, MRCP, senior lecturer in paediatrics

R A THOMPSON, MRCP, MRCPATH, consultant immunologist