SHORT REPORTS

Acute anuric glomerulonephritis in monoclonal cryoglobulinaemia

While proliferative glomerulonephritis with progressive renal failure is a common and often fatal complication of primary mixed cryoglobulinaemia,¹ the presence of monoclonal cryoglobulins is not usually associated with glomerular proliferative lesions. Nevertheless, a few well-documented cases of glomerulonephritis in patients with essential monoclonal IgG cryoglobulinaemia have been reported.²⁻⁴ In these cases the outcome of glomerulonephritis was generally favourable or very slowly progressive.

We describe a further case of a woman with a primary monoclonal IgG cryoglobulinaemia who developed a diffuse proliferative glomerulonephritis. In this patient renal damage was particularly severe and prolonged anuria complicated the outcome. Despite the ominous prognosis attributed to acute renal failure in cryoglobulin-aemic nephritis, this patient's renal function recovered.

Case report

A 42-year-old woman was admitted to our unit in December 1972. In July 1971 she had had a normal pregnancy, and in February 1972 she developed skin eruptions on her legs and arms, without fever or other symptoms. The skin lesions spontaneously disappeared but reappeared in subsequent episodes. In December 1972, during one of these episodes, we found that she had proteinuria and hypertension. After a few days she became oliguric and oedematous, and on admission she was pale, febrile, with oedema and haemorrhagic-papulo-necrotic lesions of the legs. Blood pressure was 165/100 mmHg. Serum creatinine was 831 μ mol/l (9.4 mg/100 ml), blood urea 30.4 mmol/l (183 mg/100 ml), packed cell volume 28%, haemoglobin 8.9 g/dl, white blood count 8.3×10^9 /l, platelets 220×10^9 /l. Serum C3 was 0.43 g/l, and tests for ANF and LE cells were negative. Her proteinuria amounted to 6 g daily and haematuria was present. In the serum cryoglobulin was found with cryocrit of 48%. Both serum and cryoglobulin electrophoresis showed a M component in the gammaglobulin region, and immunoelectrophoresis disclosed a biarcuate band with anti-human-IgG and anti-human-k chain sera was detected. The cryoglobulin had rheumatoid activity. Skeletal x-ray examination and bone marrow aspirate showed normal results. Renal biopsy showed intense, diffuse mesangial proliferation with coarse fibrinoid subendothelial deposits, interstitial infiltration of mononuclear cells, and tubular swelling. A skin biopsy showed vasculitis.

The patient was treated with aspirin, cyclophosphamide, subcutaneous heparin, and peritoneal dialysis. After 30 days of oliguria, a diuresis occurred and renal function recovered slowly. In March 1973 she developed a new purpuric skin eruption, fever, fall of serum C3 levels, impairment of renal function, haematuria, and severe proteinuria (5 g daily). Azathioprine and two pulses of intravenous methylprednisolone were added to cyclophosphamide, heparin, and aspirin. Again, renal function rapidly improved and the other symptoms disappeared. Thereafter, on continued immunosuppressive and anticoagulant treatment, she remained well, though proteinuria (2 g daily) and monoclonal IgG cryoglobulinaemia persisted.

Discussion

In primary mixed cryoglobulinaemia the glomerular lesions and vasculitis are due to immune complexes, in which the cryoglobulin IgG is the antigen, and IgM the antibody. The same mechanism could operate in monoclonal IgG cryoglobulinaemia, in which part of the IgG consists of antigens and part of antibodies.⁵ In our patient the finding of mesangial proliferation, vasculitis, low concentrations of serum C3, and rheumatoid activity of the cryoglobulin were consistent with an immune complex disease.

In conclusion, this case confirms that diffuse proliferative glomerulonephritis is a rare but possible complication of essential monoclonal cryoglobulinaemia and that severe glomerular damage with acute renal failure may develop in this condition. On the other hand, the severity of the renal lesions and prolonged anuria do not necessarily preclude the recovery of renal function.

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Eosinophilic fasciitis

Shulman¹ first used the term eosinophilic fasciitis in 1974 to describe two men with thickening of the subdermal tissues, eosinophilia, and hypergammaglobulinaemia. He suggested that it was a separate entity from scleroderma in that the skin itself was normal, only the subdermal tissues being affected, and there was no visceral involvement.

Case report

In September 1971 a 28-year-old man gradually developed swelling and stiffness of the soft tissues of the wrists, forearms, and legs, later spreading to his trunk. In February 1972 he had fever and night sweats. In March he was admitted to the Hammersmith Hospital for investigation. The positive findings on examination were a temperature of 102° and diffuse induration of the subdermal tissues sparing the hands and face. There were no symptoms of Raynaud's phenomenon. The ESR was 46 mm in 1 h, eosinophil count 6.5×10^9 /l (6500/mm³), and immunoglobulin G and M levels were raised (IgG 28 g/l; IgM 0.3 g/l; IgE and IgA normal). Antinuclear antibodies were not found. Renal function, barium swallow findings, creatine phosphokinase levels, electromyogram, and lung function were normal. Skin biopsy showed an increase in subdermal collagen and a perivascular infiltrate of chronic inflammatory cells. Muscle biopsy showed connective tissue heavily infiltrated with aggregates of plasma cells, lymphocytes, and occasional eosinophils.

Although the nature of the connective tissue disorder was not defined prednisone 40 mg daily was started because of the widespread subcutaneous involvement. This resulted in considerable symptomatic improvement; the temperature returned to normal and the eosinophil count was lower. He was discharged on reducing doses of prednisone. In 1974 muscle stiffness and fever returned. Prednisone was increased to 40 mg/day and penicillamine therapy started. Again there was satisfactory response, which has been maintained despite reducing prednisone to 15 mg on alternate days.

