NEUROFIBROMATOSIS AND PSEUDARTHROSIS OF THE ULNA

A Case Report

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In 1793 Tilesius published his beautifully illustrated account of molluscum fibrosum as it appeared in a fifty-year-old man. Ninety years later von Recklinghausen recognised the relationship of these swellings to the peripheral nerves, and in 1918 Gould referred to additional features of the condition, including the skin pigmentation, mental deficiency and skeletal changes.

Hatzoecher (1708) was one of several authors to report cases of intra-uterine fracture of the lower limbs, which were attributed to psychic influences upon the pregnant mother. Camurati (1930), who made this reference, suggested that many of the cases must have been examples of pseudarthrosis of the tibia. The connection of this condition with neurofibromatosis was suggested by Ducroquet (1937) and confirmed by subsequent authors.

CASE REPORT

A boy of two years fell on the grass but no definite injury was recognised until three days later when it was noticed that he was not using his left arm. A radiograph showed an undisplaced transverse fracture of the distal half of the ulna and a greenstick fracture of the

![Fig. 1](image1)

![Fig. 2](image2)

Figure 1—Radiographs of the forearm indicating an apparently simple injury in a 2-year-old boy. Figure 2—Delayed union with deformity of the radius following three months in plaster.
Eighteen months after the original injury, the radius is united. The tapering of the ulnar components is exaggerated and the gap between them further increased. Figures 7 and 8—Lysis of the bone graft during the six months after surgery. Figure 9—Radiograph of the forearm at 15 years of age showing appearances in the ulna reminiscent of pseudarthrosis of the tibia, dislocation of the head of the radius and incomplete restitution of its medullary canal.
radius at the same level (Fig. 1). The limb was rested in a plaster splint and, though the records indicate that the fracture initially became firm, it was later found that a severe angular deformity had developed in the radius. Radiographs at three months confirmed delayed union with dorsal angulation of the distal fragments (Fig. 2). This was corrected by open reduction (Fig. 3), the position being supported by plaster splintage. Six months after injury the deformity had recurred in the radius, and the ulnar fragments were tapered towards the fracture site (Fig. 4). Plaster splintage was continued until fourteen months after injury. The radiographs indicated, in addition, non-union of the radius (Fig. 5). A fibrous union was excised and the limb was again supported in plaster for five months. It was then eighteen months from the original injury and the radius was united (Fig. 6). Three months later bone blocks taken from the full thickness of the ilium were inserted to bridge the gap in the ulna. Radiographs taken over the ensuing six months suggest initial viability and subsequent lysis of the graft (Figs. 7 and 8).

The patient ceased to attend hospital until he was fifteen years of age. He then complained of weakness of grip and deformity of the forearm (Fig. 10). The range of movement at the elbow and wrist was full except for some restriction of pronation. Radiographs indicated that union of the radius was sound, though failure of development of the medullary canal suggested persisting local abnormality. The appearance of the ulna with tapered atrophic components resembled that seen in pseudarthrosis of the tibia. In addition the head of the radius was dislocated (Fig. 9).

**COMMENT**

The patient was of average intelligence and without physical stigmata apart from café-au-lait patches on the trunk (Fig. 11). Two of his five siblings, his mother and his maternal grandmother also bore café-au-lait markings but to a lesser extent. Thannhauser (1944) postulated that café-au-lait patches are as much an indication of neurofibromatosis as the...
neurofibromatous nodules themselves, though McCarrroll (1950) found isolated patches in 20 per cent of a group of 100 nurses. In the case presented, the behaviour of the forearm bones in producing deformity, the reluctance to unite and the lysis of the bone grafts, together with the radiological appearances, strongly suggest the behaviour and radiological appearances seen typically in the tibia. The presence of café-au-lait patches supports the view that it is a similarly related manifestation of neurofibromatosis.

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REFERENCES


