DERMATOMYOSITIS WITH CALCINOSIS CUTIS UNIVERSALIS

S. L. WADHWA * LESLIE MARQUIS †

Summary

A eleven year old child with dermatomyositis and calcinosis cutis universalis is presented. She showed heliotrope bloating of eyelids, Gottrons sign, proximal muscle wasting with contractures and extensive areas of calcification over the shoulder, pelvic girdles and proximal extremities. The diagnosis was confirmed by biochemical and histopatholagical studies as well as elctromyography. High doses of steroids and supportive measures made the patient ambulatory.

Dermatomyositis is a necrotising disease of striated muscle associated with a variety of inflammatory cutaneous lesions. Proximal muscle groups are commonly involved in a symmetrical fashion. The incidence of dermatomyositis is twice as common in women as in men. About 25% of those afflicted are less than eighteen years at the time of onset of the disease. Dermatomyositis is grouped with collagen disorders with which it shares rheumatic symptoms, cutaneous features and serologic abnormalities. Although malignancy is associated with adult dermatomyositis in a few cases, such association in childhood dermatomyositis seems uncommon[1].

Calcinosis cutis, a major complication of this disease develops more frequently in children than in adults and is related directly to the degree of muscle necrosis[2]. This may occur focally or diffusely in the skin, fascial planes and muscles. The term universalis denotes widespread involvement[3]. The extensive calcium deposits not only cause limitation of movements but extrusion of the calcium may lead to bacterial cellulitis, abscess formation, ulceration and scarring. Mortality in this disease is often related to respiratory insufficiency, cardiac failure and aspiration pneumonitis.

A clinical diagnosis is made mainly on the basis of proximal symmetrical muscle weakness associated with characteristic skin lesions. The diagnosis is confirmed by the presence of elevated serum enzymes, electromyographic changes and histological changes in muscle biopsy.

Case report

An eleven year old female child was admitted to the skin ward of the B. Y. L. Nair Hospital with the complaints of generalised weakness, restriction of movements of all four limbs and skin lesions. She was an active, healthy, playful child till two years before when she developed low grade

* Lecturer

† Prof., Dermatology & Venereology, T.N. Medical College
Hon. & Head, Department of Dermatology and Venereology,
B.Y.L. Nair Charitable Hospital, Bombay.

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fever, anorexia, generalised weakness and easy fatiguability. The muscle weakness and pain gradually increased leading to restriction of hip and shoulder movements. This was accompanied by the appearance of swelling and discolouration around the eyes and knuckles and discharge of chalky white material from the bony prominences in turn leading to painful ulcerations, contractures and further restriction of movements.

Examination revealed a febrile, markedly cachtic child. The skin around the eyes showed periorbital edema with violaceous hue - heliotrope bloating (Fig. 1A.) The dorsal interphalangeal joints of both hands showed flat topped violaceous papules– Gottron's sign (Fig. 1B.) Gritty, chalky white material was seen extruding from the shoulders, axillae, elbows and knees with ulceration and scar formation (Fig. 2A.) Contractures and flexion deformities of varying degrees were present at the shoulder, elbow, wrist, hip, knee and ankle joints. There was generalised wasting and tenderness of all the muscles, particularly the muscles of shoulder and pelvic girdles.

The blood counts were within normal limits except the E.S.R. which was 100 mm/hr. Additional investigations and results were total serum proteins 7.8G%, alb. 4.36G%, glob. 3.44 G%, α_1 0.63 G%, α_2 0.85 G%, β 0.81 G%, γ 1.15 G%. Serum calcium, inorganic phosphorus, alkaline phosphatase, serum enzymes (SGOT, aldolase, CPK, LDH), 24 hour creatine excretion and E. C. G. were within normal limits. The L.E. cell test and R.A. test were negative.

Skin biopsy on H & E stain revealed an atrophic epidermis, homogenisation of collagen fibres and deposition of calcium in the subcutaneous tissue.

