CALCINOSIS OF HAND AND AXILLA IN POLYMYOSITIS/DERMATOMYOSITIS

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INTRODUCTION

POLYMYOSITIS is an autoimmune disease which may present in many ways (Bradley, 1977). As a result, various classifications have arisen over the years (Walton, 1958, Currie, 1971, Pearson, 1972). This reflects the variability in clinical and pathological findings. Classification provides a guide to prognosis and the response to therapy (Pearson, 1971). The first criteria for the diagnosis was by De Vere (1975). The clinical criteria were muscle weakness, usually proximal and symmetrical, as well as muscle pain and tenderness, while the laboratory criteria included a muscle biopsy showing perivascular inflammatory cell infiltration with or without muscle degeneration, electromyography findings, and raised serum creatinine.

Earlier on (Rose and Walton, 1966) the grading system used for disability was done. Unless the criteria are specified, many clinical studies may be contaminated with a wide spectrum of neuromuscular disorders, denervating disorders, metabolic and connective tissue disorders. We would like to report a patient who initially presented with what looked like a metabolic disorder.

CASE REPORT

Patient, 15 year old male, referred from the Orthopaedic Clinic with one year history of painless whitish lumps over the left hand and the right axilla which progressively increased in size. The swelling, sometimes, flakes off. He dis not have any joint pains, haematuria nor abdominal pain.

About the same time, he experienced difficulty in getting up from the squatting position. He had no difficulty in lifting his hands. With this he noticed wasting of the shoulder girdle and hip girdle muscles. Initially the muscles were painful but in the last six months this has improved. There was some weight loss. Bowel habits were normal, so was his swallowing.

His gait was unsteady for five years and he noticed that his feet were deformed. Examination revealed periorbital oedema without heliotropic rash. No oedema was detected. Patient was thin.

There were minute ulcers at the tips of the fingers, otherwise there was no deformity. A whitish, non-tender calcinosis of about two centimeters diameter was seen and felt at the first metacarpo-phalangeal joint of the left hand (Fig. 1). No wasting of the small muscles of the hand was detected. There was loss of crease on the dorsum of the fingers. The nails showed pitting and looked fragile. Calcinosis of similar size was detected in the right axilla (Fig. 2).

Fig. 1. Calcinosis of left hand.

Neck muscle was of normal power. There were bilateral symmetrical wasting, with tenderness, of the muscles around the shoulder and hip joints. No particular distribution of wasting was noted. No was there any fasiculation. The weak muscles were of grade 3. Reflexes were slightly diminish in the upper limbs but those in the lower limbs were more pronounced. The plantars were down going.

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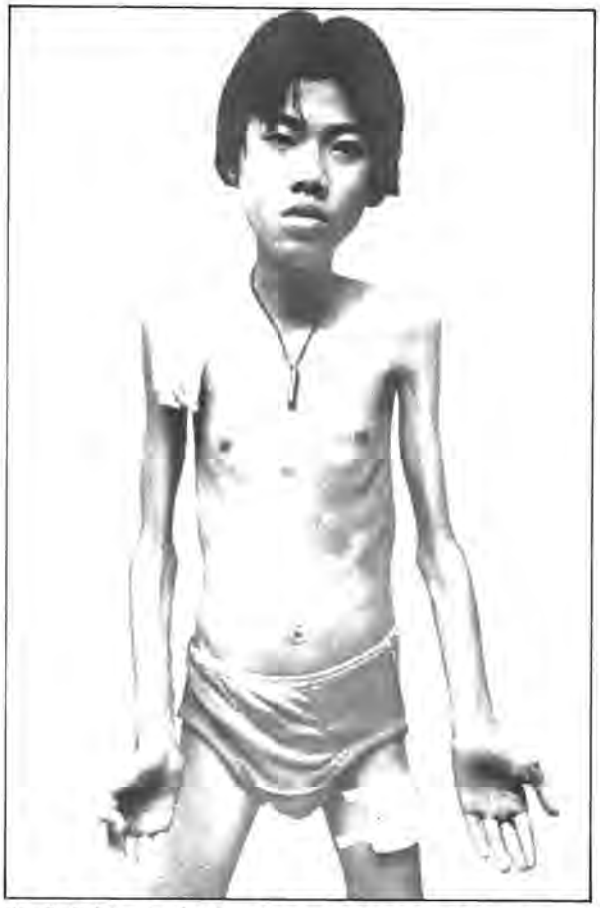

Fig. 2. Calcinosis of right axilla and left thigh which is covered by dressings.

Sensations were intact. His gait was waddling. There was talipes equinus varus and pes cavus bilaterally.

Laboratory investigations revealed a raised serum creatinine phosphokinase, while the electromyography showed short polyphasity, fibrillations, and positive short waves. Biopsy of the deltoid muscle of the right arm showed chronic inflammatory changes. X-rays of the spine showed spina bifida occulta. Investigations for metabolic disorders such as serum calcium and phosphate, fasting lipids and serum uric acid and for connective tissue disorders such as antinuclear factor and L.E. cells revealed no abnormal findings.

He was put on high dose of Prednisolone initially and his muscle power improved from grade 3 to grade 4. He is now on a lower maintenance dose of Prednisolone. However, his calcinosis remained static. So were his talipes equinus varus and pes cavus.

DISCUSSION

Our patient was about 14 years old when he developed his first episode of polymyositis. The age distribution is a bimodal curve with peaks in the groups from 5 to 15 years and 50 to 60 years of age. (Bohan, 1975). The distribution of muscle weakness though proximal in two-thirds of patient, is diffuse in pattern and does not show the specific involvement in some groups of muscles and sparing of others as in muscular dystrophies (Bradley, 1977). This case showed similar features except that the lower limbs were also involved distally with talipes equinus varus and pes cavus. X-ray of the spine revealed the presence of spina bifida occulta. Because of his walking with a waddling gait and had difficulty in getting up from squatting (Grade 5 disability, Rose, 1966), he had proximal weakness. Probably his feet deformities were the result of the spina bifida occulta though there is tendency for polymyositis to develop atrophy and contractures in children (Pearson, 1972).

Raised serum creatinine phosphokinase is not confined to polymyositis but also occurs in muscle dystrophies (Bradley, 1977). However, it is important to think of polymyositis when faced with a problem of muscle weakness since its presentation need not always be classical.

The presence of calcinosis may imply a metabolic disorder such as gout or hyperlipidaemia. Gout, though often presenting with joint pains, is sometimes associated with muscular pains. Calcinosis occur in about 4% of cases of dermatomyositis and it is more common in children (De Vere, 1975). The patient showed some improvement in muscle weakness when treated. Children show good recovery and become asymptomatic three to five years later. By eight years, the disease burnt itself in 80% of the cases.

Hence polymyositis is a treatable condition and responds well to Prednisolone. Therefore the condition should be recognised early in order to avoid contractures. This is more so in children as the response is better and contractures occur more frequently.

SUMMARY

A fifteen year old boy presented with a year's history of weakness and wasting of hip and shoulder girdles with wasting of the distal muscles of the legs associated with talipes equinus varus and pes cavus. He had calcinosis of the left hand and right axilla. Clinical examination and laboratory investigations revealed that he had polymyositis with calcinosis and spina bifida occulta. His muscle weakness responsed to treatment with Prednisolone. This condition may be mistaken for muscular dystrophies or metabolic disorders.

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