Current problem cases

Pseudoarthrosis of the ulna in neurofibromatosis

A report of four cases

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Summary. Four children suffering from neurofibromatosis with ulnar pseudarthrosis and progressive reabsorption of the middle and distal thirds of the ulna are reported. None had any pain or sensory loss, but all had progressive deformity of the forearm. Instability of the elbow and wrist were present in three cases, while one case showed good stability and function. A cross-union of the ulna with the radius to produce a one-bone forearm was accomplished using screw fixation and iliac bone grafting, and a one-bone forearm was achieved in three cases. Non-operative management was decided on in the patient with slow ulna reabsorption. The creation of a one-bone forearm is more likely to produce sound union, thus avoiding the need for further operations, while conservative management should be reserved for patients with a slowly progressive condition.

Neurofibromatosis is frequently complicated by pseudarthrosis of the tibia [1, 6, 8]. Reports of isolated ulnar pseudarthrosis in neurofibromatosis are rare, with a computer search identifying only 12 documented cases (Table 1).

We report on the management of four patients suffering from neurofibromatosis who developed ulnar pseudarthrosis.

Case reports

Case 1

At the age of 3 years and 6 months, a girl whose mother was affected by neurofibromatosis was referred with progressive deformity of the left forearm over the previous 15 months. At 23 months she had fallen on the outstretched left forearm. At that time, this had been regarded as of minor importance and no treatment sought. There were several cafe-au-lait spots, but no further manifestations of neurofibromatosis, namely a left-sided amblyopia with astigmatism.

X-ray revealed partial reabsorption of the distal end of the ulna, with a pseudarthrotic lesion at the border between the upper and middle thirds. Progressive narrowing of the ulna and its medullary canal, with complete reabsorption of the lower middle third, was evident. The proximal end of the ulna was not affected. The radius showed moderate bowing (Fig. 1a). The girl did not complain of pain, and sensation was intact in the left forearm, wrist and hand. There were no other manifestations of neurofibromatosis, X-ray of the spine, skull and pelvis revealing normal results.

It was decided to establish cross-union of the ulna with the radius to produce a one-bone forearm. The proximal fragment of the ulna and the proximal end of the radius just distal to the radial head were exposed through a posterior approach, with haemostasis achieved by means of a tourniquet. The periostium was elevated from the adjacent areas of the radius and ulna, after which the roughened cortical surfaces were opposed and secured in mid-rotation using two AO screws. The whole area was then decorticated, and cancellous bone graft from the iliac crest packed into the gap between the two bones (Fig. 1b). An above-elbow plaster of-Paris U slab was applied and changed for a full above-elbow plaster-of-Paris cast after 2 weeks. This was kept in place for 3 months, by which time solid union had been achieved (Fig. 1c) despite an accidental fall on the left hand 1 month after the operation. One year later, the forearm had excellent function, with little residual deformity and evidence of sound union. The distal end of the ulna showed evidence of progressive resorption. During the 12 months following the operation, she developed a further sign of neurofibromatosis, namely a left-sided amblyopia with astigmatism.

Case 2

A 9-year-old girl presented with extensive erosion of the distal end of the ulna (Fig. 2a). She had familial neurofibromatosis from her father's side, and 15 cafe-au-lait spots were evident but no other manifestations of the condition. Weakness of the hand, wrist instability, dislocation of the radial head and forearm shortening were present, with marked limitation of elbow and wrist movements. Radio-ulnar synostosis was performed through a posterior approach, with haemostasis achieved by means of a tourniquet. The middle third of the radius was approached by lifting the muscles from the interosseous membrane, and an osteotomy was carried out at the level of the distal end of the proximal fragment of the pseudarthrotic ulna. The distal fragment of the radius was opposed to the roughened lateral surface of the ulna and secured in mid-rotation using two screws [9]. Cancellous bone graft from the iliac crest was packed between the two bones (Fig. 2b). The patient was kept in an above-elbow plaster-of-Paris cast for 3 months to achieve solid union. Eighteen months later, the forearm was stable, and...
Fig. 1a-c. Case 1. a On presentation, radius is bowed, ulna shows tapering of its distal end, with evidence of reabsorption. b Postoperative X-ray. The proximal ends of the radius and ulna have been united using two AO screws and cancellous bone grafts taken from the iliac crest. c Twelve months after the operation there is radiographic evidence of sound healing.

Fig. 2a-c. Case 2. a Extensive erosion of the distal end of the ulna. b Post-operative view. The radio-ulnar synostosis is soundly united. c Eighteen months after the operation. A one-bone forearm has been created.

Fig. 3a, b. Case 3. a Extensive reabsorption of the middle third of the ulna. b Sixteen months after the operation, the osteotomy is fully united and stable union has taken place.

the hand function was good with evidence of sound union. The screws were removed, and the patient has since been able to lead a normal life (Fig. 2c).

Case 3

A 6-year-old boy in whom a diagnosis of neurofibromatosis had been made on clinical and histological grounds presented with extensive reabsorption of the middle third of the ulna (Fig. 3a). Clinical instability of the wrist and elbow was present, but the child was not complaining of pain or tenderness in the pseudarthrotic area. Forearm rotation was full, while some limitation of flexion was evident. Radio-ulnar synostosis using the same technique as described for the previous case was carried out [9]. When last seen, the pa-