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CASE REPORT PEER REVIEWED | OPEN ACCESS

Special case of bilateral postaxial longitudinal deficiency in a 3-year-old child

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ABSTRACT

Introduction: Considering the variety of their clinical, radiological, and etiological manifestations, congenital abnormalities of the upper limbs represent complex diseases. They can be separated into brachydactyly, duplication, differentiation failure, and training failure. Ulnar longitudinal deficiency, a deformity that belongs to the first group and is uncommon, manifests differently in the forearm and the hand. Postaxial longitudinal deficiency and unilateral radial humerus synostosis are common diagnoses.

Case Report: We present a case of a 3-year-old boy who had radio-humeral synostosis on the right side along with a typical congenital failure of longitudinal bilateral ulnar development.

Conclusion: While traditional radiography still plays a significant role in diagnosis and prognosis, ultrasonography and magnetic resonance imaging (MRI) will soon be valuable diagnostic tools. Each case is different, and the treatments and the time of surgery are specific, the optimal treatment remains the prosthetic door; however, the placement of a prosthetic on a sensitive member requires early intervention of the patient and his family.

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INTRODUCTION

There are only 6.5 to 21.5 occurrences of congenital upper limb abnormalities per 10,000 live births [1]. Based on radiographic findings, they are typically classified into four types: training failure, differentiation failure, duplication, and brachydactyly [1, 2].

The upper limbs' development is halted in the first type of training failure. Transverse, intercalary, and longitudinal are its divisions (preaxial, postaxial, and mesoaxial) [1].

In this case study, we describe a young child who had a typical bilateral postaxial longitudinal training failure deformity.

CASE REPORT

We report the case of a 3-year-old child, only son, with no significant antecedents other than a consanguinity second degree, who exhibited a congenital arrangement of the two forearms with digital residual (Figure 1A–C) and a language delay on clinical examination. Biological analysis and karyotype are normal.

Upper limbs X-ray exam showing bilateral abnormalities, with partial ulnar aplasia with absent carpal bones and all digits other than the index fingers on the right side, also exhibiting radio humeral synostosis and radial inclination (Figure 2A and B).

The rest of radiology, cerebral computed tomography (CT), abdominal ultrasound (US), Doppler echocardiography, standard X-rays: chest, lower limbs, pelvis, and spine are normal.

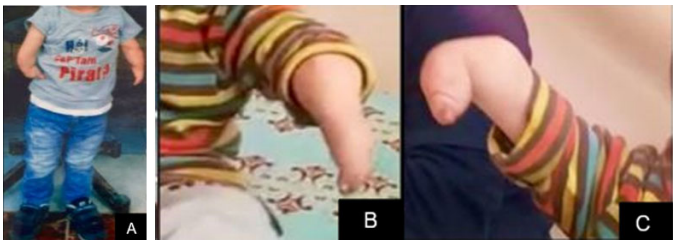

Figure 1 A–C: Photographs showing the clinical appearance of the longitudinal cubital deficiency.

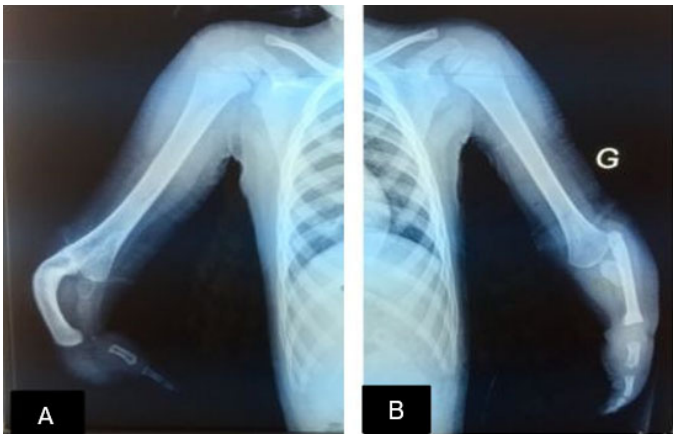

Figure 2A and B: Upper limbs X-ray exam showing bilateral abnormalities, with partial ulnar aplasia with absent carpal bones and all digits other than the index fingers on the right side in A, also exhibiting radio humeral synostosis and radial inclination.

