Radiology Case Reports 17 (2022) 4885–4888

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Ulnar dimelia and a rare variant ulnar trimelia: Case reports ☆

Jesus Alberto Mosquera-Lopez, MD[a,c,*], *Oscar Alberto Mosquera-Lopez, MD*[a], *Claudia Palomino-Diaz, MD*[b], *Jesus Alberto Mosquera-Perea, MD*[a]

[a] *Department of Diagnostic Imaging, DIAGNOSTICAR S.A.S., Choco, Colombia*
[b] *Department of Orthopedics and Traumatology, DIAGNOSTICAR S.A.S., Choco, Colombia*
[c] *Unidad de Diagnóstico Por Imagen DIAGNOSTICAR S.A.S, Calle 24 # 7-07, Quibdo, Choco, Colombia*

ARTICLE INFO

Article history:
Received 15 September 2022
Accepted 18 September 2022

Keywords:
Ulnar dimelia
Mirror hand
Congenital anomaly
Polydactyly
Ulnar trimelia

ABSTRACT

Ulnar dimelia is an infrequent congenital anomaly of the upper limb characterized by the duplication of the ulna, absence of the radial ray, and polydactyly. We report on 2 cases of young girls with upper limb abnormalities who were brought to our Advanced Imaging Department for specialized imaging evaluation. The first case is a 15-month-old girl with a typical manifestation of ulnar dimelia. X-ray images of her right forearm and hand showed 2 ulnar-like bones, absence of the radial ray, and post-axial polydactyly. Our second case is a 7-year-old girl with evident limited pronosupination and shortening of the left forearm. Findings from computed tomography evaluation included ulnar duplication, one of which had a bifid aspect, with characteristics that are similar to ulnar proximal and distal epiphysis. The latter case which, to the best of our knowledge, has not been reported in the literature, is a rare variant of ulnar dimelia, that we have called *ulnar trimelia*. In this contribution, we emphasize the importance of an adequate diagnostic imaging approach for accurate recognition of upper limb anomalies, so that the treatment selection is appropriate to achieve better health outcomes and improve patients' quality of life. Our descriptions and images presented in this contribution form the basis for further research.

Introduction

Ulnar dimelia, also referred to as "mirror hand," is one of the rarest congenital disorders of the upper limb characterized by duplication of the ulna, absence of the pre-axial structures (radius, radial carpal bones, and thumb) and polydactyly [1]. The first case of ulnar dimelia was documented in 1852 and the specimen is currently preserved at Harvard Medical School [2]. There are no cases reported in our country, Colombia. There have been about 70 cases of ulnar dimelia reported in the literature and many of them only describe the case characteristics [1], but not advanced diagnostic and therapeutic approaches to recommend a clear management path.

☆ Competing Interests: None.

* Corresponding author.
E-mail address: jesus.ml@diagnosticar-ips.com (J.A. Mosquera-Lopez).

https://doi.org/10.1016/j.radcr.2022.09.067

Ulnar dimelia affects men and women equally [3], mostly with an unilateral involvement of the left upper limb. An autosomal dominant component is conferred to this condition in some syndromes such as Laurin-Sandrow, in which there is an association between ulnar duplication and the presence of fibula duplication, absence of the radial ray and tibia, polysyndactyly, and facial anomalies [4].

The exact pathogenesis mechanism is not clearly kwon; it has been described that the zone of polarizing activity, which is responsible for the development of the anterior and posterior axes of the undifferentiated mesenchyme that will eventually differentiate into the upper limb, is altered throughout its development process; thus, leading to a post-axial duplication and ulnar dimelia. Mutations of the genes HOX and TWIST1, ALX4, GLI-3, HOXB8, HAND2 that generate some subtypes of polydactyly and dimelia of the upper limb have been also described [1].

In general, the cosmetic and functional involvement of limb defines the management of this condition [3,5,6]. Because there is still a limited amount of literature regarding ulnar dimelia, it is important to describe cases found all around the world to improve the diagnostic and therapeutic approach. In this article, we present a typical ulnar dimelia case and a special case with particular characteristics that have not been described in the literature. Moreover, we discuss therapeutic alternatives that can be followed.

Case report no. 1

This case is about a 15-month-old girl, first child, born via spontaneous vaginal delivery at 7-month gestational age. After birth, during a physical exam, 7 fingers were found in her right hand. Patient was brought to our institution for advanced imaging evaluation. Physical examination showed that head, neck, lower extremities, and left upper limb had no apparent alteration. However, the exam revealed absence of the thumb in her upper right limb, duplication of the last 3 fingers of the hands, and 2 ulnar bones. Pinch grip was not possible. A 3D reconstruction of the right upper limb and conventional X-ray of the hands and distal forearm presented in Fig. 1 show 2 bones with ulna-like characteristics; no trochlear joint in the elbow; absence of the radial ray, thumb, metacarpal and carpal bones, as well as duplication of triphalangeal fingers. Coronal T2-weighted sequence magnetic resonance shows duplication of ulnar vascular structures (Fig. 2A); and simple tomography of the shoulder, shows hypoplasia of the glenoid cavity (Fig. 2B).

