Dermatitis Herpetiformis in a Treated Coeliac Child

The relation between dermatitis herpetiformis (DH), structural and functional abnormalities of the small intestine, and coeliac disease with gluten sensitivity remains uncertain. It is well established that DH may be accompanied by malabsorption, and that such cases have abnormalities of the small intestinal villi of variable degree (Marks, Shuster, and Watson, 1966; Fraser, Murray, and Alexander, 1967; Fry et al., 1967; van Tongeren, van der Staak, and Schillings, 1967; Marks et al., 1968). Furthermore, malabsorption may precede the characteristic eruption of DH by several years (Fraser, Ferguson, and Murray, 1968). The importance or even relevance of these observations to gluten sensitivity is less clear, since the degree of villous atrophy found in DH may fall far short of subtotal villous atrophy which is the diagnostic essential in gluten sensitive coeliac disease. Nevertheless, gluten withdrawal may benefit the malabsorption syndrome of DH (van Tongeren et al., 1967; Marks et al., 1968; Fraser et al., 1968), and gluten sensitivity has been demonstrated in the small intestinal mucosa (Shuster, Watson, and Marks, 1968). In addition, improvement in the skin manifestations of DH has been noted in some patients on a gluten-free diet (van Tongeren et al., 1967; Marks et al.,

Conclusions

While there are several possible causes for this infant’s arthrogryposis, drug-induced immobilization of the fetus at the time of or shortly after the development of the joint cavities seems the most probable.

The severity of the deformities of this infant, and ubiquity of the drugs that may have caused them, would probably justify a review of the prolonged administration of relaxant drugs to women in the first trimester of pregnancy.

Summary

An infant with arthrogryposis was born to a mother who had been treated with d-tubocurarine in early pregnancy. The arthrogryposis is attributed to immobilization of the fetus at the time of joint formation.

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1968; Fry et al., 1967). Fry and his co-workers believe that the enteropathy in DH is caused by gluten sensitivity, and that the skin lesions are related to the enteropathy; Shuster and his colleagues consider that the degree of improvement in the skin lesions of patients on a gluten-free diet can be explained by the natural history of the disease process.

In the following report, an 8-year-old coeliac boy developed DH while on a gluten-free diet. The diet had been adhered to for over 5 years, and had produced a complete clinical and histological remission. It is suggested that this supplies significant evidence against the hypothesis that gluten might be an important factor in the aetiology of DH.

Case Report

This boy was born in July 1960. Mixed feeding was introduced at the age of 2 months. When he was 6 months old, he began to have bouts of loose pale stools which persisted until he was investigated in July 1962. At that time his height was on the 25th centile and his weight was on the 20th centile. A fat balance with a 40 g. fat intake gave 86% absorption; Hb was 9·6 g./100 ml., serum iron 16 μg./100 ml., and the blood film was hypochromic; a barium meal and follow-through showed a malabsorptive pattern; there was radiological osteoporosis; and a jejunal biopsy showed subtotal villous atrophy.

A gluten-free diet was given, and this produced a resolution of symptoms, with growth acceleration until both height and weight reached the 75th centile. For a period of 6 months in 1965 a normal diet was resumed against medical advice, and this resulted in a weight loss of 1 kg. A gluten-free diet was restarted, weight gain resulted, and the diet has since been firmly maintained.

In January 1968, he developed an itchy eruption affecting mainly the extensor aspects of elbows and knees, buttocks, shoulders, and face. The eruption consisted of small erythematous macules, with some raised urticarial lesions. The patient was initially treated with antihistamine preparations, but because of a poor response to this therapy he was admitted to hospital for further investigation. Oral ingestion of 350 mg. potassium iodide induced a marked exacerbation of the rash, and skin biopsy showed changes consistent with DH. Other investigations, performed while the patient remained on a gluten-free diet, gave the following results: Hb and full blood count within normal limits, serum proteins and immunoglobulins normal, 5-hour D-xylose excretion 42%, daily faecal fat excretion (on ward diet) 3·8 g., and biopsy from the duodeno-jejunal function showed normal villous architecture, well-differentiated epithelium, and no increase in the round-cell infiltrate.

The skin eruption cleared on dapsone therapy, and he is at present controlled on 25 mg. on alternate days.

Discussion

Dermatitis herpetiformis is uncommon in children, and its association with coeliac disease in adults has only recently been briefly described (Mayon-White, 1969). In addition, Dyer and Verbov (1968) described an adult male who developed DH and gave a history of treatment for coeliac disease as a child. In most of the adults, patients with DH and an associated enteropathy, the skin lesions have preceded the onset of malabsorption by months or years, though Fraser et al. (1968) recently described the onset of DH in two patients who had had coeliac disease for several years.

