Ind J Dermatol Venereol Leprol 1994; 60 : 225-7

ANGIOKERATOMA CIRCUMSCRIPTUM WITH MARFANOID HABITUS

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An 11-year-old male having zosteriform angiokeratoma circumscriptum is reported for its unusual association with Marfanoid habitus.

Key words : Angiokeratoma Circumscriptum; Marfanoid habitus

Introduction

Angiokeratoma circumscriptum (AC) is a rare congenital vascular malformation of the papillary dermis, evolves through stages. In early stage, it consists of aggregates of deep red coloured compressible papules or nodules that gradually become verrucous. The lesions may coalesce into one or several plaques with liner arrangement.[1] The majority of lesions occur on the lower leg and foot, occasionally seen elsewhere. It has been reported to occur with underlying naevus flammeus,[2] Cavernous haemangioma,[3] Klippel-Trenaunay sydrome,[4] Cobb syndrome,[5] and Cornu cutaneum.[6] Recently, we had a patient with AC who had associated Marfanoid habitus. This association seems fascinating for no such report is so far available in the literature.

Case Reports

An 11-year-old male presented with unilateral asymptomatic red coloured papules and plaques over right side of upper chest and upper limb since birth. The lesions were gradually increasing in size. Family members of the patient observed unusual increase in the height and aesthenic look in comparison to his brothers and sisters. No other family member had similar problem.

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Patient was tall, thin built with typical facies and high arched palate (Fig 1). His height was 149 cms (more than 95th percentile), weight 22 kgs (less than 10th percentile) and arm span equal to that of height. The ratio of upper segment (top of pubic symphysis to crown) to lower segment (top of symphysis pubic to sole of feet) of the body was 0.82. Arachnodactyly, positive wrist and thumb sign, prominent length of hallux,

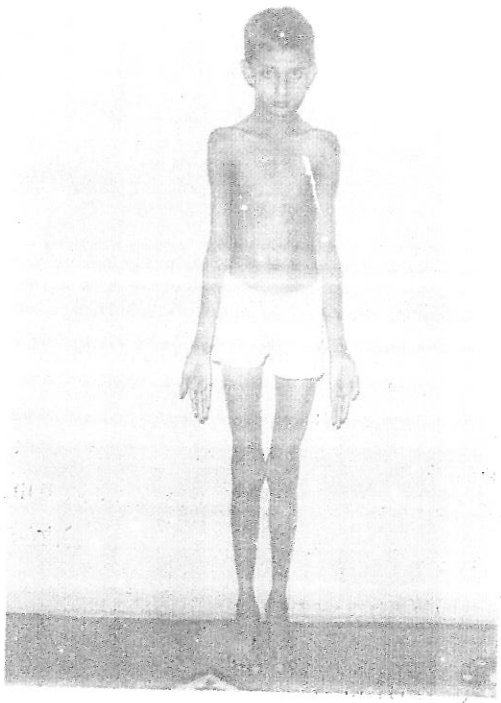

Fig. 1. Tall and thin built patient with abnormal body proportions.

pigeon chest, under-developed muscles and remarkable thinness of subcutaneous fat were other features seen in our patient. Besides, he had erythematous papular lesions, coalescing at places. Most of them had hyperkeratotic surface and bled on removal of keratotic scale. These lesions were arranged in a zosteriform pattern over right side of chest and upper limb (Fig 2). Various veins and increased girth of limb in comparison to other limb was the other conspicous feature present in the patient.

Haemoglobin, total and differential leucocyte counts, ESR, LFT, kidney function tests, electro-cardiogram and echocardio-

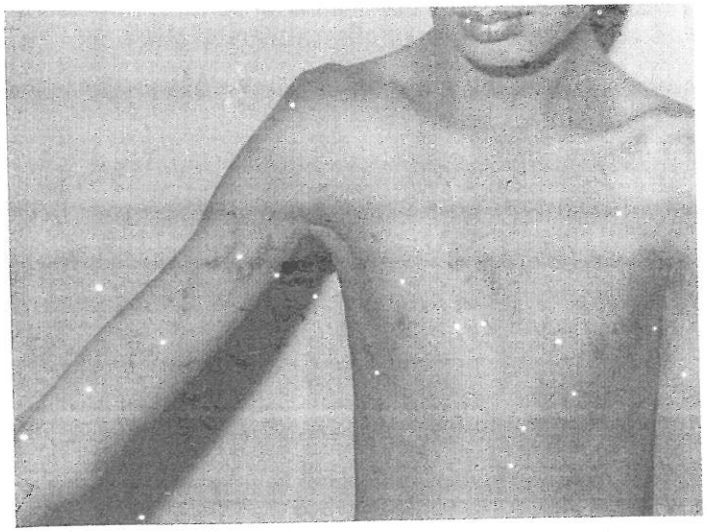

Fig. 2. Zosteriform pattern of angiokeratoma circumscriptum.

graphy were within normal limits. Sketetal survey confirmed the clinical findings of hands and feet, with metacarpal index of 10.05 (Normal individuals <8). Slit lamp examination of eyes revealed no abnormality. Microscopic examination of biopsy from right forearm lesion demonstrated ectatic dilated vessels in papillary dermis with marked acanthosis and hyperkeratotis of epidermis.

Comments

Marfan's syndrome is a rare, connective tissue disorder characterised by abnormally long extremities, arachnodactyly, ocular and cardiovascular defects, but partial forms with findings in only 1 or 2 systems are not uncommon.[7] It is an autosómal dominant disorder, however, about 5% of cases arise from new mutations. In our patient, there was no evidence of disease among any of his relatives.

The wide variability of clinical expression and delayed/later development of manifestations of this syndrome, in the absence of absolute diagnostic criteria may be responsible for dilemma in some cases. A high index of suspicion may be of prime importance in suspecting the diagnosis in infancy, but it may not be considered untill the second decade or later.[7] Skeletal defects are the most conspicous external feature of the disease. The patients are often tall and skeletal proportions are abnormal. Arachnodactyly is found in most patients with Marfan's syndrome and the calculation of the 'metacarpal index' may be helpful in its diagnosis. In addition, high arched and typical facies are other features[7] suggestive of Marfanoid habitus in our case.

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SCABIES

It all begins with the usual scratch & itch,
3-4 wks. after the arrival of the Sarcoptes witch.
To make matters even worse,
the entire family seems to acquire the curse.

Even though Avil & Calamine you may try.
The itch dose'nt subside- one wonders why?
Just when you begin to think mite is right.
The damn itch worsens at night!

It is then that the ever observant third eye;
In the web spaces, a burrow doth spy!
Add to that the many excoriated papules
and it leaves those in doubt looking like decorated fools.

For those doubting Thomas' who still harbour a doubt.
The microscope seems but the last resort.
Alas! this hapless creature lies flattened dorsoventrally;
With four pairs of short legs on an ovoid stocky body.

Sulfur, Rotenone & Malathione in its treatment have all been used down the ages;
So too we will find have
Monosufiram & Permethrin if we care to turn the pages.
Lindane & Benzyl Benzoate however are the current mainstay.
Antihistamines may be added to give a symptomatic allay.

In infants & children the scabicidals should be diluted;
For the head & neck, Crotamiton may be solicited.
The clothing & bedding must always receive a healthy wash and tan.
Not forgetting at the same time, that treatment of the entire family is also part of the plan.

To complete the picture, may I make an institutional mention;
Of a type of scabies commonly reffered to as crusted/Norwegian!
A florid form- this entity is no longer rare:
So my friends, when seen, of AIDS Beware!!

Dr Rodrigues
Banglore