Case report no. 2

A 7-year-old girl, born on time via spontaneous vaginal delivery after a normal pregnancy, was sent to our center for an imaging evaluation after resection of supernumerary fingers. Physical exam evidenced shortening of the left forearm, limited pronosupination, polydactyly, pollicization of the left hand, and adequate pinch grip strength. Computerized tomography of the left forearm and hand (Fig. 3) shows ulnar duplication; one of the ulnas in normal position and configuration, but the lateral ulna with a bifid appearance, which shares characteristics with an ulnar proximal and distal epiphysis. All of the fingers in the left hand have 3 phalanges, but no thumb was visualized. There was duplication of the carpal bones. Also, shortening of the forearm bones and postsurgical changes in the thenar region due to pollicization were observed.

Discussion

According to Al-Qattan MM et al., our first patient could be classified as a type 1A ulnar dimelia with a classic presentation. For our second reported patient, imaging examination showed ulnar dimelia, but because of the proximal thickening and the distal duplication of the lateral ulnar, is difficult to fit her case within one of the approved categories of ulnar dimelia. Thus, we consider this case a rare variation of ulnar dimelia and called it *ulnar trimelia*. To the best of our knowledge, this is the first report of this variant.

Although it is not the case of our patients, this condition could involve the complete anatomy of the upper limb; in many cases, it can involve the more proximal structures such as the scapula, humerus, clavicle and glenohumeral joint. In some other cases, there is scapular hypoplasia, recurrent glenohumeral dislocations, hypoplasia of the capitellum in the forearm articulating with 2 bones of ulnar morphology described in corpses as rotated between 70° and 180° from each other. These changes generate biomechanical alterations in the elbow joint, which result in limitation of the normal range of motion [7].

Due to the infrequent occurrence of ulnar dimelia, case reports do not provide an ideal or generalized therapeutic approach for treating this condition. Therefore, it has not been possible to establish a management protocol. Currently, individualized management according to outcome goals is the optimal approach.

The therapeutic approach to treat this condition could be surgical and nonsurgical, and the selection of the course of treatment is based on the patient's measured joint mobility and its ability to perform activities of daily living, as well as the aesthetic component that could negatively affect patient's self-esteem and mental health. In this case, the reconstructive process might be important. The treatment is focused on the correction of polydactyly and reconstruction of the thumb to achieve a hand with a similar look to a normal one, thus allowing the patient to perform daily activities. To this end, several techniques can be employed including pollicization of the more radial and more functional finger, amputation of the fingers and pre-axial metacarpals, tendon transfers to improve the position and mobility of the wrist, resection of the more lateral proximal ulnar end to allow a functional pronosupination. Finally, procedures such as arthrodesis or resection of the first row of the carpus cann be done to provide greater stability [8].

Taking into account the concept "the hand is an extension of the brain" [9], brain plasticity is crucial for the rehabilitation of these patients. This capacity produces a cortical reorgani-

