Introduction

Congenital pseudarthrosis of the radius is an extremely rare condition. It is usually associated with an autosomal dominant disorder, neurofibromatosis type 1 (von Recklinghausen’s disease). Mutation of the NF1 gene on chromosome 17 causes von Recklinghausen’s disease. The NF1 gene is a tumour suppressor gene; it encodes a protein, neurofibromin, which modulates signal transduction through the ras GTPase pathway. In all cases reported, pseudarthrosis of the radius occurred in the distal third. Numerous treatment options have been explored with varying degrees of success and the reports have demonstrated successful healing.

Case report

A 9-year-old female child was brought to our centre by her father with a complaint of gradually progressive apex anterior deformity of the left forearm for the last 1 year (Fig. 1). She had a history of fractures of both bones of the left forearm after trivial trauma at the age of 7 years (2003) and 8 years (2004). On each occasion she was treated conservatively in a POP cast for 4 weeks. She reported to us in January 2005 complaining of a gradually increasing deformity of the left forearm. Radiologically, it was an apex anterior deformity of the distal aspect of the left radius. Clinically she had multiple café au lait spots over her body. Neurological and ophthalmological examinations were normal. The fibrous tissue and the fracture ends were excised. The fracture was stabilized with a 6-hole DCP with iliac crest graft to bridge the gap along with cortico-cancellous chips. The fracture united uneventfully at 3 months post-operatively. At 17 months post-operatively there is no evidence of recurrence of pseudarthrosis with a near normal range of movements. Congenital pseudarthrosis of the radius is an extremely rare condition with only 10 cases seems to have been reported. Dual onlay bone graft, vascularised fibular graft has been the treatment options the latter being the preferred one. But the disease being extremely rare not much has been documented about the treatment options. We treated this case by excision of the sclerotic bone ends along with a cuff of periosteum and internal fixation with DCP along with iliac strut graft to restore the length. Successful union was achieved in 3 months and the patient has satisfactory follow-up at 17 months.

Key words

Congenital pseudarthrosis of radius • Deformity • Café au lait spots • Non-union • Strut grafting • Open reduction • Internal fixation
In January 2004 she sustained another injury of the left forearm due to a fall and had a fracture of both radius and ulna (Fig. 4). After being treated in a POP cast for 4 weeks she developed a deformity which had progressively ultimately culminated in an apex anterior deformity of the left radius when the patient reported to us on 8 January 2005 (Figs. 1b–d).

On detailed clinical examination we found she had multiple café au lait spots over her body (Fig. 5a) one of them could be seen over the anterior aspect of left forearm (Fig. 5b, c). There was tenderness and mobility at the fracture site with shortening of the left forearm and restriction of pronation (Fig. 5d). There was no family history of neurofibromatosis. Radiological examination revealed apex anterior deformity of the left radius in distal third with pseudarthrosis. X-rays of the spine and CT scan of the brain were normal. The blood picture was Hb 81%, ESR 25 mm/h, neutrophils 50%, lymphocytes 47%, eosinophils 1%, monocytes 2% and basophils 0%.

We referred the patient to an ophthalmologist for further investigation, and the ophthalmologic examination was normal.

On 12 January 2005 under general anaesthesia surgery was performed using the anterior approach. Fibrous tissue between the fracture ends was seen. The tips of the fracture ends were sclerosed and tapered with thickened periosteum. The fracture ends were excised till bleeding normal bone was seen. Strut graft from the ipsilateral iliac crest was used to bridge the gap and stabilised using a six-hole DCP and screws. The fracture ends were decorticated and cancellous chip bone-grafting was performed. Post-operatively an above-elbow POP cast was put on, with elbow in 70° flexion. The stitches were removed on the 12th post-operative day. After 4 weeks
the cast was removed and active exercise of the wrist and elbow joint was started. Histopathology showed fibrous tissue.

The patient was followed at 3-month intervals. Seven months after surgery the fracture had united and the graft had been incorporated (Fig. 6). One year post-operatively the fracture had completely united (Fig. 7a), with restoration of supination and pronation of the left forearm (Fig. 7b, c).

**Discussion**

Congenital pseudarthrosis of the forearm is a rare condition. About 60 cases have been reported in the literature [1]. The ulna is more commonly involved than the radius. Only 10 cases of congenital pseudarthrosis of the radius seem to have been documented [2]. Boyd and Sage [3] suggested dual onlay bone grafting as the treatment. Kameyama and Ogawa [4] reported good results after complete resection of the involved radius with the surrounding periosteum and free vascularised fibular graft. More
recently vascularised fibular grafting has become the preferred treatment [1, 5–7]. Other procedures like external electrical stimulation of the forearm and reversal of the bone segment to place the bone adjacent to the pseudarthrosis in contact with the normal bone have failed to produce union [5].

Cleveland et al [8] treated 4 cases of congenital pseudarthrosis of radius with corticocancellous bone grafting and achieved union in 3 cases.

In conclusion, we report a case of congenital pseudarthrosis of the radius treated by excision of the sclerotic bone ends, internal fixation with DCP and iliac strut graft to restore length of the radius. Successful union was obtained in 3 months and the clinical follow-up continues to be satisfactory at the latest follow-up at 18 months.

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