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A case of congenital pseudoarthrosis of forearm

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Abstract
Congenital pseudoarthrosis in pediatric bones is a rare condition. It is most commonly seen in Tibia and fibula. We encountered similar pathology in forearm. It was treated along the lines of pseudoarthrosis of tibia. We achieved stable union, with continued growth of forearm and had good functional and cosmetic results.

Keywords: pseudoarthrosis, pediatric forearm fracture nonunion, neurofibromatosis

Introduction
Fracture Union is a complex process. Disturbance of this complex process can lead to severe problems in union. Pseudoarthrosis of bone is a congenital condition where some abnormal tissue surrounds the bone and disturbs its remodeling potential [1, 8]. This tissue also promotes osteoclastic activity in the bone so that this bone becomes weak and prone to fracture [5]. Once the bone is sufficiently weakened due to the presence of this hamartomas tissue, a fracture happens in this bone. Then the fracture doesn't unite normally [5, 8]. This leads to development of pseudoarthrosis. Pseudoarthrosis is most commonly seen in lower limb at middle third lower third tibia junction. Incidence of this pathology, pseudoarthrosis in forearm is exceedingly rare [2]. We present a case of a 6 year old child , whom we found out to be having similar pathology in the forearm and applied the principles of treating pseudoarthrosis in tibia .We had an excellent long lasting result

Case report
A four-year-old child presented to us with history of fall at home in a trivial trauma and in the process broke his forearm. He was seen by an orthopedic surgeon who advised X-rays of forearm. This x-ray showed an undisplaced radius fracture at middle third lower third level. There was abnormality and lysis of distal end of ulna. Since it was an undisplaced fracture, splint support was given. At removal of plaster at 1 and a half months the forearm deformity had increased. There was persistent pain and swelling in forearm. Child was seen by another doctor who advised closed reduction and cast. Child underwent this procedure under anaesthesia and an approximate reduction was achieved. This cast was continued for another 4 months. On removal of cast, the deformity had worsened and X-ray showed sclerotic bone ends and increased bone gap, suggestive of no progression of union [4]. At this stage, movements of wrist and pronation and supination of forearm were painful.
At this stage, child was seen by us. There was no family history of similar condition. However we noticed Café-au-lait spots on child's groin area and fathers chin area. We suspected neurofibromatosis inheritance in this child. However since the child was from an unfavourable economic background, we could not get genetic testing done. We suspected that the forearm fracture nonunion was a skeletal manifestation of neurofibromatosis. It was decided to get an MRI Done. It confirmed abnormal hamartomous tissue around the radius fracture and the distal end of ulna was also absorbed by this hamartomas issue.
We offered surgery to this child. The surgical principles of treatment of congenital pseudarthrosis of Tibia where followed [10]. During surgery we explored the site of pseudo arthrosis. Abnormal white tissue was found at the fracture site and the surrounding periosteum was thickened and fibrotic [8]. Abnormal nerve branches from superficial radial nerve were seen to be entering the fracture site. We debrided the hamartomatous tissue and circumfrentially excised the periosteum till normal periosteum was found. Bone ends were found to be cartilaginous of fibrocartilage type and were excised till healthy bleeding bone was seen. The bone ends were opposed and a single K Wire from the dorsum of radius was passed from distal end.

The K Wire was passed through the physis in a single attempt. External fixator device was used to provide continuous compression at the fracture site.

The histopathology findings showed membranous tissue composed of fibrillar cells with ovoid nuclei; the chromatin was granular and sparse and showed no nucleoli. There was some differentiation into nerve like structures, articular cartilage

The fracture united within one and half months. We removed external fixator device at the end of one month and the intramedullary nail k-wire was retained. The child was started on rehabilitation and gradually gained full function within four months. Child was called for regular six monthly follow up to monitor the quality of union.

At last follow up, child has no pain, instability and a normal functional range of movement of affected limb. The growth of radius has not been affected by the transphyseal k wire. The forearm range of movement is -- normal supination but, there is 50° pronation lag. Wrist flexion extension is normal. Elbow flexion extension is normal. Wrist is slightly radially deviated on the forearm.

