

CONGENITAL BILATERAL RADIO-HUMERAL SYNOSTOSIS*

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SUMMARY

A rare case of congenital bilateral undifferentiated elbow with radiohumeral synostosis, with bilateral absence of ulnar ray components in the forearm and hand, with deformities of hands i.e. syndactyly in left hand fingers and hypoplasia of thumb with webbing of first space in the right hand, in a 3 years old female child, is being reported in this communication.

Congenital deformities in upper limbs are well known, ranging from partial deficiencies in longitudinal and transverse components to the total absence of the limb.

We are presenting here a rare case who had bilateral congenital deformities in the upper limbs and had associated deformities in the hands as well.

Case History

A three years old female child was brought to the out patient department of Orthopaedics, Nehru Hospital, B.R.D. Medical College, Gorakhpur with complaints of deformities in both the upper limbs since birth. The child had full term, normal, home delivery, without any antenatal supervision. History of intake of any teratogenic medicines or exposure to the radiation was denied by the mother.

On general and systemic examination following congenital anomalies were detected :

On local examination (Fig. 1) : The patient had normal height and weight for her age, but both the upper limbs were proportionately shorter. The shape of both the upper limbs were normal at the arm level. The elbow joint was absent. A small bud was present on the medial side of both the upper limbs at presumptive elbow level. At forearm, wrist joint and hand the components of the ulnar ray were absent on both the sides. Only thumb, index and middle finger rays in both the hands were present. There was syndactyly in the left hand in between the index and the middle

fingers. Right thumb was short, adducted, hypoplastic with webbing of the first space. The muscular contractability could be observed in flexor and extensor compartments of the forearm during finger and wrist movements.



Fig. 1. Photograph showing short upper limbs with syndactyly in left hand fingers and hypoplastic short right thumb. The shape of arm and forearm well demarcated.

X-ray findings of both the upper limbs were symmetrical (Fig. 2) : The shoulder joint was normal. Humerus was abnormally long in relation to other long bones in the body.

An osseous spur was present, medially projecting from the humerus at the junction of upper

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2/3rd and lower 1/3rd of the present single bone (Radiohumerus). At the distal end of the radiohumerus the wrist carpal bones were attached. The components of the ulnar ray in hand and forearm were absent and were represented only by the bony spur.



Fig. 2 Radiograph showing single radiohumeral bone with bony spur on both sides.

Discussion

Malformations of the upper limb have been variously classified (Frantz-O'Rahilly, 1962). However, Swansons (1976) classification is being followed by the International Federation of Societies for Surgery of Hand and other reputed Societies in the World. The main categories of this accepted classification are :

(i) Failure of formation of parts (arrest of development). (ii) Failure of differentiation of parts (Separation of parts). (iii) Duplication, (iv) Overgrowth (gigantism), (v) Undergrowth (Hypoplasia), (vi) Congenital constriction band syndrome, (vii) General skeletal abnormalities.

A congenital anomaly having bilateral absence of the elbow joint with radiohumeral synostosis along with congenital deformity of the hand is an extremely rare occurrence in the available literature.

So far only few cases of radiohumeral synostosis have been reported by Daniel, 1986. In the case reported by Daniel the bony spur was attached at the lower end of the radiohumerus and all the metacarpal and phalanges were present in the hand.

In extreme cases the ulna may be completely absent with or without radiohumeral synostosis in extension or flexion at elbow or else the ulna may be absent partially in its proximal or distal part in its milder form (Daniel, 1986). It is to be differentiated from distal phocomelia in which fully developed hand is attached directly to the arm, the forearm being absent.

In the present case there was unduly long single bone representing radiohumerus, symmetrically present in both the upper limbs. Symmetrical, bilateral osseous spurs were present on the medial aspects of radiohumerus at the junction of upper 2/3rd and lower 1/3rd i.e. the probable site of elbow. The osseous spur represented the under developed ulnar ray. The radial ray components were present in the forearm and hand. The upper radial end was fused in continuity with the lower end of the humerus (Non differentiated elbow). In the hand, little and ring finger rays were absent on both the sides.

Embryologically the limb bud is formed at the end of 4th week of intra uterine life, consisting of mesenchymal tissue covered by ectoderm. Mesodermal condensation forms the future bone, while interzonal mesenchymal tissue forms the joint. The self differentiation of bones and joint is based on genetically controlled, intrinsic factors (Gray, 1977 and Hamilton, 1976).

However, other workers are of the opinion that there is failure of segmentation of elbow joint during development whereby the distal humeral epiphyses and the proximal radial epiphyses are not formed and hence there is fusion of these two bones.

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