

DISCUSSION

DCM is a rare condition. A literature review of DCM with neonatal onset is shown in Table I. This form of DCM is more frequent in males and blistering is common. The prognosis appears to be unfavourable compared with disease onset after the neonatal period, when a good prognosis is expected (7).

The present patient probably has a good prognosis, as illustrated by the clinical improvement and the regression of both cutaneous mast cell infiltration and hepatomegaly. The favourable prognosis may be related to the disease onset occurring late in the neonatal period.

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Dermatomyositis Induced by Penicillamine

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Abstract. A case of dermatomyositis is reported in a 50-year-old woman receiving D-Penicillamine therapy for rheumatoid arthritis. There was no evidence of neoplasia on full investigation. Remission of dermatomyositis occurred on withdrawal of D-Penicillamine.

Key words: D-Penicillamine; Rheumatoid arthritis; Dermatomyositis

D-Penicillamine has been used successfully in the treatment of Wilson's disease and is being used with increasing frequency in the treatment of rheumatoid arthritis and other 'collagen' diseases. Side effects are unfortunately frequent and have been well reviewed (5).

Less common side effects include the development of other auto-aggressive disorders: Systemic lupus erythematosus (3), myasthenia gravis (2), and polymyositis (1). There has been a single case report of dermatomyositis (4) and we report a second case.

CASE REPORT

A 50-year-old housewife developed a sero-positive rheumatoid arthritis (Rosewaaler 1:512) in April 1976. Treatment with D-Penicillamine was started at 250 mg/day and continued with satisfactory clinical improvement on that dosage. After 19 months of treatment the patient complained of a month's history of tenderness, redness and swelling of the fingers and knuckles of both hands and also redness and puffiness of her face and eyelids. Examination revealed a dusky violaceous discoloration of her eyelids and forehead; dilatation of nail fold capillaries on all fingers with a dusky violaceous discoloration of the skin overlying all metacarpophalangeal joints, and extending along the dorsal aspect of the fingers to overlie the first inter-phalangeal joint. There was no clinically detectable muscle weakness.

Investigations

ESR was 5 mm in 1 hr. Antinuclear factor 1:10, 24-hour urinary creatine 1920 $\mu\text{mol}/24\text{ h}$ (normal <380). Other investigations including haemoglobin, full blood count, serum transaminases, creatine phosphokinase and elec-

tromyography were normal. Chest X-ray, barium meal, barium enema, intravenous urogram and mammography were normal. Pelvic examination by a gynaecologist was also normal. Skin biopsy showed a vasculitis with overlying sub-acute dermatitis and direct immunofluorescence demonstrated positive immunofluorescence for IgM and C₃ in superficial vessels and patchily along the dermo-epidermal junction. Muscle biopsy was not performed.

All therapy was stopped and over the next 2 months the rash gradually faded and a feeling of well-being returned; at no time was there a clinically evident weakness of muscles, but 24 h urinary creatine levels remained elevated at 1140 µmol even after 9 months.

DISCUSSION

This 50-year-old lady presented with the characteristic rash of dermatomyositis and biochemical (but not clinical) evidence of myopathy. Muscle biopsy was not performed in our patient because of the normal electromyograph and lack of muscle symptoms. The diagnosis of myositis in this patient is based on the elevated level of creatine in the urine. Several authors (7, 8) cite this test (urinary creatine) as the best index of disease activity. Serum glutamic oxaloacetic transaminase (SGOT), serum aldolase and serum creatine phosphokinase may also be elevated, but some patients continue to show activity in the presence of normal levels (7). We cannot explain why our patient's urinary creatine is still abnormal, while the dermatological features have resolved. It could be suggested that she has both rheumatoid arthritis and polymyositis. The association between these two disorders has been established (6), but none of the patients in this series had dermatological manifestations.

There has been one previous case report of dermatomyositis occurring during D-Penicillamine therapy (4) though there are several differences between the two cases. The patient concerned received up to five times the daily dosage taken by our patient, she developed a high titre of anti-nuclear factor and required systemic therapy with Prednisolone to induce a remission of dermatomyositis.

We suggest that our patient had both rheumatoid arthritis and dermatomyositis and that, because of the rapid remission of the rash on withdrawal of D-Penicillamine, the drug was the causative factor, especially as investigations for neoplasia and an 18-month follow-up have revealed no other associated conditions.

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Local Photochemotherapy in Nodular Prurigo

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Abstract. Fifteen patients with nodular prurigo were treated with a trioxsalen bath (50 mg/150 l of water) and UVA given in an ordinary PUVA cabin. Good results were observed in 8 and moderate in 7 patients in the initial phase. The result was good or excellent in 13 of the 15 patients during maintenance treatment. The good results are promising in the treatment of this chronic and otherwise therapy-resistant disease.

Key words: Nodular prurigo; Itching; Photochemotherapy; Trioxsalen; PUVA

Nodular prurigo (syn. Prurigo nodularis Hyde, Lichen cornuus obtusus) is seen most often in middle-aged women. The exact cause and mechanism of this fairly rare disease are unknown. It may be some kind of cutaneous neurosis with intense itch-