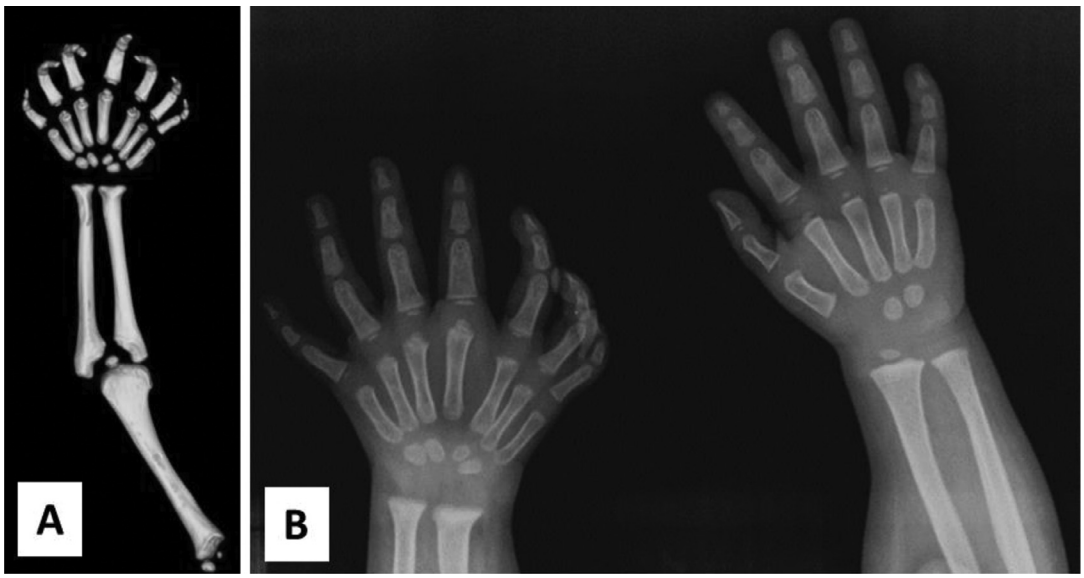

Fig. 1 – (A) CT of the right upper limb. Duplication of the ulna, single ulnar epiphyseal growth nucleus, absence of the radius, duplication of carpal, and metacarpal bones. (B) Comparative X-ray, right mirror hand, left hand normal.

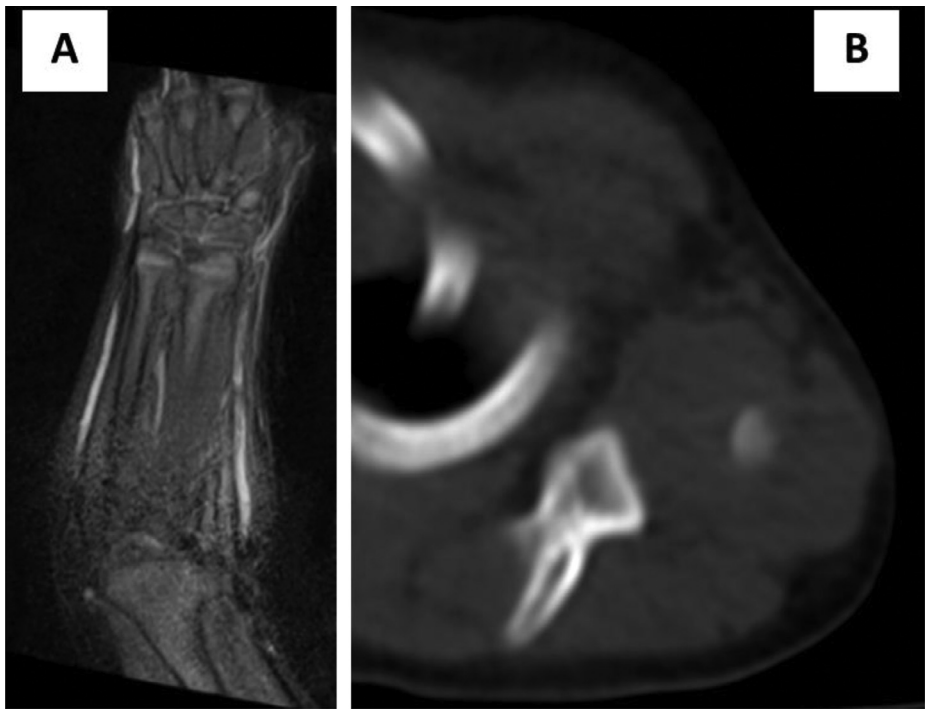

Fig. 2 – (A) MRI of the right forearm. Duplication of the vascular structures of ulnar morphology. (B) Axial CT, glenoid cavity hypoplasia.

zation in response to changes in the way each finger is used in a repetitive way or to changes in sensitivity. In patients with congenital alteration in the mobility of the thumb, when performing the flexion reconstruction, new synaptic pathways are generated, allowing the area of the cortex previously dedicated to flexion of the elbow to adapt to control de flexion of the thumb. Although the significance of age in brain plasticity is not yet clear, it is known that younger patients have greater potential for plasticity and reorganization of the motor cortex than adults [10].

In conclusion, we have presented 2 cases of ulnar dimelia/trimelia, a rare congenital malformation of the upper limb with significant functional and cosmetic compromise for the patient. In this contribution, we reported a special case, which has the characteristic of a duplication of one of the mirror ulnas, and therefore we propose a new category

