

A CASE OF ULCERATIVE COLITIS WITH AMYLOIDOSIS PRESENTING AS FEVER OF UNKNOWN ORIGIN

Pages with reference to book, From 112 To 113

Javaid A. Khan, Wasim Jafri, Hizbullah Sheikh, Ata Khan (The Aga Khan University Hospital, Karachi.)

Amyloidosis is a disorder of protein metabolism which is characterized by deposition of eosinophilic material intercellularly which has characteristic ultra-structural morphology and biochemical features. Amyloidosis may be primary but more often it is secondary to various inflammatory, neoplastic and suppurative conditions. Inflammatory bowel disease is an unusual cause for secondary amyloidosis. The incidence of amyloidosis in Crohn's disease varies from 1-8%^{1,2}; however it is extremely rare in ulcerative colitis. We describe a case which was admitted to determine the cause for fever of unknown origin (F.U.O.) which turned out to be a case of secondary amyloidosis due to ulcerative colitis.

CASE REPORT

A 37 years old lady was admitted in Aga Khan University Hospital for investigation of fever for the past 5 months. The fever was intermittent ranging from 37.2°C to 38.2°C. She also complained of anorexia and progressive weight loss (25 to 30 lbs in 5 months), and dry cough and had become amenorrhoeic 3 months prior to her admission in hospital. Her bowel habits were normal till 3 days before her admission, at that time she developed loose motions, the frequency of which was 2 to 3 per day. Two of the motions contained fresh blood. There was no previous history of bowel disturbance. Her past medical history was unremarkable apart from jaundice at the age of 10. During 5 months of her illness she had received various courses of antibiotics and three weeks before her admission she was put on antituberculous treatment.

ON EXAMINATION

She was anaemic. Her pulse was 120/min and BP 100/70mm Hg. There was no jaundice or lymphadenopathy. She had a normal chest and cardiovascular examination. Her abdominal examination showed no organomegaly. Rectal and vaginal examinations were normal.

INVESTIGATIONS

Investigations showed hemoglobin of 8.1 g/dl, blood film showed macrocytosis and anisocytosis and red cell indices revealed a microcytic hypochromic picture. ESR was 140 mm/hour. Her blood sugar and serum creatinine was normal but serum albumin was 1.0 mg/dl. She also had prolonged PT of 20 sec. (control 13 sec). Stool analysis was positive for occult blood but stool cultures were negative. Urine analysis showed + 3 proteinuria. 24 hour urine for protein was 4336 mg/24 hours. Urine, stool and sputum were negative for AFB. Bone marrow examination and culture was negative. Serum protein electrophoresis showed polyclonal gammopathy. Her chest X-ray, abdominal ultrasound, barium meal follow through and barium enema all were normal. Liver biopsy was done after correction of her coagulation profile with fresh frozen plasma. She had 2 further episodes of blood in stools therefore flexible sigmoidoscopy was performed upto 60 cms. Mucosa was found to be inflamed in patches with friability and contact bleeding, linear ulcers covered with pus were also seen, the inflammation appeared discontinuous. Multiple biopsies were obtained for histopathological diagnosis. Mucosal

biopsy showed active ulcerative colitis (Figure 1).

