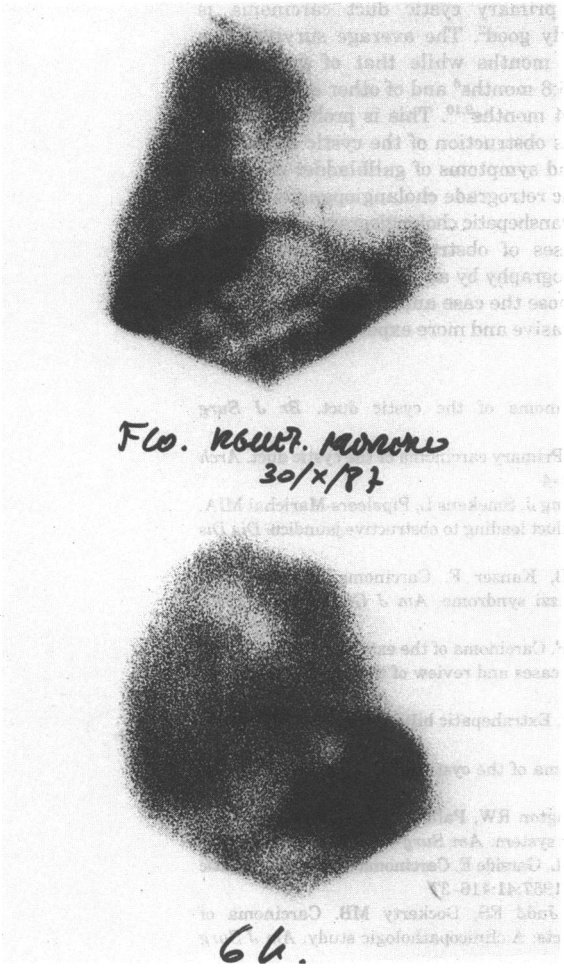

Figure 1.

performed, but 6 days later the patient died in a final phase of hepatic failure.

Discussion

Our results confirm the abdominal origin of hepatic hydrothorax even in patients without sonographic evidence of ascites[6] and serve to suggest the conditions in which spontaneous or diuretic-induced reabsorption occur and the mechanisms by which hydrothorax can be associated with ascites or not, even in the same patient.

The passing of ascites towards the thorax is determined by the sum of the negative pleural pressure (NPP) and the positive abdominal pressure (PAP). When the pleural filling pressure (PFP) equilibrates the former forces (NPP+PAP) the effusion remains stable, but a flow of ascites persists only to replace the liquid which is reabsorbed in the pleural serosa. However the characteristics of this equilibrium vary according to the mechanism of production of the pleural defect. If the hole develops when there is ascites, or is even due to it, the PAP will be very high and, given the limited capacity of the pleural cavity, the persistence of hydrothorax and ascites is logical. Instead, in those patients without ascites a diaphragmatic defect must exist before effusion develops, which permits the passage of liquid as soon as there is a minimal production of ascites. Consequently, in some of these patients, an equilibrium can be established in conditions in which the production of ascites is equal to the pleural capacity of reabsorption, so that ascites will not accumulate. Any decrease in the production of ascites can easily make it smaller than the pleural capacity of reabsorption and the effusion would finish being reabsorbed, as happened in the patient reported here. On the contrary any increase in the production of ascites will lead first to its accumulation and later to its decrease or disappearance if the increase in the PAP forces a net passage of ascites into the thorax, reaching a new equilibrium at a higher level of PFP and a greater hydrothorax, as was observed finally in our patient.

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(Accepted 31 January 1990)

Rare type of visceral myopathy mimicking anorexia nervosa

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Keywords: small bowel; genetic disease; smooth muscle degeneration

We report a case of visceral myopathy involving small bowel without obstructive symptoms or radiologic intestinal dilatation.

This rare type of visceral myopathy acquired by autosomal recessive transmission is characterized by small bowel involvement without clinical obstruction or radiological dilatation.

Case report

A 19-year-old woman who had been treated for anorexia nervosa for two years was admitted with severe cachexia (height 1.52 m, weight 26 kg), oedema and ascites. She complained of chronic diarrhoea (6 stools/day), abdominal pain and vomiting, but she had no signs of obstruction. Examination threw doubt on the initial diagnosis of anorexia nervosa and led to her having extensive digestive tract investigations culminating in laparotomy.

A plain film of the abdomen did not reveal any bowel obstruction. A barium meal showed a hiatus hernia and a normal oesophagus and duodenum. Oesophago-gastro-duodenal endoscopy confirmed the hiatal hernia and was used to provide proximal small bowel biopsies. Colonoscopy was negative and intubation of the terminal ileal loop was unsuccessful. However, barium X-rays of the small bowel were abnormal showing a diffuse enteropathy involving the entire small bowel (especially its middle part). There was mild thickening of intestinal wall, moderate contraction of

