A 5-year-old with abnormal hand and forearm: diagnosis and discussion

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Diagnosis

Ulnar club hand/ ulnar dysplasia [type I variant].

Discussion

Ulnar club hand, also known as ulnar dysplasia or postaxial longitudinal deficiency, is a rare condition with an incidence reported by some authors to be as low as 1:1,000,000 [1]. The abnormality occurs as a result of distal tethering of the ulnar fibrocartilaginous structure to the carpus, with resulting bowing of the radius. Most cases are sporadic and causes include postinfective [2]. Association with rare inherited disorders such as Klippel Feil and ulnar mammary syndrome as well as nongenetic syndromes such as Cornelia de Lange has also been reported [1,3]. Associated major organ system malformations are uncommon though musculoskeletal malformations, such as

phocomelia, aphalangia and radial deficiency, are seen in up to 50% of cases [3].

Patients usually present with a foreshortened, internally rotated forearm with associated hand malformations. Dislocation of the radius head (present in our patient) with decreased range of movement of the elbow is reported in 40% of cases [3].

The condition was classified by Bayne according to musculoskeletal abnormalities of the elbow and forearm into four groups [3]. Type I presents with hypoplasia of the ulna, both proximal and distal ulnar physes present and nonprogressive radial bowing. Type II consists of a partially absent radius, a fibrous anlage extending to the carpus from the distal ulna, radial bowing and ulnar deviation of the hand. Type III involves absence of the ulna with no anlage present, unstable elbow due to absent olecranon and radius, wrist and hand are usually straight. Type IV presents with synostosis of the distal humerus and proximal radius with marked radial bowing and an ulnar anlage extending from the humerus to the carpus. Our patient was classified as type I due to the presence of a completely bowed radius, visible proximal ulnar cartilage structures and trochlea as well as the probable fibrous anlage distally.

Further classification on the basis of presenting hand malformations was established by Cole and Manske and they divided the dysplasia into four types based on the first webspace and the thumb. Type A presents with a normal thumb and webspace, type B with contracture of the webspace, type C with hypoplasia and rotation of the webspace into motion plane of the other digits and type D presents with an absent thumb [3]. These classifications are mostly used by clinicians to establish a treatment protocol for patients, which are mostly designed to improve function and address cosmetic concerns. Our patient presented with

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a normal thumb and first webspace and therefore was classified type A.

Radiographs are helpful in establishing the diagnosis whereas MRI is useful in further classifying the dysplasia depending on the presence of a fibrous anlage, presence of the distal and proximal ulnar ossification centres and in confirming dislocation of the radius when this is not obvious on radiographs.

References

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