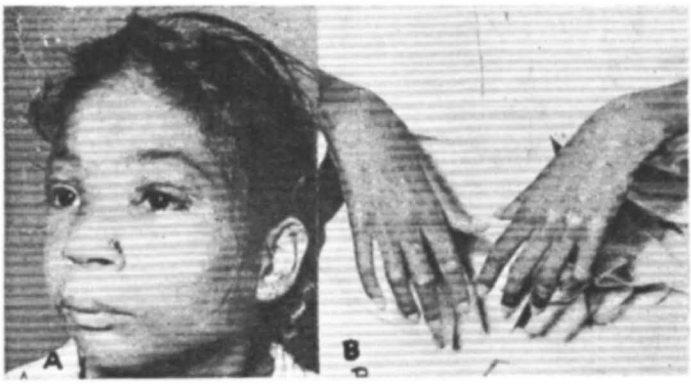

Fig. 1

(A) Heliotrope bloating

(B) Gottron's Sign.

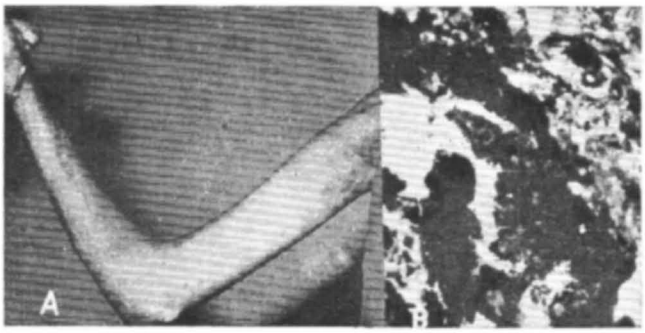

Fig. 2

(A) Extrusion of Calcium with resulting ulceration. Scar formation over wrist, elbow and axilla.

(B) Calcium stained black by Von Kossa Stain.

The presence of calcium deposits was confirmed by Von Kossa stain (Fig. 2B.) Muscle biopsy from the quadriceps showed vacuolar degeneration with loss of transverse striations. In some areas there was sclerosis and atrophy and fibrotic connective tissue replacing muscle bundles.

Roentgenograms showed widely distributed extensive sheets of calcification in the skin, subcutaneous tissue and muscular and fascial planes around the shoulder and pelvic girdles and proximal extremities (Fig. 3 & 4). Roentgen ray examination of chest and oesophagus revealed no abnormality. Electromyographic studies from the affected muscles showed presence of denervation, pseudomyotonia, reduced size of motor units, and interference pattern in very weak muscles. These findings were compatible with those in dermatomyositis.

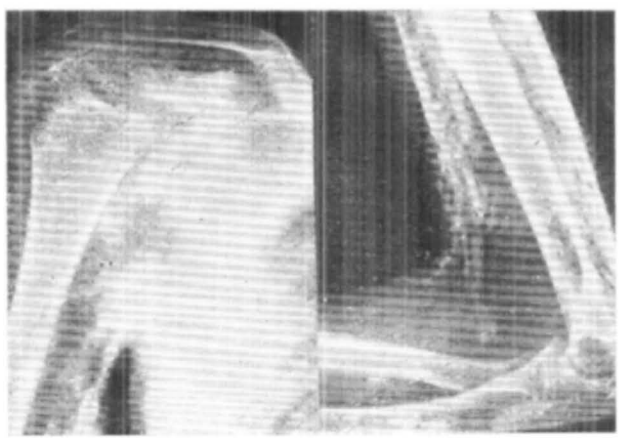

Fig. 3 X-rays of shoulder & elbows regions showing extensive linear plaques of calcification in various tissue planes.

The patient was put on tab. Prednisolone 80mg/day in divided doses along with general supportive and ancillary measures. Within two months of therapy the child showed remarkable improvement. She regained her appetite and started to take interest in her surroundings. The muscle pain and weakness improved and the calcium extrusions were less distressful. The steroid dosage was then reduced and physiotherapy initiated. This child who was completely incapacitated and bedridden at admission was able to perform her elementary chores.