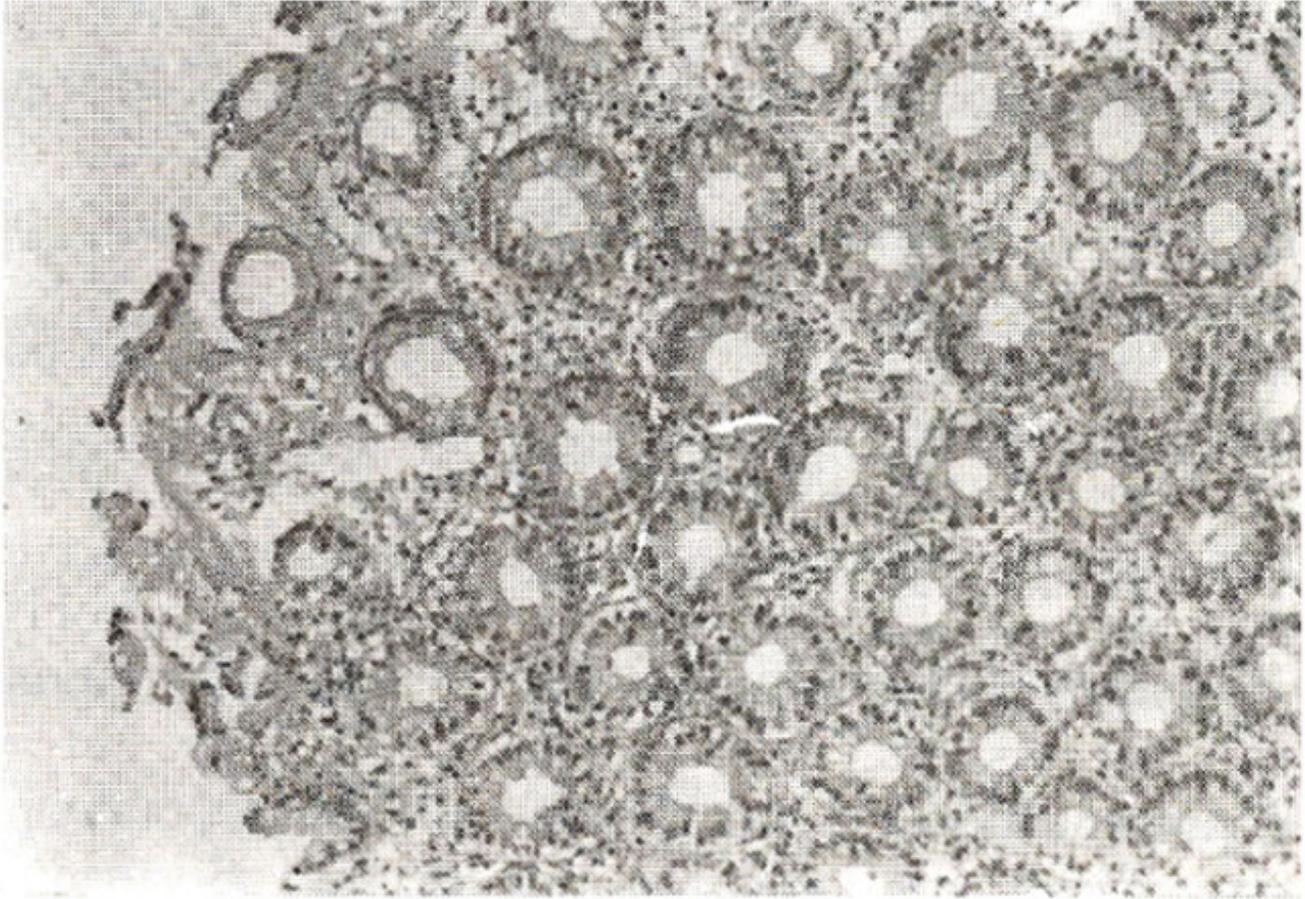


Figure 1. A cellular, homogenous amyloid (arrow) present in space of Disse and compressing sinusoids and liver cells. Needle biopsy H & E x 400.

Congostain of both hepatic (Figure 2)

