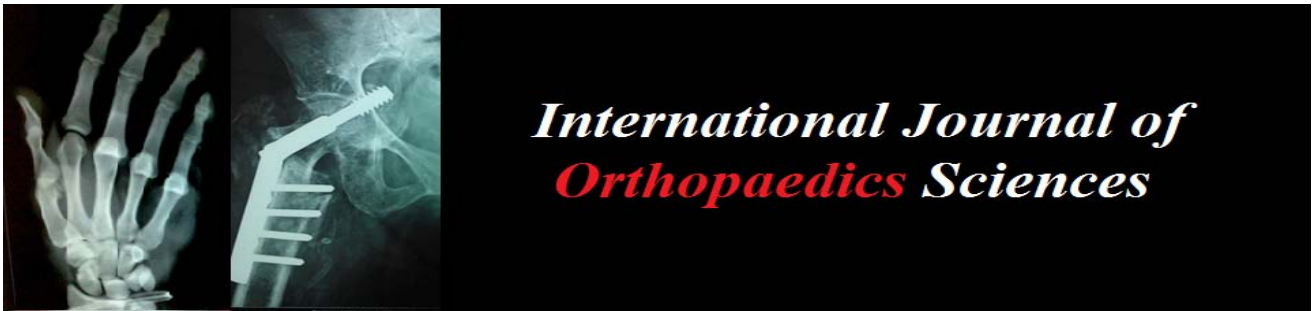

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Novel correction and fixation technique of forearm deformity in cases of hereditary multiple exostoses

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Abstract
We present three cases of forearm deformities which were corrected and fixed with a novel method using intra-osseous wire loops. Two cases of Masada Type I and one case of Masada type IIa forearm deformity were treated with an excision of the distal ulnar osteochondroma and corrective osteotomy of the radius at two sites with intramedullary nail, which was secured with two intra-osseous stainless steel wire loops. At 5 years follow-up of the first case, pronation improved from 35° pre-operatively to 70° post-operatively. At 3 years follow-up in the second case, pronation improved from 20° to 40°, whereas no significant improvement was noted in the third case at 2 years follow-up. The Radial Articular Angle and Carpal slip improved significantly for all the three cases. Thus, a good functional outcome can be obtained with this easy technique with minimal and cheap inventory.

Keywords: Hereditary multiple exostosis, Forearm, deformity, osteochondroma, osteotomy

1. Introduction

Hereditary Multiple Exostoses (HME) is a disorder of enchondral bone growth affecting the metaphyseal region of the growing bone. Multiple forearm deformities occur in upto 30%-60% of these patients [1]. The indications for forearm deformity correction include pain, functional deficit and concerns of cosmesis.[2] Multiple surgical techniques are available for the correction of deformity. We describe here an innovative correction and fixation technique involving excision of the distal ulnar osteochondroma followed by corrective osteotomy of radius at two sites with an intramedullary nail, which was secured with two intra-osseous stainless steel wire loops on the tension side to close the osteotomy site. With this minimal and affordable inventory, we had a very good functional outcome.

Case 1

A 16 year old male presented with gross deformity of forearm and pain while carrying weight. He had multiple hard bony swellings over multiple sites namely, proximal humerus, proximal and distal regions of femur and tibia. A clinical diagnosis of hereditary multiple exostoses was made. There was a radially convex deformity of the forearm with an ulnar deviation of the wrist. The ulnar border of the forearm was foreshortened. There was a bony swelling present along the distal ulna (Figure 1a). The range of motion of flexion and extension at the elbow was normal. The pronation was restricted pre-operatively to 35°. The
radiographs confirmed the presence of Masada Type I deformity of the forearm with distal ulnar osteochondroma, bowing of the radius, ulnarly directed carpus, located radial head and a foreshortened ulna (Figure1b).The radial articular angle was 49.12° and carpal slip was 61.6%. Considering the patient was skeletally mature, a definitive corrective procedure was planned. At 5 year follow-up, the patient did not have a recurrence of deformity with an excellent range of motion (Figure 2, 3a). At the end of a 5 year follow-up, the pronation improved from 35° pre-operatively to 70° post-operatively. The radiographs at the end of 5 years, showed union and remodelling at the osteotomy site (Figure 3b). The radial articular angle decreased to 21.67° and the carpal slip was 48% (i.e. a centralised carpus). The patient had good grip strength, though we did not objectively measure the same in this case with a Jamar Dynamometer.

Case 2

A 13 year old female with hereditary multiple exostosis having distal ulnar osteochondromas and shortening (Masada type 1). There was convex radial bowing of the radius shaft and wrist was ulnarly deviated. (Figure 4) Pronation was restricted to 20° on the left side and 30° on the right. There was full supination on both side. Elbow flexion and extension range was normal. Left side was operated as it was more symptomatic. The pre-operative radial articular angle was 55.6° and the carpal slip was 83.1%. Correction was done by a similar technique using intramedullary nailing and 'figure of 8' intraosseous SS wire loop on the tension side at the osteotomy site (Figure 5). At latest 3 years follow-up pronation on the left side improved to 40° and supination remained full. Post-operatively the radial articular angle improved to 24.1° and the carpal slip was 41%.