Discussion

In our patient who presented with bilaterally symmetrical proximal muscle weakness and pain, heliotrope bloating, Gottron's sign and extensive calcinosis cutis, a clinical diagnosis of dermatomyositis with calcinosis cutis was made. The usual diagnostic tests employed for other connective tissue are of little value in dermatomyositis. Apart from muscle biopsy and electromyography, determination of serum enzymes has been considered as most useful in diagnosis and evaluation of activity in dermatomyositis. However, since in our patient the disease had been smouldering for a long time, the serum enzyme levels were within normal limits, high lighting the fact that the value of serum enzyme determination in diagnosis has been felt to be limited when the disease becomes chronic. This has been attributed to replacement of muscle with connective tissue and fat (or calcium salts in our case) resulting in a reduction in parenchymal muscle tissue and as a result, decrease in the extent of serum enzyme elevation[4].

The disease was present for over a year before calcification became evident clinically. The serum calcium and phosphorus values were normal. It is obvious that calcium deposition occured in a diseased tissue and thus is of a secondary nature.

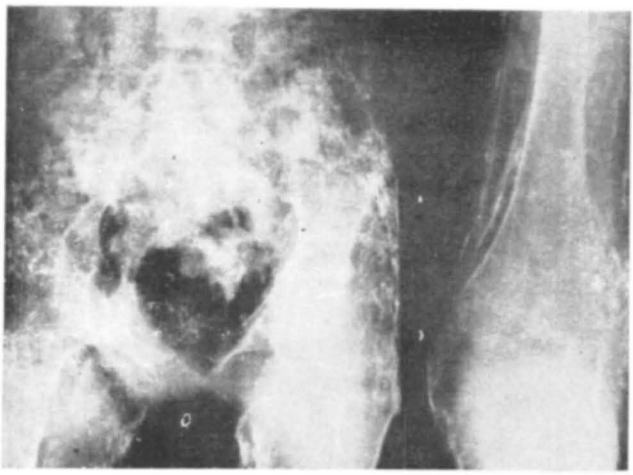

Fig. 4 X-rays of pelvis and knee showing widespread calcification of soft tissue of upper thighs and knee.

A greatly increased incidence of calcinosis cutis in females is observed in dermatomyositis as well as in scleroderma[5]. This would seem to indicate some hormonal influence in the initiation of calcification as is known to occur in the avian egg laying cycle[5].

Calcinosis usually occurs in muscles showing maximum involvement. The increased incidence in children may thus be due to a severe degree of muscle destruction. It may be due to an increased tendency to calcification during active growth period[9]. Calcification is usually progressive over a period of several months and thereafter stops. Ulceration with extrusion of calcified debris occur periodically, particularly in children. This is probably due to the large amounts of calcium deposits in muscles and associated muscle necrosis. Calcinosis has a poor outlook with respect to complete functional recovery since such dystrophic calcification is not amenable to therapy.

Steroids form the mainstay of therapy. Although these do not appear to shorten the duration of the disease, these reduce the incidence of complications and facilitate remission. Steroids should be started early and in adequate doses. Subsequently the dose may be reduced depending upon clinical response and a fall in serum enzyme levels. Maintenance therapy must be continued for upto 5 years or more in most cases[6]. Systemic steroids do not apparently affect calcinosis cutis. Chelation therapy with EDTA[7], Sodium versenate[8] and EHDP[9] in patients with calcinosis cutis universalis has not produced consistent clinical response. Excision of calcified deposits may benefit in selected cases and may provide relief from pain, infection and cellulitis. Regards those patients who fail to respond to even large doses of steroids, cytotoxic agents such as methotrexate[10] and azathioprine[11] have given favourable results in isolated cases. When muscle strength has returned and signs of inflammation have subsided, physical therapy should be instituted as vigorously as is feasible to improve muscle performance and prevent contractures[12,13].

Acknowledgement

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Announcement

Pediatric Dermatology Seminar IX

The 9th annual Pediatric dermatology seminar will be convened at the New Carillon Beach Hotel, Miami Beach, Florida, February 25th - 28th, 1982. The seminar fee remains at $ 190. A two week post - seminar tour to East Africa is planned. The cost will be approximately $ 2000 per person.

For information, contact: Guinter Kehn, MD (305/552/8600) 16800 NW 2 Ave., Miam, Florida 33169