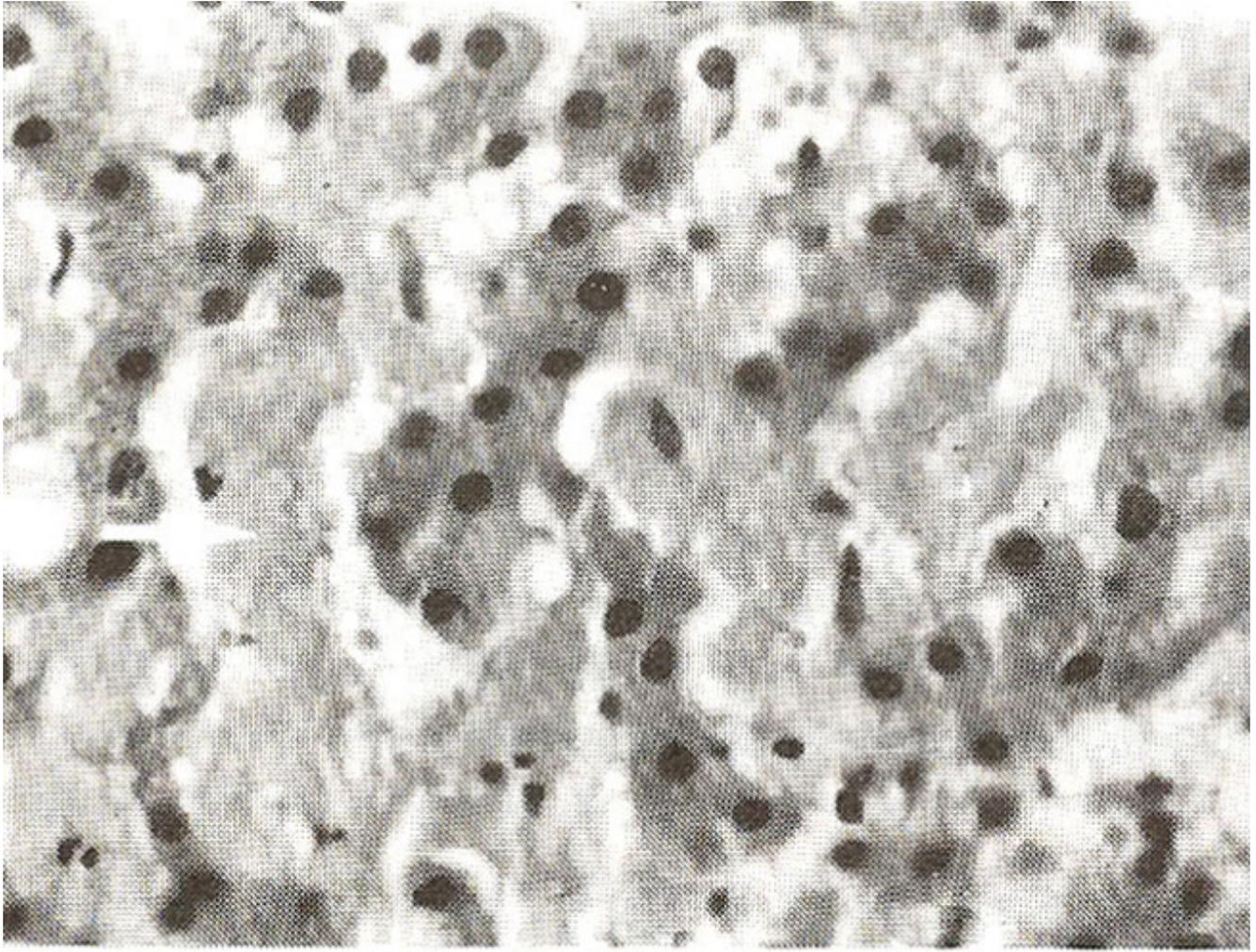


Figure 2. A cellular homogenous material (amyloid) in surface epithelium with inflammation in the lamina propria. H & E X 200.

and colonic (Figure 3)

