AMYLOIDOSIS occurring in association with regional enteritis was first described by Cohen and Fishman in 1949. Eleven years later, Werther, Schapira, Rubinstein and Janowitz (1960) reviewed the literature and found a total of five cases only. Since then two more cases have been written up in America (Briggs, 1961; Palmer, Kirsner, Goldgraber and Fuentes, 1963), but none has yet been described in Great Britain.

As there is some evidence that this complication of regional enteritis is commoner than these figures suggest, the present cases are recorded in the hope that this association will be recognized more frequently.

Case No. 1

R.W. first developed intermittent diarrhoea in 1951 when he was 20 years old, and 2 years later was admitted to the Central Middlesex Hospital for surgical treatment of multiple fissures in ano.

After two years his diarrhoea became associated with abdominal pain and weight loss and in April 1956, he was admitted to the gastroenterological ward. At that time he appeared generally unwell although no other clinical abnormalities were detectable. However, his stools contained occult blood and a small bowel enema revealed irregularity and narrowing of the terminal ileum. A diagnosis of regional enteritis was made and he was discharged on prednisone which he continued to take intermittently throughout his illness. He was also given antituberculous drugs which had to be discontinued after a month as he developed a hypersensitivity reaction.

His general health improved, but he continued to have painful episodes suggestive of intestinal obstruction and, as further X-rays suggested spread of his disease, laparotomy was advised. This was performed in December 1956 by Mr. Gummer who found extensive regional enteritis involving the terminal ileum with multiple fistula formation. He resected the diseased gut and carried out a right hemicolectomy.

Following a stormy convalescence he did well until 1962 when he was readmitted complaining of swelling of the face and ankles. At that time he was passing 4 g. of protein daily in his urine. His serum albumin was 1.9 g./100 ml and his blood urea was normal. His stools contained a normal amount of fat. Renal and rectal biopsies showed changes of amyloidosis.

He was treated with dietary supplements but shortly after this developed a recurrence of his regional enteritis with intestinal obstruction that required a shortcircuiting operation for its relief. On this occasion his proteinuria amounted to 8 g. daily and his blood urea had risen to 79 mg./100 ml.

In January 1964 he developed acute haemorrhagic pancreatitis which was confirmed at laparotomy and which led to an acute exacerbation of his chronic renal failure. This responded to conservative treatment but his blood urea remained above 100 mg./100 ml. His convalescence was further complicated by venous thrombosis in the legs and recurrent pulmonary emboli. He deteriorated steadily and died in renal failure 6 months later. Autopsy confirmed the presence of extensive amyloidosis with renal vein thrombosis.

Case No. 2

S.L. first developed abdominal pain and diarrhoea in 1940 when he was sixteen years old. Two years later he was admitted to St. Bartholomew's Hospital and, at laparotomy, was shown to have terminal ileitis. Seven feet of bowel that were grossly and microscopically typically affected by regional enteritis were removed.

Three years later his disease relapsed with involvement of his colon, and he was noted to have developed enlargement of the spleen. He continued to have intermittent diarrhoea and bleeding and during the next five years had four operations for recurrent fistulae-in-ano. In 1950 proteinuria was recorded for the first time; his blood urea was, however, normal.

His first admission to the Central Middlesex Hospital was in October 1956 following a severe relapse of his enteritis. He was passing up to six fatty stools daily and had lost three stone in weight during the previous fifteen months.

On examination, he was asthenic and pale with finger clubbing and slight ankle oedema. His blood pressure was 75/50 mm. Hg. and there was some skin but no buccal pigmentation. His spleen was palpable one inch below the left costal margin and there were several perianal fistule.

Investigations: Hb., 62%. Serum sodium 118, potassium 4.1 mEq./l., blood urea = 120 mg./100 ml. serum albumin = 2.5 g./100 ml. with an excess of gamma globulin. The urine contained 4 g. of protein daily and the stools contained 81 g. of fat in three days. Barium follow-through investigations showed recurrence of the disease in the terminal ileum. Amyloidosis was suspected and confirmed by the Congo Red test (only 27% of the injected dose being retained in the circulation at one hour). He was transfused and treated with prednisone, streptomycin and isoniazid with slight improvement. After a month he was discharged from hospital but was readmitted, moribund, with uremic acidosis a few days later. Renal and hepatic biopsies obtained after death showed typical histological appearances of amyloidosis.
Discussion

Both these patients closely resemble those that have been described before. In three of Werther’s cases amyloidosis had contributed substantially to the cause of death. Symptoms of enteritis had preceded the diagnosis of amyloidosis by intervals of eight, ten and fourteen years and death had occurred in each case before the age of forty years. Two of the patients had died of uraemia and one had died during an episode of acute pancreatitis which had developed during a nephrotic stage of his chronic renal disease. The occurrence of acute pancreatitis in Case 1 is of some interest as this is an uncommon disease at this age (Rocker and Bartlett, 1953). This may possibly be related to steroid therapy as the present case had had several years’ treatment with prednisone and Werther’s case had had a short course of ACTH. This association has been suggested several times in the past (Nelp, 1961; Carone and Mebow, 1957).

The development of renal involvement by amyloidosis, as shown by proteinuria, is a bad prognostic sign and Werther’s three cases all died within two years of discovery of this. Case 2 appears to have done relatively well in this respect.

It is well known that amyloidosis is frequently associated with chronic inflammatory disease which is sometimes of a tuberculous nature and that it is sometimes difficult for the pathologist to distinguish intestinal tuberculosis from regional enteritis. Both the above cases were investigated extensively with this in mind without any evidence of tuberculosis being found. Antituberculous drugs were given because it was the routine to treat cases of regional enteritis in this way, but neither case received an adequate course of treatment for tuberculosis.

On the other hand it is hardly surprising that regional enteritis, which is a disease characterised by the persistence of lesions of an inflammatory type, should be associated with amyloidosis. In fact, Chapin, Scudamore, Bagenstoss and Bargen (1956) in an analysis of post-mortem material obtained at routine post-mortem on thirty-nine cases of regional enteritis found two cases with severe renal amyloidosis.

They suggest that this complication may be much more common than is generally recognized although it may not necessarily contribute to the patient’s death.

In view of the poor prognosis of a patient who develops manifestations of renal amyloidosis, Werther’s recommendation that this complication should provide an indication for further surgery seems reasonable. However, it will seldom be applicable as these patients have usually had severe prolonged enteritis with several previous operations.

Summary

Two patients with regional enteritis who developed renal failure due to amyloidosis are described. The literature is reviewed briefly and the hope expressed that this association will be recognized more frequently in the future.

Thanks are due to Dr. Aver Jones, under whom both these patients were admitted, for permission to publish these cases and for his help with the preparation of this paper.

REFERENCES


VILLOUS ADENOMA OF THE RECTUM WITH ELECTROLYTE DEPLETION, DIABETES AND HYPOGONADSIM

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VILLOUS tumour of the lower bowel has been recognised as a clinical entity for over a century; Quain used this descriptive term when writing in 1855. However it was not until 1954 that the association of a state of severe fluid and electrolyte depletion with villous adenoma of the rectum
Regional enteritis associated with amyloidosis.

C. S. Ogg

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