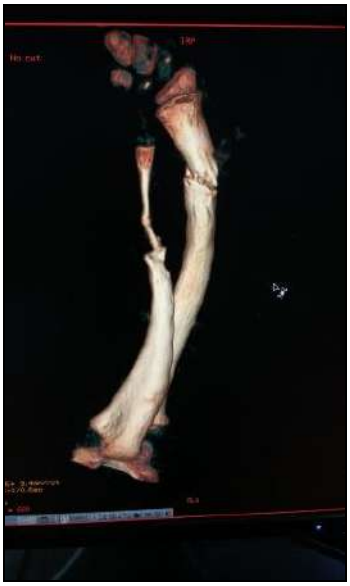

Fig 1: Preoperative X-Ray

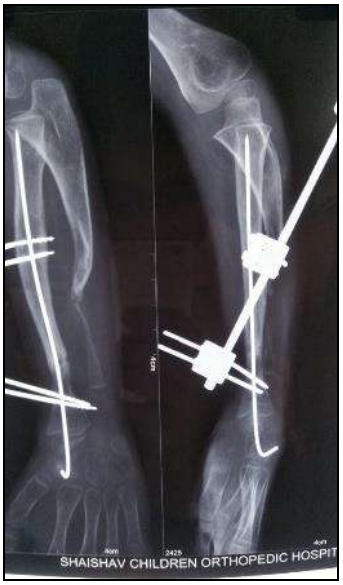

Fig 2: Postoperative X-Ray at union

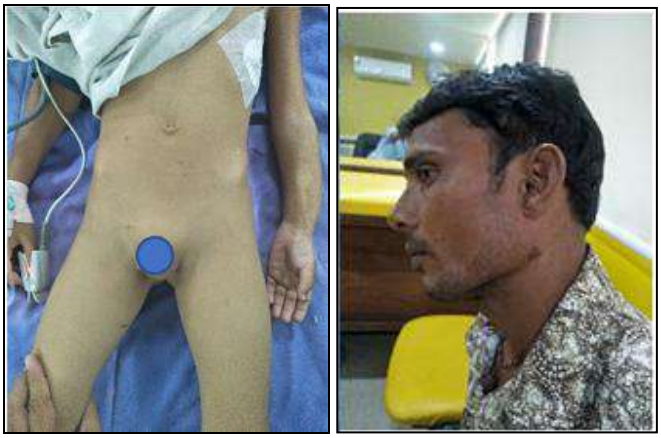

Fig 2: Freckles indicative of neurofibromatosis in child's groin area and fathers chin area

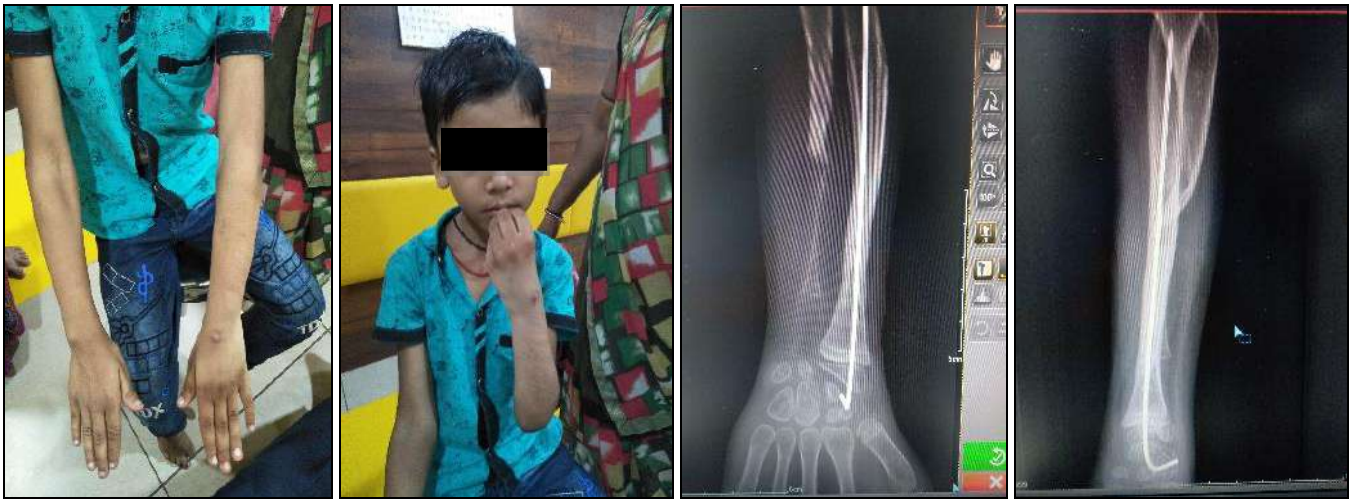

Fig 3: Final Followup Xray and Clinical Result

Discussion

Neurofibromatosis is a rare genetic, autosomal dominant condition [6]. There are three types, of which type 1 is known to cause skeletal manifestation. NF1 gene, located on chromosome 19 is responsible for this type. Along with other manifestations like auditory problems, vision problems, freckling or macrocephaly, it can have orthopedic manifestations also [6]. The most common orthopedic manifestation is Pseudoarthrosis of tibia [5, 10]. Occurrence of similar pathology in forearm is a rare occurence and as such treatment principles are not well defined [2, 7]. Our case had no progressive union of radius with complete absence of distal ulna. We decided to attempt union of radius using the same principles of pseudoarthrosis in tibia. The single bone forearm thus created will provide good cosmetic and functional result.

During the surgery, we found abnormal nervous tissue around the metaplastic perisoteum which needed to be debrided. This sort of proves the pathology as being, orthopedic manifestation of a neural tissue tumor[8]. Restoration of alignment of radius was critical as it balances the biomechanical forces across the bone and prevents refracture through the pathological bone. We retained the intramedullary nail till last follow up, so as to provide additional internal support to the bone [10].

A review of literature, reveals no high level of evidence [7]. There are multiple case reports and some case series', reflecting the rare occurence. The presentation varies….it can be...gradual deformation of bones of forearm, to a fracture that refuses to heal. No standard surgical procedure has been described[7]. All the cases show involvement of both bones. Some authors have described good results with plating only [4. There are reports of treating this nonunion with cancellous bone grafting or a vascularised fibula grafting [7, 11]. There are many reports of creating a single bone forearm [10]. In our case we did not create a single bone forearm, because we found the supination and pronation to be quite functional. All case reports and series indicate a good functional result after surgery.

Conclusions

If a fracture is not uniting or a fracture happens after a seemingly trivial trauma, we should look for an alternative diagnosis [9]. Thorough debridement of abnormal periosteum in a pseudoarthrosis case gives excellent results [10].

Conflicts of Interest Statement

The author declare that there is no conflict of interests regarding the publication of this paper

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