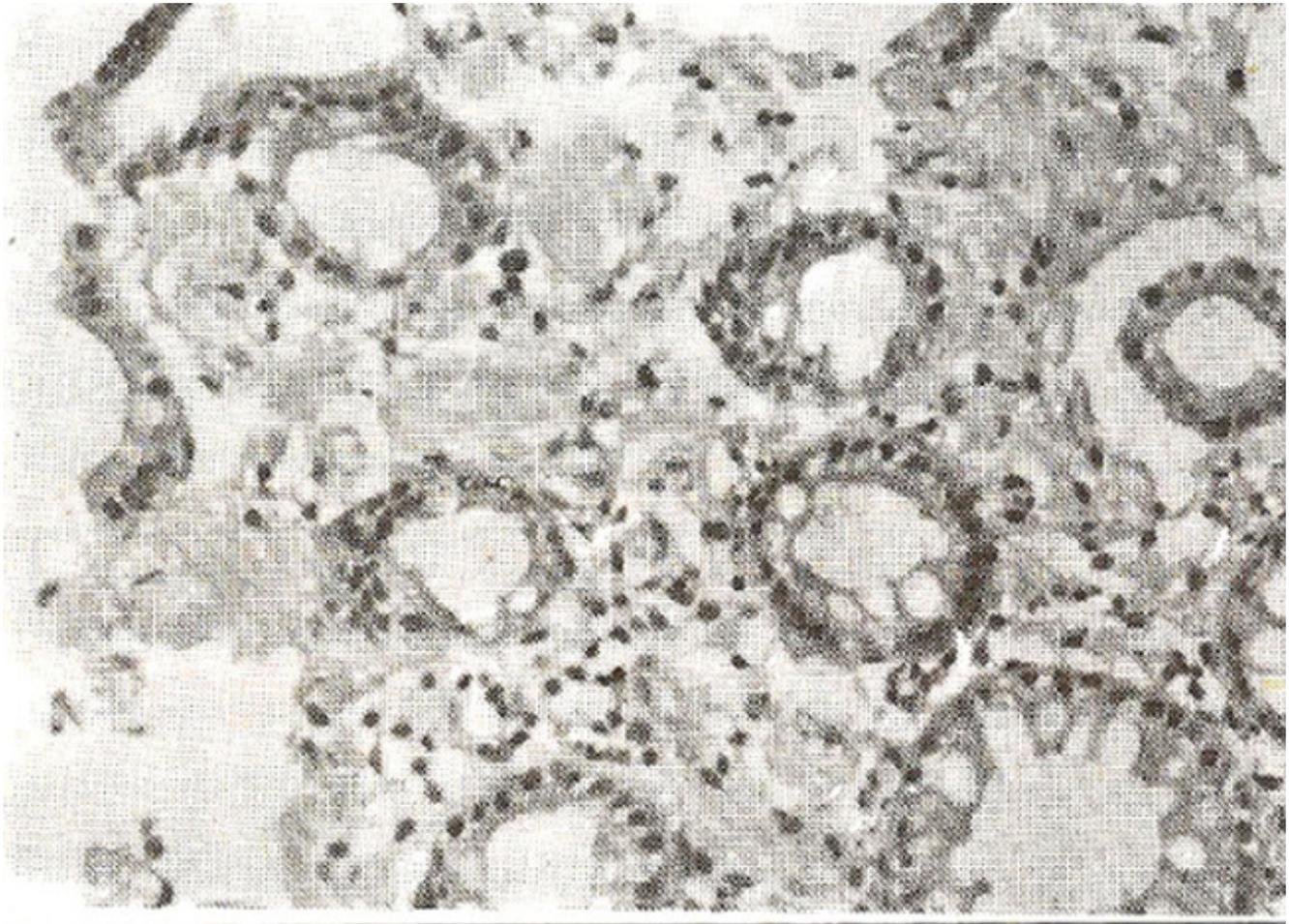


Figure 3. Amyloid material present as nodules and amorphous material. H & E X 400.

biopsies revealed the presence of amyloid. The patient was treated with blood transfusion, salazopyrine and steroids. Initially she responded to treatment but later relapsed. She progressively lost weight, continued to have diarrhoea and failed to respond to any treatment. She expired at home after 2 months of discharge. Because of her very poor general condition and severe nephrotic syndrome, surgery was not contemplated.

DISCUSSION

Inflammatory bowel disease (Crohn's disease and ulcerative colitis) may be complicated by secondary amyloidosis affecting the kidneys and rarely adrenal glands as well³. Amyloid may produce frank nephrotic syndrome and even frank renal failure^{4,5}. Patients who die because of complications of inflammatory bowel disease, amyloidosis may be an important cause of death, it was reported as 29% in one series⁶. Pen-intestinal suppuration and fistulization usually precede the development of secondary amyloidosis in ulcerative colitis. Primary disturbed immunologic mechanism has been postulated as responsible for both enteritis and the amyloidosis in Crohn's disease which may be true for ulcerative colitis as well. Clinical evidence of amyloidosis complicating inflammatory bowel disease either as nephrotic syndrome, hepatomegaly or splenomegaly, constitute an indication for resection of the diseased bowel⁶. Mild tubular dysfunction and non-specific glomerulitis has also been discovered at autopsy in patients with both acute and chronic inflammatory bowel disease. Hepatic

amyloidosis complicating inflammatory bowel disease usually Crohn's disease has been reported very rarely. Shorvan found 33 cases in literature upto 1977⁷ and Eade found amyloid deposits in 2 of 20 liver biopsies in patients with Crohn's disease⁸. Our case has demonstrated evidence of amyloidosis complicating ulcerative colitis in biopsies obtained from the colon and liver (Figure 2 and 3) although renal biopsy was not obtained but the evidence of nephrotic syndrome leaves no doubt that kidneys were also involved in secondary amyloidosis.

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