Calcinosis Universalis Complicating Dermatomyositis : A Case Report

SUMMARY

Calcinosis Universalis is a rare complication of dermatomyositis. Dermatomyositis is a connective tissue diseases, and they occur infrequently in Nigeria. We hereby report a female patient who had both, so as to heighten the index of suspicion among practitioners.

KEY WORDS: CALCINOSIS, *Soft Tissues, Dermatomyositis.*

INTRODUCTION

Collagen vascular diseases occur infrequently among Nigerians (1) Dermatomyositis is one such disease. We hereby present the case of an adult female patient whose dermatomyositis was complicated by calcinosis universalis. It is to our knowledge the first reported case in our part of the country.

Case Report

RA is a 39 year old female who was referred to JUTH in 1991 for symmetrical proximal and distal inter phalangeal joint swelling, stiffness, body aches and hypopigmented rashes which started when she was 25 years old. She was then diagnosed as a case of dermatomyositis and referred to us for continuation of management. She was maintained on oral Celestone 0.5 mg daily and stable.

This treatment was continued since she was stable. While on this, she intermittently complained of transient one sided headache as well as transient chest pain worsened by food and exercise. The pain usually passed off spontaneously but at times required analgesics to subside. She also complained of early satiety, dysphagia and constipation. On examination, she looked asthenic, had thin dry skin as well as sub cutaneous calcinosis with ulcers and scars. She also had proximal myopathy in the lower limbs.

Sometime in 1995, she could no longer tolerate oral steroids and had to be changed to Depomedrol. This was able to hold down the symptoms at 80mg 6 weekly. She was on this up till 1997 when she insidiously developed a sinus behind the right elbow which discharged a chalky material. This was associated with fever, pain and limitation of movement in the affected limb. The upper arm and shoulder girdle muscles were tense and warm. She was thought to have developed infection in her subcutaneous calcinous ulcer with muscle involvement. Chronic osteomyelitis with surrounding soft tissue involvenient was also considered.

Investigation results returned as follows: PCV - 36% (low), WBC - 13,300/mm3 (high) with 79% neutiophils. ESR - 127mm/hr (westergren) and peripheral film showed toxic granulations. Both her serum albumin and calcium were low. Her renal functional status was normal. X ray showed soft tissue opacification in the upper arm. There was no evidence of osteomyelitis (fig 1). Though swab of the discharge grew Staph aureus, no pus was got on diagnostic aspiration of the boggy upper arm muscles.

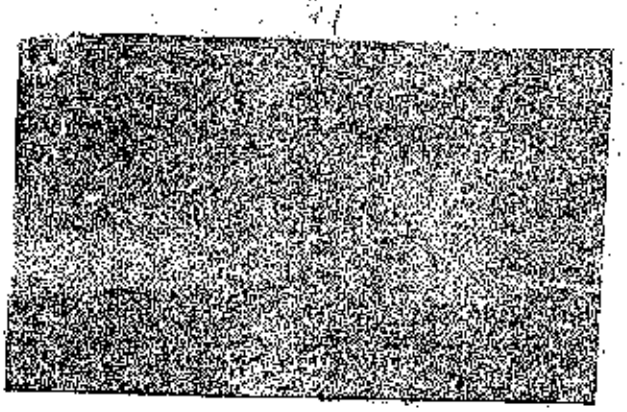

FIG. 1: *Shows calcification in the biceps and the triceps compartments, and a dense subcutaneous calcification at the extensor aspect of the elbow. This was before steroid treatment.*

The diagnosis of calcinosis universalis was made at this point and she was put on piroxicam, Augmentin (R) and prednisolone. Her fever and discharge subsided. A repeat x ray showed a marked reduction in muscle calcification and linear streaks of calcification (fig. 2). The oral steroid dose was then tapered to a maintenance dose and she remained well till recently when she developed pyomyositis of the calf muscles which responded to inci-

*B. N. Okeahialam
and
J. E. Ekedigwe

*Department of Medicine, and Department of Radiology, Jos Univeristy Teaching Hospital

*Correspondence

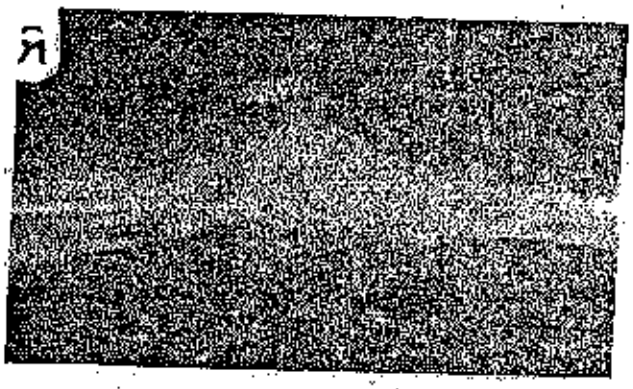

FIG. 2: *shows a reduction in the opacity of the biceps and triceps compartments. Linear streaks of calcification are now evident. Subcutaneous calcification around the elbow joint is still present. This was after initiation of steroid treatment.*

sion, drainage and systemic antibiotics. She is still being followed-up.

Comments

Dermatomyositis can occur at any age and is more in females. It could co-exist with neoplasm, vasculitis or other collagen vascular diseases. Some of them present with dysphagia and oesophageal dysfunction, myocardial involvement, vasculitis and arthritis. Calcinosis of the subcutaneous and soft tissues also occur but are rare (2). Infact calcinosis universalis is a late complication of the juvenile type. In it, calcium is diffusedly deposited in fascial planes of the skin and muscles (3). It is said to confer a good prognosis to the cases in which it occurs (4). However, some workers contend otherwise (5) believing that being a late complication, only those who live long enough for whatever reasons do develop it. Going by the fact that this patient on presentation had circumscribed subcutaneous calcinosis with ulcers, she actually started off with calcinosis cutis circumscripta. This is also found in dematomyositis (6).

We did not bother to confirm the diagnosis of dermatomyositis with muscle biopsy and enzyme studies as going through the referral notes and examining the patient left little if any room for doubt. Her ESR is suggestive. Her mild transient small joint arthritides does not exclude the diagnosis. She also had vascular headaches and unstable angina with normal ECG as well as dysphagia. These are all possible associations of the disease. She did not have chronic renal failure; Therefore her ectopic calcification could not have been due to it.

This case is presented to highlight this rare complication of an uncommon connective tissue disorder. It will expectedly improve the index of suspicion in clinicians who may encounter similar cases. This will enable proper diagnosis to be made and appropriate management instituted promptly.

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