Case 3

A 12 year old female presented with a gross deformity of the left forearm (Fig. 6a) with multiple bony swellings noted over her scapulae and distal femur. A clinical diagnosis of hereditary multiple exostoses was made. She had similar deformities as the first case but her range of motion at the elbow was restricted from 10° to 110°. Her supination and pronation was restricted to 60° each. The radiographs confirmed the presence of a Masada type IIa (Fig.6b) but the radial head was deformed and subluxed (not dislocated). Since the range of motion at the elbow was functional, proximal radius osteochondroma was not
excised. The radius shaft deformity was corrected by the similar technique as described above after the excision of the distal ulnar osteochondromatous mass. Radius intramedullary nail was removed at 1.5 years post-surgery. Post-operatively at latest 2 years follow-up, no significant improvement was noted in the pronation and supination but the Radial Articular Angle decreased from 49.43° to 22.26° while the Carpal Slip improved from 93.88% to 54.42% after the corrective surgery (Fig. 7a, b).

2. Surgical technique

First, the distal ulnar osteochondromatous mass (Figure 8a) is excised along with the band tethering the carpus ulnarward, through an ulnar approach. Then, through a volar approach, single stage corrective osteotomy of the radius is performed at one or multiple levels (at the Centres of Rotational Axes). An intramedullary nail is inserted into the radius to ensure rail-roading of the segments while closing the osteotomy radially. The osteotomy is closed by manually deviating the wrist and forearm radially. The osteotomy is secured with two intra-osseous stainless steel (SS) wire loops in a figure of '8' configuration (Figure 8b). Post-operatively, the patient is immobilized in an above elbow slab for 8 weeks before range of motion is begun for the elbow and wrist.

Fig 1: Pre-operative Forearm deformity (a) Clinical image showing ulnar shortening, bowed radius, distal ulnar bony swelling (b) Antero-posterior and Lateral Radiographs confirming Masada Type I deformity of the forearm

Fig 2: Clinical image of the forearm deformity at 4 weeks post-op showing a good cosmetic outcome as compared to the opposite normal forearm

Fig 3: 5 year post-operative follow-up (a) clinical image showing good correction of the deformity without a recurrence of deformity with a normal grip (b) Radiographs showing union and remodelling at the osteotomy sites, restoration of Radial Articular Angle and a centralised carpus

Fig 4: Preoperative a) clinical image showing cosmetic deformity in forearm with ulnar deviation of wrist (b) Preoperative anteroposterior and lateral radiograph showing distal ulnar osteochondroma (Masada Type 1)

Fig 5: 1 year Post-operative anteroposterior and lateral radiograph showing healing of the osteotomy site and restoration of radial articular angle

Fig 6: Pre-operative Forearm deformity (a) Clinical image showing ulnar shortening, bowed radius, distal ulnar bony swelling (b) Antero-posterior and Lateral Radiographs confirming Masada Type IIa deformity of the forearm with distal ulnar and proximal radius osteochondroma but with only a subluxation of the radial head.

Fig 7: At 2 years follow-up: (a) Clinical images showing correction of the deformity of the radius shaft (b) Radiographs showing union and remodelling at the osteotomy sites, restoration of Radial Articular Angle.

Fig 8: Intra-operative Images (a) showing the distal ulnar osteochondromatous mass (b) Extraperiosteally resected osteochondroma and corrective osteotomy of the radius secured with two intra-osseous stainless steel wire loops

3. Discussion

Forearm deformities are very common in Hereditary Multiple Exostoses (HME) due to ulnar shortening and differential lengthening of paired bones. Ulnar shortening occurs due to the distal ulnar physis being affected to a greater extent because of its smaller crosssectional area than that of the distal radius. Moreover, the distal ulnar physis contributes more to total length than the distal radial physis does to the total length of the radius [2]. Many surgical procedures have been described for treating the forearm deformity like simple excision of osteochondroma, acute or gradual ulnar lengthening, corrective radial osteotomy, hemi-epiphyseal stapling of the distal radius, creation of a one-bone forearm and Sauvé-Kapandji procedure [2]. Excision of the osteochondromatous mass is done in both mature and immature skeleton. But the recurrence rate is lower in case of patients nearing skeletal maturity [3] In most cases, an ulnar lengthening procedure was performed, using either a monolateral or ring fixator [4, 5] It has been stated in literature that ulnar lengthening is a difficult procedure associated with complications [6, 7]. It is also reported that only symptomatic radial head instability is an indication for an ulnar lengthening procedure [8]. Considering that the radial head was stable in our case, we did not perform an ulnar lengthening. A corrective osteotomy of radius is recommended at near skeletal maturity [8]. In most cases the radius osteotomy was fixed with a plate. In the past, an intramedullary guide wire has been used to guide forearm lengthening with an external fixator [6]. We used an intramedullary nail in the radius to guide the segments of radius while compressing the osteotomy site. The nail thus enabled the segments of radius to rail-road while closing the osteotomy. However, we have used intra-osseous stainless steel wire loops to secure the osteotomy rather than a plate. There is no English literature on the use of an intramedullary nail and SS wire loops in a radial corrective osteotomy in a mature skeleton. This fixation technique is cheap, easy to perform and requires very little inventory. This technique is only meant to highlight the technique of radius deformity correction. The status of the radial head or proximal radius anatomy will determine the functional outcome in terms range of motion at the elbow joint. We hereby suggest that in patients of HME with a forearm deformity who are skeletally mature or who are nearing skeletal maturity, a favourable clinical outcome can be attained with this cost-effective technique of osteochondroma excision with a corrective osteotomy of the radius with easily available implants, provided the patient selection is appropriate.

5. Conflict of Interest: None

6. No financial support was received for this study

6.1 Ethical standards

All patients gave an informed valid consent prior to being included into the study. This study was performed in accordance with the Ethical standards of the 1964 Declaration of Helsinki as revised in 2000.

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