The response of the enteropathy of DH to a gluten-free diet is variable (Marks et al., 1966; van Tongeren et al., 1967; Marks et al., 1968; Fraser et al., 1967); even when there is complete symptomatic remission, there are usually persisting histological abnormalities in the small intestine. This has been considered analogous to the situation in adult coeliac disease. The complete histological remission achieved in the present case is compatible with the expected improvement in a child with gluten-sensitive enteropathy (Anderson, 1960).

The skin lesions of DH have been reported to be improved by a gluten-free diet (van Tongeren et al., 1967; Fry et al., 1967), leading Fry and his colleagues to suggest that the skin lesions in DH are related to the enteropathy and thus to gluten sensitivity. The present case supplies considerable evidence against this theory, since the skin lesions first appeared when the child was on a strict gluten-free diet, with no evidence of an enteropathy, clinical, biochemical, or histological.

The relation between the skin and the bowel in DH remains unknown. The features of the child described here suggest that the relation may be indirect, and that gluten is unlikely to be an important factor in causing the skin lesions. It remains possible that DH can induce a gluten-sensitive enteropathy, perhaps in the genetically predisposed.

Summary

An 8-year-old coeliac boy, who had been on a gluten-free diet for 6 years, developed dermatitis herpetiformis (DH) while the coeliac state was in complete remission. This confirms an association between DH and coeliac disease, previously described in adults, but suggests that gluten is unlikely to be an important factor in the aetiology of the skin manifestations of DH.
Familial Duodenal Atresia

Congenital atresia of the bowel is infrequent (Harris and Steinberg, 1954); its familial incidence therefore constitutes a finding of special interest. Very few reports have been published on familial congenital intestinal obstruction not associated with mucoviscidosis (Winter and Zeltzer, 1956; Mishalany and Najjar, 1968; Esterly and Talbert, 1969). To our knowledge no instance of familial duodenal atresia has been described. We wish to report the occurrence of duodenal atresia in 3 sibs.

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The parents are young, healthy first cousins. No history of abortions, stillbirths, or premature deaths in the family could be elicited, nor could they recall any periods of ill-health during pregnancy. Their firstborn, a girl, was the product of a normal pregnancy and delivery, and is healthy. The following three pregnancies, during three consecutive years, were complicated by polyhydramnios in the last two months of pregnancy. The infants were a girl weighing 2300 g. at 35 weeks’ gestation, a boy weighing 2800 g. at term, and a girl weighing 2250 g. at 36 weeks’ gestation. The first two were born at the maternity centre in her Arab village, and the last girl was born at the Hillel Yaffe Government Hospital. The clinical, radiological, and operative findings were essentially the same in all three affected babies: continuous vomiting of bile, epigastric distension, and colourless meconium obtained by rectal examination. Abdominal x-rays showed the classical ‘double bubble’ pattern. Laparotomy revealed atresia of the third part of the duodenum, with marked dilatation of the proximal loop. The first two babies were brought to our hospital at the age of 2 days. The third affected sib, born in our hospital, was operated on 12 hours after birth. End-to-oblique duodenojejunostomy was performed, with good initial results, but all 3 babies died 5 to 6 days after operation. Necropsy revealed bronchopneumonia and localized peritonitis, but no evidence of mucoviscidosis, rubella, or other local or systemic illness which could be considered responsible for the anomaly.

Chromosome studies from the last case showed a normal karyotype.

Discussion

The literature contains very little information on familial congenital intestinal obstruction. 2 brothers reported by Blanck, Okmian, and Robb (1965) had atresia of the middle ileum in association with meconium ileus; Winter and Zeltzer (1956) reported 2 sibs with ileal atresia; Mishalany and Najjar (1968) described 3 sibs with jejunal atresia; Esterly and Talbert (1969) reported twins with jejunal atresia and ascribed the lesion to the effects of rubella arteritis. Our report is the first on familial duodenal atresia, and raises again the problem of aetiology of intestinal atresias. The concept of faulty recanalization after an embryological ‘solid stage’ of the gut as the cause of intestinal atresias has been losing ground. There is mounting evidence that injury to the fetal bowel, especially interference with blood supply to a portion of it, is the main cause of atresias (Grob, 1960; Santulli and Blanc, 1961). This approach has been substantiated by experimental observations (Louw and Barnard, 1955), and is in accordance with the frequent association with mucoviscidosis (Bernstein et al., 1960; Andersen, 1962).

It is, therefore, not surprising that ileal atresia with mucoviscidosis in the kindred reported by Blanck et al. (1965) was ‘familial’, but the actual familial condition was mucoviscidosis. The twins with jejunal atresia described by Esterly and Talbert (1969) were simultaneously exposed to rubella, and the authors assume that vascular damage by the infection disturbed the blood supply to the fetal jejunum, thereby causing atresia. The