

## CASE REPORTS

### CONGENITAL CONSTRICTIONS

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A young man having congenital annular constrictions of the legs with lymph-oedema of the left foot below the constriction, webbing of the fingers of both hands, anonychia of two fingers and intra-uterine amputation of terminal phalanges of two fingers and one big toe is reported.

**Key words :** Congenital constrictions, ainhum, pseudo-ainhum, congenital ring constriction, intra-uterine constriction bands.

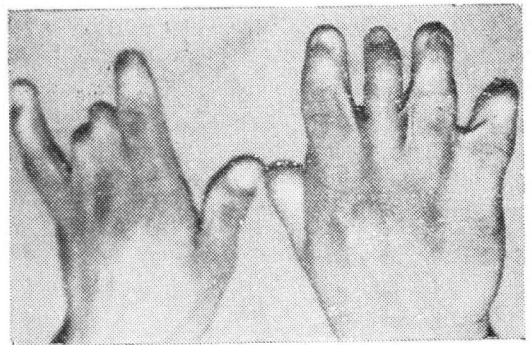
Congenital constriction of the limb is a rare anomaly. Its cause is still uncertain, though various postulations are put forward. Only five cases have been reported from India.<sup>1-3</sup> We are reporting a young male patient who had congenital constrictions of the legs associated with deformities of the fingers and toes.

#### Case Report

A 21-year-old male college student had multiple deformities of the hands and feet and annular constrictions of the legs since birth. For the last 10 years, he was gradually developing lymph-oedema of the left leg and foot. He was the first of two siblings and none in his family had similar deformity. There was no history of parental consanguinity or any illness or drug intake during pregnancy. He was born in a hospital after a full-term normal delivery. There was no history of birth injuries. The developmental mile-stones were normal. There was no other abnormality except the deformities of the hands and feet and constriction bands of legs.

There was a 2-cm wide constricting annular fibrous band on the left leg about 8 cm above the ankle producing a groove on the leg. The skin of the leg and the foot below the ring was

oedematous and the surface showed verrucous thickening. The terminal phalanx of the left big toe was missing since birth. The right leg had another fibrous band 13 cm above the ankle. This band was slightly oblique, incomplete and did not produce any groove on the surface. There was no lymph-oedema or any other deformity on that limb. Both the hands showed skin folds in between the fingers resulting in webbing of the fingers (Fig. 1). Peripheral arterial pulsations were normal except the left dorsalis pedis which could not be palpated due to the gross lymph-oedema. Cutaneous sensations were normal and there was no thickening of the peripheral nerves.



**Fig. 1.** Webbing of the fingers of both hands, anonychia of both index fingers, absence of terminal parts of a few fingers and crumbling of some nails.

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Routine investigations of blood, urine and stools were normal. Repeated examination of the blood smear did not show microfilaria. Blood VDRL was non-reactive. Serum uric acid was 5.4 mg%. Ear-lobe smears did not show any acid-fast bacillus. X-ray of the hands, feet, legs and chest showed soft tissue swelling of the left leg and foot and missing of the distal phalanges of the middle and ring fingers of the left hand and that of the left big toe. Biopsy of a part of the constriction band showed prominent sub-epithelial collagenisation.

#### Comments

Congenital constrictions are only rarely seen in dermatological practice. These cicatrizing bands develop usually around the limbs and are present at birth. The deformity can be prevented by early surgical removal of the band during childhood. The webbing of the fingers seen in our case is an uncommon deformity in association with the annular constrictions. Patterson<sup>4</sup> described four types of constrictions in this disease : (1) Simple constriction. (2) Ring constriction with deformity of the distal part (lymph-oedema may or may not be associated with it). (3) Ring constriction associated with fusion of the distal parts. (4) Intra-uterine amputations. In the present case, the right leg showed only a simple constriction while the left leg had a ring constriction with lymph-oedema of the foot, fusion of distal parts of the hands, absence of the distal phalanges of two fingers and left big toe as a result of intra-uterine amputations. So all the four changes described by Patterson were evident in our case. Anonychia seen on the fingers is an uncommon association.

The mechanism of development of these cicatrizing bands and deformities is not well understood. Turner<sup>5</sup> is of the view that these are caused by amniotic bands or adhesions. Seeds et al<sup>6</sup> also favoured this view and suggested that these deformities result from extra-amniotic, intra-chorionic entanglement with mesodermic strands. Another view is that these are the result of a primary constitutional defect of the germ plasm which results in defective growth of the tissue affected.<sup>7</sup> Association of congenital constrictions and amputations in the present case, favours the amniogenic aetiology. Stock and Stock<sup>8</sup> reported four cases and concluded that these lesions were not primary embryologic defects but a secondary super-imposed disease process of unknown aetiology.

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