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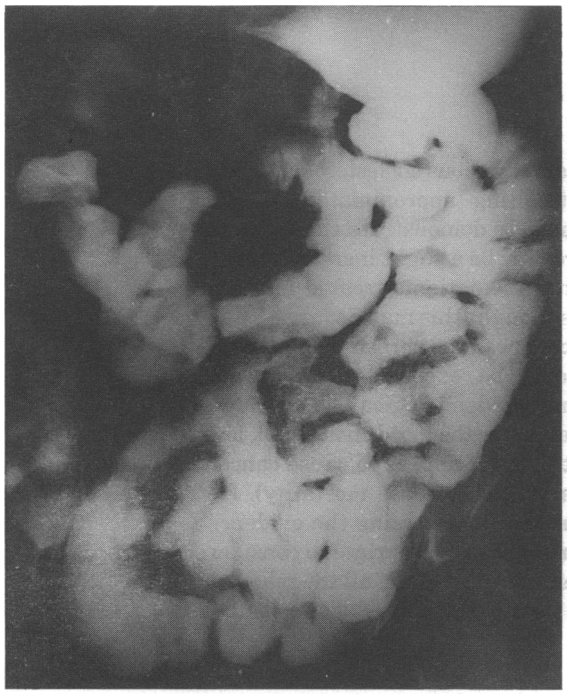

Figure 1. Diffuse enteropathy involving the entire small bowel, mild thickening of intestinal wall, sacculation of the antimesenteric border, total disappearance of the valvulae conniventes and coarse mucosal pattern. No intestinal dilatation

the mesenteric border with early sacculation of the antimesenteric border, diffuse ulceration, total disappearance of the valvulae conniventes and a coarse mucosal pattern (Figure 1).

Oesophageal manometry was normal.

There was severe malnutrition (total serum proteins: 2.9 g/100 ml) and intestinal malabsorption.

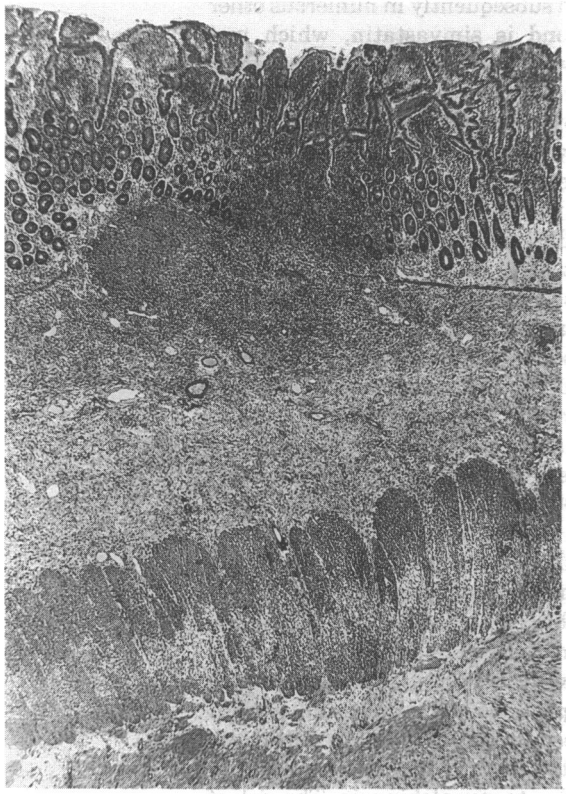

Figure 2. Light microscopy of the ileal biopsy - complete atrophy and fibrosis of the outer muscular layer and partial vacuolar degeneration of the inner layer. The mucous membrane and submucosa are not involved (H&E stain, magnification×15)

All the endoscopic biopsies were superficial and insufficient for diagnostic assessment; therefore, a laparotomy was indicated. The small bowel appeared to be thickened, and although the entire small bowel was involved, the main lesions were located in the ileum. The ileum and jejunum were biopsied.

Light microscopic examination showed that only the muscular layer was affected. The smooth muscle cells presented a vacuolar degeneration responsible for a fascicular atrophy and many were replaced by fibrosis (Figure 2). The outer longitudinal layer of muscle was more affected than the inner circular layer and these changes were more severe in the ileum than in the jejunum. Jejunal biopsy did not show lesions of circular muscle, whereas, on the ileal biopsy, the two layers were involved: the longitudinal outer muscle was almost entirely atrophic and was replaced by thick fibrosis. The inner circular layer showed only vacuolar degeneration. The myenteric plexus appeared normal.

Electron microscopy confirmed the diagnosis with a vacuolar degeneration of the smooth muscle cells. No abnormality of myenteric plexus were noted.

Genetic investigations revealed that her parents were first cousins. The probands sister probably also had visceral myopathy but she refused investigations. Two brothers were asymptomatic.

Discussion

Visceral myopathy was recognized as a distinct entity among many abnormalities[1]. The diagnosis is made by histopathology and is characterized by three abnormalities all of which were present in this case:

(1) vacuolar degeneration and replacement of the smooth muscle cells by fibrous tissue.

(2) outer longitudinal muscle more affected than the inner circular muscle and

(3) the absence of lesions in the myenteric plexus.

In 1987, Kristnamurthy and Schuffler identified four types of visceral myopathy[2]. Our patient appeared to belong to the rarest type which combines gastroparesis with isolated involvement of the small intestine. Unlike the commoner types it is not associated with intestinal pseudo-obstruction[1,3,4]. Only one other family with this type of presentation has been described so far[5]. These also presented chronic diarrhoea, abdominal pain but no dilatation. Unlike the findings in our patient, Jacobs and colleagues[5] found no degeneration in the inner muscle which was, on the contrary, hypertrophic. They attributed the absence of intestinal dilatation to the inner muscle hypertrophy but our observation seems to refute this hypothesis.

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(Accepted 8 February 1990)