

## LETTERS TO THE EDITOR

### Polymyositis and cyclosporin A

Sir: Polymyositis is an inflammatory disease of striated muscle of unknown cause. When it is accompanied by the characteristic skin lesions the disease is called dermatomyositis.<sup>1</sup> Although controlled studies have not been carried out, it seems that corticosteroids are beneficial, particularly in acute polymyositis/dermatomyositis.<sup>1,2</sup> Certain patients, however, need very high doses or do not respond to this treatment. Such patients have been treated with a wide spectrum of immunosuppressive agents, alone or combined with steroids, with variable response.<sup>2</sup> Successful treatment with cyclosporin A in acute polymyositis/dermatomyositis has been reported,<sup>3-5</sup> but long term follow up of these patients is lacking. We present a report of three patients with polymyositis/dermatomyositis,<sup>1</sup> in whom cyclosporin A treatment was tapered after their recovery from an acute bout of the disease.

#### CASE 1

A 46 year old woman presented with progressive weakness in hip and shoulder girdles, arthritis, Raynaud's phenomenon, and heliotrope rash. A diagnosis of type II dermatomyositis was established by clinical, laboratory, electromyographic, and histopathological studies. Studies to rule out neoplasia were negative. The patient started treatment with methylprednisolone (1 mg/kg/day), with partial clinical improvement. Five months later the patient's clinical status worsened and she developed Cushingoid changes. Azathioprine was added (2 mg/kg/day), without clinical improvement. This treatment was then stopped and cyclosporin A treatment was started (5 mg/kg/day). A good clinical response was obtained during the first week. Two months later she suffered a new bout related to a voluntary withdrawal of cyclosporin A treatment. Treatment was restarted and muscle strength recovered again completely. Cyclosporin A was stopped 10 months later. After remaining asymptomatic for more than 17 months the patient was readmitted because of severe dyspnoea and cervical lymphadenopathy. An interstitial pattern was noted on a chest radiogram. Six days later the patient suffered a fatal pulmonary thromboembolism. A lymph gland biopsy that had been performed some days before disclosed the presence of a metastatic adenocarcinoma.

#### CASE 2

A 48 year old woman presented in September 1984 with weakness in both girdles, fever, and heliotrope rash. A diagnosis of type II dermatomyositis was established by clinical, laboratory, electromyographic, and histopathological studies. Neoplasia was ruled out. Treatment with methylprednisolone (1.5 mg/kg/day) was started. Three months later, after no clinical response, the patient developed Cushingoid signs and upper gastrointestinal bleeding secondary to duodenal ulcer. Previous treatment was then stopped and treatment with cyclosporin A (7.5 mg/kg/day) was started. Fever and rash disappeared in a few days, and muscle strength recovered completely. Cyclosporin A treatment was stopped in July 1986. From that moment and so far the patient has remained asymptomatic.

#### CASE 3

A 53 year old woman was admitted in March 1983 to our hospital because of oedema in eyelids, hands, and malleolar regions, stiffness, Raynaud's phenomenon, progressive dyspnoea, telangiectasia, and proximal sclerosis. Oesophageal manometry showed impaired motility. Antinuclear antibodies were positive (1/6400) with a nucleolar pattern. A diagnosis of progressive systemic sclerosis was made. In May 1985 she was readmitted because of severe weakness of both girdles. Electromyographic and histopathological studies showed myositis. Corticosteroid treatment (1 mg/kg/day) was started, without clinical improvement. Treatment with cyclosporin A was then started (5 mg/kg/day), with muscle strength recovery. This treatment was stopped in January 1987. From then the patient has been asymptomatic.

Polymyositis/dermatomyositis left to its spontaneous evolution has a high mortality, estimated to be more than 60%.<sup>6</sup> Most authors agree that steroids are useful in the treatment of acute forms.<sup>2</sup> Failure to respond to steroid treatment occurs in 25-50% of patients, however.<sup>7</sup> In this 'steroid resistant' group other immunosuppressant treatments, including cyclosporin A, have been tried, with variable results.<sup>2-8</sup>

Cyclosporin A is a peptidic drug with immunosuppressant activity that interferes with the synthesis and release of lymphokines from the T helper subset.<sup>9</sup> Some authors agree with the usefulness of cyclosporin A in the acute forms of polymyositis/dermatomyositis,<sup>3-5</sup> but its effectiveness in chronic polymyositis is not clear. Cyclosporin A was given to all three patients because of failure or severe side effects of conventional treatment. A good correlation was found between clinical improvement and cyclosporin A administration in all the patients.

Side effects that might be attributed to cyclosporin A treatment in our patients are hirsutism and tremor in all of them, and mild hypertension with moderate impairment of renal function in patient No 1. All these abnormalities, very common in patients receiving cyclosporin A,<sup>10</sup> were reversed after the drug was tapered off. These few cases show that cyclosporin A can be useful in patients with acute forms of polymyositis/dermatomyositis. Suppression of cyclosporin A treatment in polymyositis/dermatomyositis can produce a new bout of the disease, but probably in some patients the cyclosporin A treatment given for an indefinite period might stop the activity of the disease even after withdrawal of the treatment for many months. Thus cyclosporin A may be the treatment of choice when conventional immunosuppressant therapy fails or when adverse effects of this treatment are important. More studies are needed to corroborate these clinical observations.

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### Toxic shock syndrome associated arthropathy

Sir: We read with interest the recent paper by Foley-Nolan *et al*,<sup>1</sup> which states 'the only previous report of a patient with the toxic shock syndrome and associated arthritis was a 15 year old girl'.

We previously reported arthritis as a manifestation of toxic shock syndrome<sup>2</sup> and would draw this to the attention of the author and your readers.

This recalls a couplet which is apt:  
When I am dead, I hope it may be said:  
'His sins were scarlet, but his papers were read.'  
(After Hillaire Belloc, *On His Books*.)

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### Destructive arthropathy after successful renal transplantation

Sir: In their recent article Duncan *et al* reported two patients treated for chronic renal failure, who developed a severe erosive arthropathy at a relatively young age.<sup>1</sup> Case 2 had slight degenerative changes of both hands after two years of haemodialysis. Eleven years after successful renal transplantation he had developed a severe erosive arthropathy. We report here a patient with chronic renal failure who developed her first joint complaints and later a destructive arthropathy after successful renal transplantation.

A 52 year old white woman attended the outpatient clinic of the department of rheumatology in 1983 with a six month history of pain and swelling of the hands. In 1968 she developed renal failure secondary to chronic pyelonephritis. She underwent haemodialysis from 1971 to 1976 and then received a renal graft. After initial problems, for which she needed antirejection treatment on four occasions, the graft functioned well (mean creatinine clearance 86 ml/min). There was no family history of osteoarthritis or psoriasis.

On examination she had a synovitis of the interphalangeal joint of both thumbs, the proximal interphalangeal joints of the right middle finger and left ring finger, and the distal interphalangeal joint of the left index finger. All other joints were unremarkable. Radiographs of her hands showed soft tissue