and it was postulated that a compensatory reactive hyperplasia of the reticulo-endothelial system had occurred, with hyperphagocytosis, as a result of repeated infections due to the almost complete lack of gammaglobulin and deficiency or absence of circulating antibodies.

In the patient at present described it seems probable that the pathology is of the same nature, with hemolysis a direct result of hypersplenism, the splenic enlargement having now reached nearly to the umbilical level. An alternative cause of the hemolysis as due to a specific antibody formation seems unlikely, in that the patient showed no demonstrable antibody response in the provocative tests employed. Supporting evidence of hypersplenism was given by the absence of leucocytosis throughout the course of illness, the white cell count never being higher than 10,000/c.mm. at the height of the severe pulmonary infection. The platelet count was unaltered. The sedimentation rate was persistently raised, being from 100 to 110 mm. in one hour (Westergren) at the height of the pulmonary infection, and between 24 and 40 mm. subsequently. This was an unusual finding, although present in some recorded cases, the levels in this condition being more commonly low during the infective episodes, in conformity with the known association between the sedimentation rate and the plasma globulins.

During treatment with gammaglobulin, despite a fall in the reticulocyte count to 2%, with rise of hemoglobin to 85% and serum haptoglobin to 90 mg./100 ml., the faecal stercobilinogen excretion was still found to be increased, and the survival time of transfused red cells became further shortened, so that it must therefore be concluded that abnormal haemolysis is still continuing. It is not yet known whether with more prolonged control of the infective episodes by gammaglobulin therapy the hypersplenism will be abated. Should this not follow and a relapse of anaemia develop, the possible need for splenectomy must then be considered.

Summary

A case is described in which idiopathic hypogammaglobulinemia was associated with hemolytic anemia. The evidence for this combined diagnosis is presented and its implications discussed.

We wish to express our acknowledgments and thanks to the Medical Research Council for supplies of gammaglobulin for use in treatment of this patient, to Dr. J. F. Soothill for estimations of serum-gammaglobulin, to Mr. P. M. G. Broughton for many biochemical investigations and to the Radiotherapy Department of the North Middlesex Hospital for red-cell survival rate estimations.

REFERENCES


A CASE OF ACUTE ILEITIS WITH PERFORATION

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Acute ileitis is uncommon in surgical practice and some doubt exists about its pathology and natural history. A case report of acute ileitis with perforation in a child is presented.

Case Report

The patient, a boy aged 10, was admitted to hospital on 8-9-59 with a diagnosis of acute appendicitis. He gave a history of colicky lower abdominal pain of 36 hours' duration. He had vomited several times and had some diarrhea, the motions being loose, but not containing blood. On examination there was tenderness and rigidity in the right iliac fossa. The temperature and pulse were normal.
A diagnosis of acute appendicitis was made. The possibility of gastro-enteritis was considered, but operation was advised, as the local signs were well marked and an expectant policy in children is only justified if acute appendicitis can be excluded with confidence. The abdomen was opened through a grid-iron incision. The appendix was normal, but the terminal ileum was acutely inflamed. The bowel was bright red and hyperemic and its wall was edematous. The mesenteric glands were enlarged. Appendicectomy was performed and the abdomen closed without drainage.

Two days after the operation he showed signs of paralytic ileus, which was treated by gastric suction and intravenous fluids. A course of penicillin and streptomycin was started. Three days later he was still distended, but faint bowel sounds were heard. On the seventh post-operative day he complained of some pain, the abdomen was more distended and his general condition had deteriorated. Operation was advised and the abdomen reopened through a right paramedian incision. A moderate amount of free purulent fluid was present. The distal small bowel was adherent and inflamed and its wall felt thickened, edematous and friable. There were two perforations at its antimesenteric border. A small abscess cavity was opened, exposing another perforation. The perforations were sutured, the adhesions separated, and the bowel deflated with the sucker. The abdomen was then closed with drainage. His general condition was very poor after the operation, but after an anxious two or three days he made good progress. Bacteriological examinations for typhoid, dysentery and abortus were performed later; culture of the feces and serological tests were both negative.

He was seen four weeks after discharge and again 18 months later. His general health was good and he was symptom free.

Discussion

Acute ileitis is fairly uncommon and is usually diagnosed as acute appendicitis, the correct diagnosis only being made at operation. When ileitis is found the abdomen should be closed without any further manipulation. Appendicectomy is contraindicated because of the danger of causing a fecal fistula but Talbot (1958) states that it does not lead to fistula formation unless the cecum is obviously diseased.

Acute ileitis of this type is sometimes called acute Crohn's disease but some doubt exists as to whether it is actually the same disease. Follow-up studies show that the majority of these patients recover uneventfully and have no further symptoms. Crohn's disease, on the other hand, is particularly liable to recur.

Meyer (1960) regards acute and chronic ileitis as two separate diseases. In a review of several previously published series there were only 18% of proved or possible recurrences. There were also histological differences; in the chronic form the submucosal edema is the earliest change, whereas in the acute disease inflammation is present from the start. It has been pointed out that a true chronic regional enteritis can exist with minor symptoms for months or even years and then suddenly have an acute exacerbation (Armitage and Wilson, 1950). If the previous history is not forthcoming these patients may often be regarded as having an acute ileitis. Armitage and Wilson, therefore, recognise two varieties of acute ileitis, one being non-specific and self-limiting and the other, with some previous history of diarrhea or abdominal discomfort, an acute Crohn's disease.

Perforation in the acute stage of Crohn's disease is very rare (Gow and Walsh, 1952). These authors reported three cases including one from the literature, two of which had an acute onset, the other had a previous history suggestive of Crohn's disease. The one patient who survived showed no evidence of Crohn's disease when seen six months later.

Although Crohn's disease typically occurs in young adults, it has been reported in children and Ebrill (1945) reported a case of acute regional enteritis with perforation in a child of nine. He quoted a series of 178 cases from the Mayo Clinic in which three were children under ten. Talbot (1958) also points out that acute ileitis may occur in children.

This patient is of particular interest in view of the two operations. On the first occasion the findings were typical of acute ileitis. It is most unusual for this condition to progress to perforation. In view of the rarity of further complications any operative treatment of acute ileitis, such as resection or even a short circuit, seems inadvisable. As the boy now shows no clinical evidence of recurrence it is probable that he had an acute ileitis rather than Crohn's disease.

Summary

A case of acute ileitis with perforation is presented. The relationship between acute ileitis and acute Crohn's disease is discussed: there is evidence that the two conditions are distinct. Acute ileitis has a good prognosis. The rarity of perforation is emphasized. Both acute ileitis and acute Crohn's can occur in children.

REFERENCES

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