Macrodactyly of the hand and foot

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Summary. Five patients with macrodactyly are described. The foot was affected in 3 of them and the hand in 2. Two patients were operated on and the histological features are described. The literature is fully reviewed and the aetiology is discussed.

Résumé. Description de cinq sujets porteurs d'une macrodactylie. Elle atteignait trois fois le pied et deux fois la main. Deux malades furent opérés et les constatations histologiques sont présentées. La littérature a été revue de façon exhaustive. L'étiologie est discutée.

Key words: Macrodactyly, Fingers, Toes

Local gigantism of the fingers or toes (macrodactyly) is uncommon and most reports are of single cases of only small series. Most sources indicate that macrodactyly affects the hand more often than the foot. Barsky [3] reported a large number of cases of congenital macrodactyly of the hand with a review of the literature of the past 140 years. Since then, a few more reports of macrodactyly of the hand have been published [2, 7, 10, 15, 18, 19, 23, 26, 27, 28]. In 1925 Feriz [11] reported a large series of cases of macrodactyly of the foot. A few more cases were added [2, 8, 9, 12, 19, 22, 28, 29]. No reports of macrodactyly of the foot during the past 5 or 6 years could be found.

The present paper describes 5 cases (2 affecting the hands and 3 affecting the feet) seen by the authors during the last 5 years.

Materials and methods

All 5 patients with macrodactyly of the hand or foot were male and between 3½ and 16 years of age. The foot was affected in 3 patients and the hand in 2. Two came to the hospital specially for the treatment of the deformity, and the other 3 cases were recognised by chance when they had come to the hospital as attendants of other patients. A detailed history and clinical examination was carried out in all cases, and radiographs were taken. Specimens obtained from operation were examined microscopically.

There was no family history of a similar condition in any of the patients and none had features suggestive of neurofibromatosis, haemangioma, lymphangioma, hamartoma or any other congenital deformity elsewhere in the body. The deformity, which was noticed by the parents of all the patients soon after birth, was progressive. The patients who were admitted for operation seemed only partially satisfied. One patient who had macrodactyly of the hand underwent amputation of the enlarged finger, whereas one patient with macrodactyly of the foot underwent several operations, including staged defatting with subsequent skin necrosis, and delayed staged defatting with subsequent skin necrosis and delayed healing of the wound with infection.

The clinical and other features are summarised in Table 1, and illustrative clinical and histological photographs with examples of radiographs are shown in Figs. 1–5.

Histology

Specimens were prepared from the amputated finger and the excised fibrofatty tissue of the foot. In the finger all the elements from skin to bone were hypertrophied. The median nerve in the palm was very much thickened and the digital branches were as thick as the sciatic nerve. Nerve bundles were normal and there was no evidence suggesting neurofibromatosis. The skin showed thickening with dermal fibrosis and flattening of rete pegs as reported by Tuli et al. [21]. The bone showed marked periosteal thickening and fibrosis. The trabeculae were thin and poorly calcified. Increased fibroblastic activity near periosteum as previously reported [18] could not be seen. Fibrofatty tissue obtained from the foot showed tissue resembling the subcutaneous tissue of an adult [3]. It consisted of large and small lobules of adipose tissue separated by fibrous septa of variable thickness.

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Fig. 1. a Clinical photograph showing involvement of all the 5 toes with swelling of the plantar aspect of the foot; b Photograph of the same foot showing dorsal curling of the toes; c Histology of the excised subcutaneous fat showing an adult type of fat (× 100)

Fig. 2. Anteroposterior and oblique radiographs of a patient with pedal macrodactyly