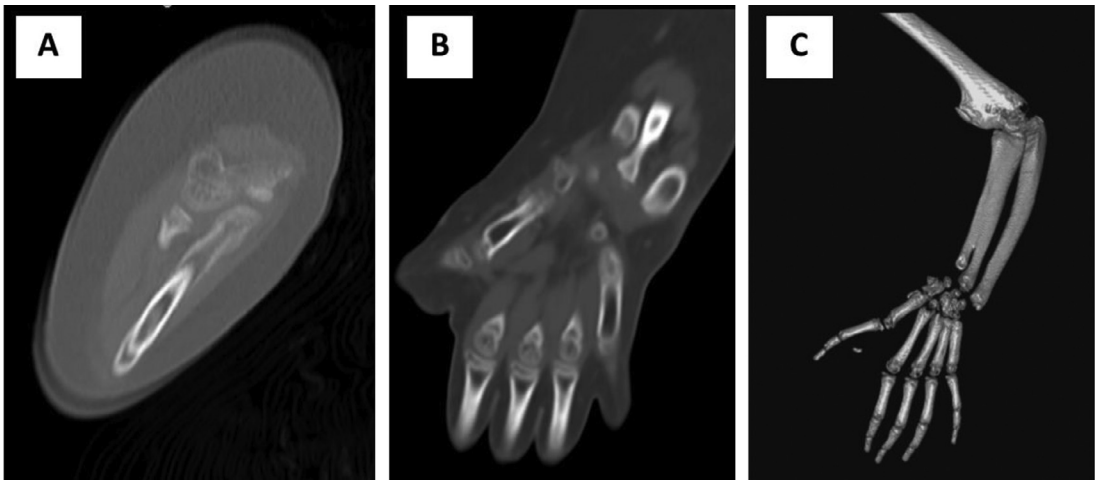

Fig. 3 – (A) Sagittal CT of the elbow, ulnar duplication with different height without articulating with each other. (B) Coronal CT, distal forearm, medial ulna normal appearing, lateral ulna bifid appearing, shaping distal ulnar TRIMELIA. (C) 3D reconstruction of the arm and forearm, humerus-ulnar joint, without configuring the trochlea. In the hand, the fingers have 3 phalanges and no thumb.

called *ulnar trimelia*. Our findings form the basis for a generalized imaging-based diagnosis and potential therapeutics options for patients. Also, these reports open doors for further research on additional ulnar dimelia variants.

Patient consent

Written informed consent to perform the imaging test and authorization for personal data collection, usage, and analysis was obtained from the patients' guardians during the admission procedures. The authorization form complies with Colombian laws and regulations on privacy and data protection (Habeas Data).

REFERENCES

[1] Tomaszewski R, Bulandra A. Ulnar dimelia-diagnosis and management of a rare congenital anomaly of the upper limb. J Orthop 2015;12:S121–4. doi:10.1016/j.jor.2015.01.027.
[2] Afshar A. Ulnar dimelia without duplicated arterial anatomy. J Bone Joint Surg Br 2010;92-B(2):293–6. doi:10.1302/0301-620X.92B2.23057.
[3] Chinegwundoh JOM, Gupta M, Scott WA. Ulnar dimelia: is it a true duplication of the ulna? J Hand Surg 1997;22(1):77–9. doi:10.1016/S0266-7681(97)80024-1.
[4] El Hage S, Ghanem I, Megarbané A, Razzouk C, Dagher F, Kharrat K. Main en miroir : une nouvelle forme avec revue de la littérature. Rev Chir Orthop Réparatr Appar Mot 2008;94(2):174–8. doi:10.1016/j.rco.2007.09.003.
[5] Aucourt J, Budzik J-F, Manouvrier-Hanu S, Mézel A, Cotten A, Boutry N. Congenital malformations of the hand and forearm in children: what radiologists should know. Semin Musculoskelet Radiol 2012;16(2):146–58. doi:10.1055/s-0032-1311766.
[6] Gropper PT. Ulnar dimelia. J Hand Surg 1983;8(4):487–91. doi:10.1016/S0363-5023(83)80213-5.
[7] Al-Qattan MM, Al-Kahtani AR, Al-Sharif EM, Al-Otaibi NJ. Thumb reconstruction without formal pollicization in mirror hand deformity: a series of four cases. J Hand Surg Eur 2013;38(9):940–7. doi:10.1177/1753193412475129.
[8] Takagi T, Seki A, Takayama S. Elbow and forearm reconstruction in patients with ulnar dimelia can improve activities of daily living. J Shoulder Elbow Surg 2014;23(3):e68–72. doi:10.1016/j.jse.2013.12.001.
[9] Lundborg G. Brain plasticity and hand surgery: an overview. J Hand Surg 2000;25(3):242–52. doi:10.1054/jhsb.1999.0339.
[10] Bunketorp Käll L, Cooper RJ, Wangdell J, Fridén J, Björnsdotter M. Adaptive motor cortex plasticity following grip reconstruction in individuals with tetraplegia. Restor Neurol Neurosci 2018;36(1):73–82. doi:10.3233/RNN-170775.