

Research Journal of Pharmaceutical, Biological and Chemical Sciences

A New Variant of Forearm Dysplasia in Children, in Addition to Those Described by Masada et al.

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ABSTRACT

A new variant of Masada, with distal radial and distal ulna exostosis with forearm deformity is reported.

Keywords: forearm dysplasia, children, deformities, exostosis.

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INTRODUCTION

Forearm deformities in the children and adolescents do occur sporadically. Some of them have associated exostosis. Such deformities are due to dysplasia occurring in the parent bone and problems of remodeling [1]. In India such patients present themselves late due to poor literacy [2]. Masada et al's classification is widely used to classify such forearm deformities [3]. We present a case of a fore arm exostosis with dysplasia of the forearm bones not reported in the Masada classification.

Case report

A 12 year old girl was brought by her parents for having developed multiple swelling over her upper and lower limbs. She had swelling of the left forearm with bowing and restricted rotations of the forearm. She also had swelling of her left leg. Figure 1 shows relative bowing of the left forearm and Figure 2 shows the restricted pronation of the left forearm of the girl. Figure 3 shows the leg of the patient. Figure 4 shows the X-ray of the left forearm of the girl.

With the presenting features a diagnosis of exostosis of the forearm- Distal radial and ulnar exostosis with DRUJ disruption was made. The girl's parents were not willing for any surgery and no intervention was made.

DISCUSSION

When the literature was searched for the classification of this particular presentation, it was surprising to find that such a type was not included in Masada's classification [1]. In 1989 Masada et al described four types of forearms with deformity. These are Type I, Type IIa, Type IIb and Type III. Of these type I had a distal ulna exostosis with Distal Radio Ulnar Joint (DRUJ) disrupted; Type IIa had distal ulna and proximal radius exostosis and the Proximal Radio Ulnar Joint (PRUJ) and DRUJ disrupted; Type IIb had distal ulna exostosis and both the PRUJ and DRUJ are disrupted. Type III had distal radius exostosis with both PRUJ and DRUJ intact.

Logically one expects that in such a classification of a new entity, the researcher should have diagnosed all the cases, treat them and then classify them. Naturally not all the cases could have come to them at one point of time. Thus this classification might be only a retrospect. Since there is no mention of the retrospective nature of the classification, the possible method is that the first case with forearm deformity seen by them is Type I, the second case is Type IIa, the third case can be Type III and the fourth type of case might have been Type IIb. Of the total 36 cases only 16 were operated. Among the operated cases only one is type 3. This means type 3 Masada carries less favourable outcome. The paper [1] does not mention in the table the age of the individual patients. This is important as the progress of this disease is age dependent. In general causes of all the deformities were not explained by any worker. Naturally one would just think that an exostosis mechanically pushes the bones apart in that part. However in type 2b of Masada et al, they were not able to explain why the proximal radial head got dislocated with a distal ulna exostosis. Thus the cause of deformity cannot be simplified as mechanical.

There had been quite a few terms used to describe Multiple Osteochondroma(MO), after John Hunter described it in 1786, e.g. diaphyseal aclasia, diaphyseal aclasis, multiple osseous exostoses etc. To reduce this confusion, WHO in 2002 had suggested the term Multiple Osteochondromatosis to be used if there were a minimum of two chondromata of the metaphyseal regions of long bones. Majority (75%) of the MO individuals have an osseous deformity and these osseous deformities mostly involve the forearm (50%), followed by the ankle (45%). 40 % of MO patients have a short stature. There is no specific prediction to any sex. When extensively affected, range of movement of forearm may be limited [4].

The MO are described even in bones excavated from archeological sites .In a study on 16 such samples are described with specific reference to MO . This gives an insight about how the MO will progress if not treated. Of these,one specific case (Sk 331) from Ballyhanna, CO, Donegal, Ireland had MO involving distal radius and distal ulna. But in that paper this specimen was classified as type 1 Masada [4]. Actually it should have been the new variant proposed in the present paper. Figure 3 in the reference of Murphy et al shows such variants are seen even in medieval era [4].



Figure 1 showing available supination of the left forearm compared to the right forearm



Figure 2 : showing available pronation of the left forearm



Figure 3 showing the swellings of the left leg



Figure 4. Radiograph of the forearm showing the exostoses of distal radius and ulna with DRUJ disruption, with arrows marking the exostosis both in distal radius and distal ulna.

Thus with all previous reports of MO have been based on the type of Masada et al, this paper presents a variant and was not even operated. How this case report is going to make any difference in the manner by which the MO is viewed now? In their original paper Masada et al advised excision of the exostosis and ulnar lengthening. But they themselves, in a later paper of a longer follow up of their cases, have reported [5] that corrective osteotomy and lengthening of forearm bones are futile in these cases. This is concurred by another author[1] who found that ulnar lengthening did not affect the clinical and radiological outcome of these cases. Thus when it comes to deformities with forearm exostosis, any operative procedure is controversial.

Thus the new variant proposed in this paper which might have even been present from the medieval times can be included for proper classification and communication. As deliberated earlier with enough confusion in the nomenclature, I only want to add one thing

that is so far not reported – exostosis in both distal ulna and distal radius. In our case even though both the bones had distal exostosis – since the diameter of distal radius is significantly more than the ulna - relative overgrowth of the radius over the ulna causing the DRUJ disruption [6,7].

CONCLUSION

In the presented case the clear difference from the existing classification is the presence of distal ulna and distal radial exostosis with Distal Radio Ulnar Joint (DRUJ) disruption. This variety is not reported in Masada's paper [3].

REFERENCES

- [1] Shin EK, Jones NF, Lawrence JF. J Bone Joint Surg Am 2006 88(2) 255-60
- [2] Illiterate patients: greater care and caution required in the Indian context. Med law Cases Doct, vol 5 issue page 180
- [3] Masada K, Tsuyuguchi T, Kawai H . J Bone Joint Surg 1989;71 –B ; 24.
- [4] Murphy E M, Mc Kenzie CJ, J Archeolog Sci 2010;37:2255-64
- [5] Akita S, Murase T, Yonenobu K, Shimada K, Masada K, Yoshikawa H. J Bone Joint Surg Am. 2007;89(9):1993-9.
- [6] Bilen FE, Eralp L, Balci H I, Kocaoglu M, Ozger H . Acta Orthopeda Belgica 2009 75 p743-7
- [7] Hilton P. Gottschalk Yumiko Kanauchi Michael S. Bednar Terry R. Light. J Hand Surg 2012;37(11):2286-2293.