DISCUSSION

Postaxial longitudinal impairment, also known as partial or whole longitudinal ulnar impairment, has been described as a rare upper limb ailment in a number of papers [1, 3]. GOLLER provided the first account in 1698 [4]. Radial dysfunction is ten times more common [4, 5]. The majority of ulnar impairment patient sets were more rare compared to radial dysfunction [5]. While many individuals have bilateral deformity, which is the situation with our patient, a study revealed that it is typically unilateral [1–3]. The majority of ulnar impairments are intermittent. Only in rare instances where they are a part of a syndrome (Cornelia de Lange syndrome, Schinzel-Giedion syndrome), dominant autosomal inheritance has been recorded in a study, which is also compatible with our situation [3].

Congenital malformations of the upper limbs usually occur between the third and eighth weeks of embryogenesis [1]. Golberg and Bartoshesky divided the causes of birth defects into three categories [3]: 1: environment (agent affecting the fetus): irradiation, certain infections (varicella, rubella, toxoplasmosis), hormones, alcohol, and especially drugs, all these factors are absent in our case; 2: genetic: (chromosomal and Mendelian model), in our case the karyotype was normal; 3: uncertain.

Postaxial structures such as the ulna, cubital carp bone (triquetrum, pisiform, and hamate), or the ring and small fingers, may be affected by longitudinal damage [1]. In the instance of our patient, all of these abnormalities are bilateral and include partial ulna aplasia, the lack of carpal bones, and absence of all fingers other than the index finger. However, postaxial longitudinal impairment can also be accompanied by femoral and fibular impairment, craniofacial malformations, right radio-humeral synostosis, radial tilt, which is also present in our case, and abnormalities of the thumbs (first insufficient web space, aplasia, hypoplasia, and losing opposition), which is confirmed in our case where there is bilateral aplasia of the thumb [1].

So the ulnar longitudinal impairment presents itself with varying degrees of severity, both in the forearm and in the hand [4]. The percentage of involvement of the radial side of the hand in this ulnar longitudinal gap is important [3].

The classification proposed by BAYN in 1986 lists 4 degrees of increasing severity [4]:

1. hypoplasia of the ulna;
2. partial aplasia of the ulna with radius curvature and possible radial head dislocation;
3. aplasia of the ulna with radius curvature and radial head dislocation;
4. aplasia of the ulna with fusion of the radius and the humerus.

Obstetric ultrasound is currently used more frequently to diagnose congenital anomalies (especially when there is a considerable impairment), but for the majority of them, conventional radiography is still the imaging method of choice, mostly to identify bone abnormalities [1]. However, information on cartilage, muscles, tendons, arteries, and nerves are not available via standard radiography. In the near future, pre-operative evaluation of congenital abnormalities of the upper limbs using MRI and ultrasound is anticipated to become highly helpful [1].

According to the cases and classes of ulnar deficiency, therapeutic abstinence, radio-humeral osteotomy, elbow-type prosthesis, and single-bone forearm are the relative means and therapeutic indications [4, 5].

A study was carried out on 29 patients with 34 ulnar limbs defiant, to establish a classification system useful for predicting prognosis and planning treatment [5]:

Type A: Dislocated radial head with inclined radius. These patients do not need osteotomy to position the hand in front of the body. A bony forearm construction is not required since the development of the radial bow.

Type B: Dislocated radial head, straight radius, ulnar cloth with bent elbow. The function is improved by an early adjustment with an above elbow-type prosthesis. Placing a prosthesis on a sensitive member requires early intervention by the patient and his family.

Type C: Radio-humeral synostosis with bowed radius, variable presence of ulna.

Patients in this group, with more severe combinations of deformities, will benefit from radial or humeral osteotomy to position the hand in front of the body.

Type D: Radio-humeral synostosis with straight radius and diminutive ulna. In this type, patients are easily identified at birth and should require no treatment for their elbow and forearm deformities.

CONCLUSION

Despite this, molecular diagnostics has advanced. To correctly detect congenital abnormalities of the upper limb, clinical and radiographic expertise is required. Standard X-rays are essential for diagnosis and functional prognosis, but in the near future, ultrasound and MRI will be fantastic instruments for more accurate evaluation of these disorders.

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Author Contributions

Chaimae Abourak – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.