Severe Coombs positive autoimmune haemolytic anaemia associated with ulcerative colitis

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Summary: We describe a case of severe autoimmune haemolytic anaemia developing in newly diagnosed ulcerative colitis of moderate severity. Full immunosuppression with steroids and azathioprine failed to bring about a full remission and a splenectomy was performed which resulted in a remission enabling immunosuppression to be discontinued. This case is compared with three previously described cases of autoimmune haemolytic anaemia associated with severe colitis requiring splenectomy.

Introduction

Ulcerative colitis, a condition considered to have an immune aetiology, is frequently complicated by anaemia. This anaemia is usually ascribed to iron depletion secondary to haemorrhage, or to impaired haemopoiesis related to inflammation or infection, but rarely an autoimmune haemolytic anaemia can occur (Allan *et al.*, 1983). In a series of 624 patients with ulcerative colitis only one had a Coombs positive autoimmune haemolytic anaemia (Edwards & Truelove, 1964). Occasionally a severe anaemia can result; such a case is described here.

Case report

A 63 year old man presented with loose motions mixed with blood, opening his bowels four times a day. A diagnosis of ulcerative colitis was made on sigmoidoscopy, and treatment was commenced with sulphasalazine 0.5 g four times a day and hydrocortisone enemas. The haemoglobin was 11.0 g/dl, and ESR 70 mm in the first hour (Westergren).

Three months later he felt listless, the haemoglobin was 10.9 g/dl and a blood film showed spherocytes and polychromasia. The reticulocyte count was $0.19 \times 10^{12}/l$. The direct antiglobulin test was strongly positive (IgG mainly with some C3d; no specificity could be found). The sulphasalazine was discontinued and prednisolone 45 mg/d started and gradually tapered to 15 mg daily. A bowel relapse necessitated an increase to 30 mg/d.

One month later he became tired and dyspnoeic. He continued to open his bowels three to four times daily. His haemoglobin had fallen to 7.9 g/dl with a reticulocyte count of $0.33 \times 10^{12}/l$, total white cell count $8.8 \times 10^9/l$ with a normal differential, platelet count $610 \times 10^9/l$. Electrolytes and calcium were normal. Total protein 55 g/l, albumin 34 g/l, bilirubin $35 \mu\text{mol/l}$ ($2.0 \mu\text{g/dl}$), alkaline phosphatase 81 IU/l (normal 25–120 IU/l), lactate dehydrogenase 992 IU/l (normal 115–480 IU/l), aspartate transaminase 36 IU/l (normal 5–45 IU/l), alanine transaminase 36 IU/l (normal 5–42 IU/l), ferritin $657 \mu\text{g/l}$, B₁₂ 197 ng/l, normal folate $5.8 \mu\text{g/l}$, autoimmune antibody screen negative. Immunoglobulins: IgG 8.1 g/l, IgA 1.0 g/l, IgM 0.7 g/l. No monoclonal present. Stool culture was negative. Sigmoidoscopy showed active colitis with ulceration and pseudopolyp formation. Biopsies taken at 20 cm, 30 cm and 40 cm showed active colitis.

He was treated with prednisolone 90 mg daily, azathioprine 100 mg daily and blood transfusion. He continued to require transfusions every ten days, and after three weeks a decision to perform elective splenectomy was made. This was delayed by episodes of septicaemia and herpetic infection of the lips which were treated with the appropriate antimicrobials. The steroid dose was reduced and the azathioprine was stopped.

A splenectomy was performed six weeks after admission. He developed a pulmonary embolus post-operatively for which he was anticoagulated. He has not required any further blood transfusions since, and the dose of steroids has been gradually tailed off. Two months post splenectomy he was well, opening his bowels twice daily, the prednisolone having been reduced to 10 mg daily. The direct antiglobulin test

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remained positive and his haemoglobin had risen to 12.6 g/dl.

Discussion

Six cases have previously been described in which a splenectomy has been performed for severe autoimmune haemolytic anaemia associated with ulcerative colitis (Altman *et al.*, 1979; Lorber *et al.*, 1955; Keene, 1966; Lorst *et al.*, 1975). Other cases (Altman *et al.*, 1979) have responded to steroids alone, required azathioprine as well, or have had colectomy performed for bowel symptoms leading to a remission of haemolysis.

Of the splenectomy cases, two did not have colitis at the time (Keene *et al.*, 1966; Lorst *et al.*, 1975) and one with inactive colitis did not receive a trial of medical treatment (Lorber *et al.*, 1955). Three splenectomy

cases are thus described which had active colitis and received a trial of medical treatment (Altman *et al.*, 1979). In one case associated with severe colitis there was a total failure of medical treatment and splenectomy was combined with colectomy resulting a complete remission. A second case made a partial response to steroids, made a further response to splenectomy, but required azathioprine to bring about a complete remission. A third case, a relapsed colitic, made a partial response to steroids, and a complete response to splenectomy.

Our case had colitis of moderate severity and received a trial of full medical treatment, but unfortunately developed the complications of immunosuppression before a splenectomy could be performed. It is suggested that if autoimmune haemolysis persists, despite heavy immunosuppression early consideration should be given to splenectomy.

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