

## CENTRAL ACRO-OSTEOLYSIS

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A case of bulbous swelling of the tips of fingers and toes, central-acro-osteolysis of terminal phalanges and bound-down skin of the extremities and face in a 14 year old girl, is reported.

**Key Words:** Sclerosis, Pseudoscleroderma, Pseudoclubbing, Central acro-osteolysis

### Introduction

Central acro-osteolysis of the terminal phalanges was typically described in occupational polyvinyl chloride (PVC) poisoning by Sucio et al in 1963.<sup>1</sup> Loss of distal phalangeal cortex, sparing the tufts like a 'half-moon', referred to as 'slice-effect' is seen. Central acro-osteolysis may also be seen in idiopathic familial acro-osteolysis of Hajdu & Cheyne, progressive systemic sclerosis (PSS), progeria, pachydermoperiostosis and hyperparathyroidism.<sup>1</sup>

### Case Report

A 14-year-old female student presented with progressive asymptomatic bulbous swelling of the tips of all fingers and toes of 4 years duration. There was no Raynaud's phenomenon and no symptoms suggestive of systemic involvement. There was no history of intake of drugs or contact with PVC and other chemicals. The personal, past and family histories were noncontributory.

On examination, she was found to have bulbous swelling and broadening of the tips of fingers and toes, involving the distal half of the terminal phalanges. The nails were broadened transversely, but there were no other nail changes or

paronchia (Fig. 1). The skin over the upper and lower limbs and also trunk was found to be sclerosed and bound down, more so proximally. The facial skin was also bound down with ironed out appearance and slight beaking of nose. There were no other skin lesions and Raynaud's phenomenon was not demonstrated even after prolonged dipping of the hands in ice cold water.



Fig. 1. Bulbous swelling of finger tips with transverse broadening of nails

Investigations on blood and urine were normal. X-rays of the hands and feet showed soft tissue swelling of the tips of fingers and toes and acro-osteolysis affecting the central part of the terminal phalanges sparing the tufts forming the 'half-moon cut', called 'slice-effect' (Fig. 2). X-rays of other bones were normal. H & E stained biopsy sections of the sclerosed skin showed mild hyperkeratosis, normal epidermis with some flattening of rete

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ridges and a subepidermal zone of fragmentation of collagen. The dermis appeared to be thinned out and appendages pulled up, but there was no inflammatory infiltrate and vessel walls were normal. Verhoeff Van Gieson staining showed absolute absence of elastic fibers in the subepidermal zone and loss of the wavy elastic nature to appear stretched out in reticular dermis.

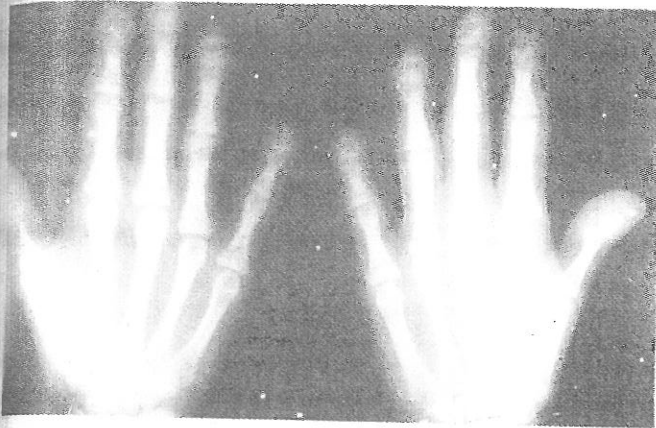


Fig. 2. X-ray hands showing central acro-osteolysis with "half-moon cut" called "slice-effect"

### Comments

The unusual features in this patient were pseudoclubbing and acro-osteolysis. The association of sclerosis of the skin of face and extremities was suggestive of scleroderma or toxic pseudosclerodermas. Though sclerosis of the skin, pseudoclubbing of fingers and central acro-osteolysis is typical of PVC poisoning,<sup>1-4</sup> the absolutely noninflammatory dermis, normal vessel walls and fragmentation of collagen in only the subepidermal zone, along with lack of history of contact with PVC or

other chemicals in any form, rules out PVC poisoning.

Though conical pointing and resorption of the 'acra' is more common in systemic sclerosis, thickening of soft tissues around terminal phalanges of fingers may be the earliest change.<sup>1</sup> Exceptionally, symmetrical acro-osteolysis involving central parts of the terminal phalanges producing 'half-moon' cut also may be seen.<sup>1</sup>

Broadening of the nails was a notable feature in this patient and it prompted the observation of similar changes in other cases of PSS, where also, acro-osteolysis in the corresponding terminal phalanges was seen though, the central type of acro-osteolysis was not seen. The unusual presentation and X-ray findings warranted the reporting of this case.

### Reference

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