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Case Report

Ulnar dimelia-diagnosis and management of a rare congenital anomaly of the upper limb

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ABSTRACT

Ulnar dimelia is a rare congenital disorder, characterized by duplication of the ulna, absence of the radius and polydactyly. Authors present a case of a girl treated due to ulnar dimelia. Physical and radiological findings, surgical treatment and postoperative effects are described.

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1. Introduction

Ulnar dimelia is a rare developmental anomaly, which is characterized by absence of radius, duplication of ulna and symmetric polydactyly (seven or eight fingers and lack of thumb).

Authors present a case of a girl treated because of ulnar dimelia in our department.

2. Case presentation

A girl, was admitted to the Department at the age of two month by the reason of polydactyly. At admission, a left upper extremity, not shortened, with abducted hand and eight fingers was found. Four post-axial fingers were normal but four pre-axial digits presented several deformities. Physical examination revealed a normal range of movement in the brachial joint and a restricted range of movement in the ulnar

joint (flexion-extension 90–5°, pronation–supination 0°) as well as in the wrist joint (palmar flexion 80° dorsal flexion 20°, adduction and abduction 0°). The mobility of post-axial fingers was normal and of pre-axial fingers restricted. The examination of the right extremity didn't show any abnormality.

X-ray examination, performed in frontal and sagittal planes (Fig. 1.) revealed:

- a normal humerus with an ossification center in the post-axial part of distal epiphysis,
- an absence of radius,
- two ulnar bones: one (post-axial) presenting a proper morphology and the second (pre-axial) which showed a mild hypoplasia of proximal epiphysis,
- a wrist with two ossification centers correspondent to two capitates,
- a normal morphology of metacarpals and phalanges of four post-axial fingers,
- a hypoplasia of all pre-axial metacarpals,

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Fig. 1 – Radiogram of left hand – at admission: (A) – anterior-posterior, (B) – lateral.

- a shortening of middle and distal phalanges of pre-axial index (I) finger,
- normal pre-axial middle (M) and ring (R) fingers,
- a hypoplasia of pre-axial little (L) finger.

Right upper extremity showed no abnormalities in the X-ray examination.

The girl was qualified to the two-stage surgical treatment. At the first stage the pre-axial L and R fingers and metacarpals were amputated (Fig. 2).

During the second stage, after five month, amputation of pre-axial M finger and pollicization of pre-axial L finger were made.

Performed surgical procedures and systematic post-operative rehabilitation produced a good functional and cosmetic effect (Fig. 3).

At age of 10 years old child presented a wide range of movement in the ulnar joint (flexion-extension 110-5°, pronation-supination 15-0°) and in the wrist joint (palmar flexion 80° dorsal flexion 45°, adduction 5° and abduction 20°)

Fig. 2 – Radiogram of left hand – after first stage of surgical treatment: (A) – anterior-posterior, (B) – lateral.

Fig. 3 – Final result of treatment.

as well as a good hold function of the hand were achieved with thumb flexion MCP 0–40°, PIP 5–45° and abduction –adduction 0–50°.

3. Discussion

Ulnar dimelia is a very rare developmental anomaly of the upper limb. There are only approximately 70 cases described in the literature.^{1–9} Ulnar dimelia, called also the mirror-hand, is classified to the 3rd group of congenital hand malformation, according to the classification proposed by Swanson [Swanson 1976] and adopted by the American Society for Surgery of the Hand (ASSH) and the International Federation of Societies for Surgery of the Hand (IFSSH) (see Table 1).

There are two types of ulnar dimelia:¹⁰

- type I – characterized by duplication of ulna, presence of 5 proximal carpal bones (2 triquetral, 2 pisiform and 1 lunate) and 5 distal carpal bones (2 hamate, 2 capitate and 1 trapezoid) one index finger in the middle and three other fingers (M, R and L) on both side of index,

- type II – differs from the type I in presence of 2 lunate and 2 trapezoid bones in wrist as well as 2 index fingers in the hand.

Skeletal malformation may be followed by arterial and nervous anomalies as an absence of the radial artery, duplication of the ulnar artery, presence of abnormal arterial arches in the hand, shortening of radial nerve, duplication of ulnar nerve (sometimes with collaterals to median nerve).

Pathogenesis of ulnar dimelia is related to the upper limb embryology. The first step of limb formation is the rise of limb buds (Wolff crest) which appear on the ventrolateral side of the embryo. The buds are formed by a mesenchyme covered with a layer of ectoderm. The ectoderm layer – the apical ectodermal ridge (AER) – is responsible for induction and modulation of the limb growth. The mesoderm of the buds is divided into the zone of polarizing activity (ZPA) proximally and a progress zone (PZ) distal. The ZPA mesenchyme induced by AER differentiates into cartilaginous and muscular tissue.^{11,12} The disturbance of differentiation of ZPA along the axis of the limb may lead to post-axial duplication and ulnar dimelia. A few reports concerning genetic mechanisms of ulnar-dimelia pathology suggest HOX genes mutations: 7p15.3 (HOXA@), 17q21.3 (HOXB@), 12q13.3 (HOXC@), and 2q31 (HOXD@) or translocation breakpoint at 14q13.^{13,14} The treatment of ulnar dimelia should lead to the best functional and cosmetic effect. Anatomical correction is possible in these cases in which there are no severe deformations of the hand. The polycization of the index finger (in type I of dimelia) or middle finger (in the II type) and amputation of other pre-axial fingers and metacarpals should be done. In those cases in which anatomical correction is impossible the re-creation of hold function of the hand is the target of treatment.

Table 1 – Classification of congenital hand deformities.

Type	Description
Type I	Failure of formation
Type II	Failure of differentiation
Type III	Duplication
Type IV	Overgrowth
Type V	Undergrowth
Type VI	Constriction band syndromes
Type VII	Generalized anomalies and syndromes

4. Conclusion

In the presented case of type II ulnar dimelia an anatomical correction of the hand was possible. The result of surgical treatment in both functional and cosmetic aspects was, in authors' opinion, good.

Conflicts of interest

All authors have none to declare.

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