Course of disease and treatment.

The patient is now very well and leading a normal, active life, though the subdermal tissues in his arms and legs remain bound down and tight. Skin biopsy shows no skin atrophy or loss of appendages but some subdermal fibrosis.

Comment

Since the initial description several authors have emphasised the distinctive features of eosinophilic fasciitis.^{2,3} While the appearances may be confused with scleroderma, the pattern of skin affection is unusual in that the hands, feet, and face are spared. Furthermore, the thickening and induration affect the subcutaneous tissues but not the skin itself. Raynaud's phenomenon does not occur. The blood eosinophilia is a striking accompaniment, with counts as high as $6 \times 10^9/l$ ($6000/mm^3$). Nevertheless, eosinophils are not often noted on histological examination of the skin, muscle, or subcutaneous tissues. Hypergammaglobulinaemia is prominent, though IgE levels are normal. Antinuclear antibodies are absent.

The histology of the skin and appendages is normal, without atrophy. There is a prominent increase in subcutaneous collagen with an associated inflammatory infiltrate composed largely of lymphocytes and plasma cells. The underlying muscles may be infiltrated but there is no evidence of muscle fibre necrosis. There seems to be no associated visceral involvement and the response to corticosteroid therapy is usually prompt and sustained. For these reasons the condition merits recognition as a separate entity from scleroderma.

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Involution of mucocutaneous pigmentation of the Peutz-Jeghers syndrome

The Peutz-Jeghers syndrome consists of mucocutaneous melanin pigmentation with gastrointestinal polyposis.¹ There may also be polyps in the respiratory and genitourinary tracts. There is a risk of gastrointestinal malignancy of 12.6% and an association with ovarian neoplasia.² As originally described, pigmentation develops during childhood and gradually fades with age. The growth rate of those polyps is variable, with a tendency for more rapid growth in younger patients.³ Large polyps have been described in a patient in whom there had been no radiologically detectable polyps seven months previously. At the other extreme, a solitary polyp appeared radiographically unchanged in size over 11 years. Polyps may slough off, diminish in size, or even disappear.³ Small bowel hamartomata have been described in the absence of mucocutaneous pigmentation.

Case report

A 43-year-old Jamaican woman presented in May 1975 with a year's history of recurrent abdominal colic and a six-hour history of severe abdominal pain. She was obese with pigmentation of the oral mucosa and on the volar aspect of the fingers. She had left hypochondrial tenderness, guarding, and rebound tenderness with high-pitched bowel sounds. Findings on rectal examination were normal. Plain x-ray examination of the abdomen showed dilated loops of small bowel with multiple fluid levels. Laparotomy confirmed ileocaecal intussusception with a pedunculated fleshy polyp at its apex. There were also three pedunculated polyps proximal to the intussusception with multiple ileal polyps of about 1 cm in height. The four polyps were excised through separate ileostomies. Histological examination

of the polyps showed hamartomatous lesions consisting of branching strands of smooth muscle and connective tissue compatible with the Peutz-Jeghers syndrome.^{1,4} At review of the patient 11 months later there was no evidence of mucocutaneous pigmentation. A double contrast barium enema, a barium meal, and a small bowel enema failed to show any evidence of gastrointestinal polyps. The patient's daughter also presented to us with similar pigmentation and iron deficiency and was found to have a single large bowel pedunculated polyp, which was resected via colonoscopy.

The mother and daughter have Peutz-Jeghers syndrome as confirmed by clinical history, physical examination, and histological appearances of resected tissue. Nevertheless, in the mother's case surgical resection of the large polyps resulted not only in the disappearance of the mucosal and digital pigmentation, but also in regression of the unresected small intestinal hamartomata.

Discussion

The disappearance of the mother's pigmentation occurred over 11 months. It is unlikely that this change was due to the regression that is associated with aging. All these findings therefore pose certain questions regarding the natural history of the syndrome. Firstly, is there a relation between fading of pigmentation and regression of polyps, and could this be explained by certain hamartomata secreting biologically active amines? Secondly, does the removal of the large polyps influence the growth rate of the small sessile hamartomas, as it appears to have done in this case? Finally, if disappearance of mucocutaneous pigmentation indicates regression of polyps, is the risk of malignancy therefore reduced in such patients?

In view of these suggestions has the daughter's colonoscopic resection had an unexpected therapeutic benefit? Should the daughter's pigmentation fail to regress over the same period of time, will further investigation disclose a large pedunculated polyp?

References

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Perforated jejunal diverticulum due to local iron toxicity

Jejunal diverticulosis is a rare acquired condition, developing progressively with age. The diverticula occur at the mesenteric border of the bowel, mainly where the vasa brevia pierce the muscularis mucosae. They are false diverticula, being lined with mucosa and muscularis mucosae only. The condition is usually asymptomatic. A straight x-ray film of the abdomen often shows fluid levels in the diverticula, which may be misdiagnosed as intestinal obstruction. Nevertheless, abdominal discomfort from gut dysfunction, malabsorption, intestinal obstruction, diverticulitis, haemorrhage, or perforation may occur. Perforation of a jejunal diverticulum is rare and fewer than 30 cases have been reported.¹ In most of these cases perforation was secondary to diverticulitis, though foreign body perforation has been recorded.^{2,3} The present case of jejunal diverticular perforation, however, appears unique.

Case report

A 74-year-old woman presented as an emergency with a 24-hour history of severe generalised abdominal pain of insidious onset but rapidly increasing severity. There was no history of indigestion or analgesic ingestion. Five