tempt to replace the necrotic and fibrotic tissue by hopefully viable cartilagenous tissue. As a second procedure, osteotomy, was anticipated. This has, however, not yet been necessary as the perichondrial graft has shown a substantial regenerative capacity (Homminga et al. 1990) and reconstituted the mechanical axis, simply by growth. As far as we know, such a pronounced regeneration in a knee, restoring normal mechanics, has not been reported before.

The present clinical status of this patient is impressive, with no pain and a high level of activity. However, complications have been reported in one third of the patients after 2.5–5 years (Bulstra et. al. 1993). Perichondrial grafting must be regarded as an experimental procedure and this patient runs a clear risk of future failure, either by loosening or wear of the graft.

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Monstrous congenital macrodactyly with syndactyly of the foot—a case report

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A 16-year-old Ovambo girl came cumbrously walking to the Oshakati Regional Hospital with an extremely bulky congenital deformity of the left foot (Figure 1). This girl, weighing 48 kg and 155 cm tall, showed also a thickening of the left leg and major labium. The left foot was 48 cm long, the right was 25 cm long. Chromosome analysis (blood) revealed a normal karyotype 46 XX.

The first 4 toes were grossly enlarged and misplaced, including the metatarsal bones. The oversized toes were stiff. The distal end of the big toe appeared as a short stump out of the dorsum of the foot. The normal-sized little toe looked like a small rudiment on this monstrous foot.

Radiographically, the misplaced metatarsal and phalangeal bones were markedly enlarged, but normally linked and articulated (Figure 2). The tarsal bones were also enlarged compared with the normal foot.

. The foot was amputated through Lisfranc's joint. Lipectomy on the medial aspect of the lower leg and thigh, and on the foot sole were also performed. After lipectomy on the sole of the left foot, the skin on the frontal and frontomedial plantar aspects of the stump

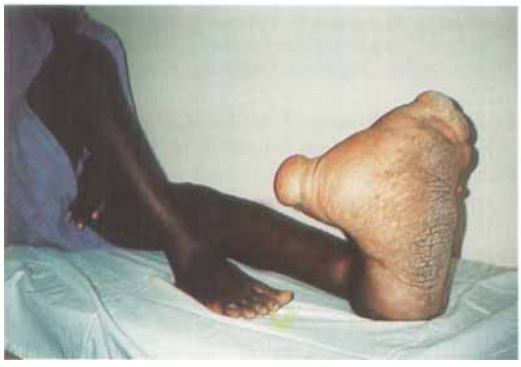

Figure 1. Monstrous macrodactyly with syndactyly of the left foot in a 16-year-old girl.

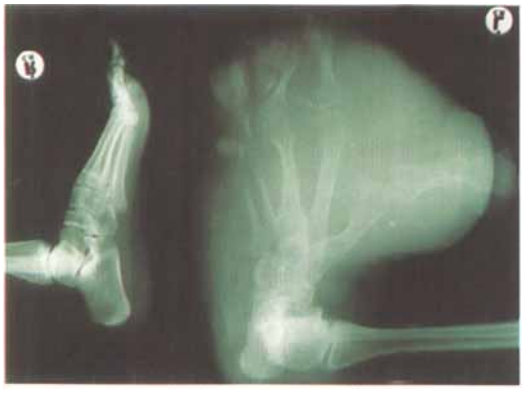

Figure 2. Extreme macrodactyly of the misplaced metatarsal and phalangeal I–IV bones of the left foot. Note also the enlarged tarsal bones.

became partly necrotic and required a skin transplantation (Figure 3). After surgery and improved function, the patient could walk more easily and she was satisfied with the foot, now 13 cm long.

Histology of the excised material revealed benign fatty tissue infiltrating the skeletal muscle by separating its fibers into small groups. It also separated and surrounded normal vascular and nervous structures. The hypertrophied bones showed mild-to-moderate periosteal thickening. Nerve axon and sheath structures were not affected, either by hyperplasia or by neurofibromatosis.

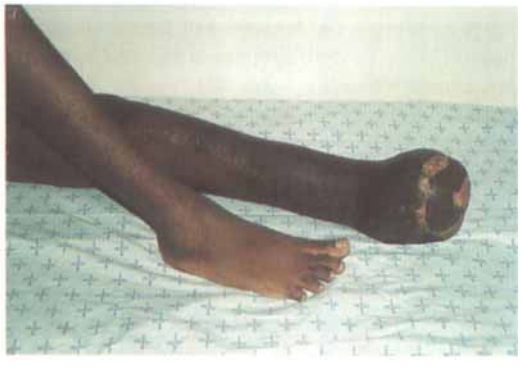

Figure 3. Postoperative findings.

Discussion

2 types of macrodactyly are distinguished: that due to bone overgrowth alone and that due to neurofibromatosis and with lymphatic overgrowth (Barsky et al. 1964, Lauschke 1988).

Holmes (1869) distinguished between symmetrical and asymmetrical varieties of macrodactyly. In the symmetrical type, all morphological structures of the phalanx are proportionally enlarged. In the asymmetrical type, the different structures are variously enlarged by fatty excrescences and overdevelopment of the joint end of the phalangeal bones.

Our case is of the asymmetrical type because of the disproportionate enlargement of the different structures of the forefoot within the monstrous bulk, where only the little toe was normal.

Our patient had a hyperostotic variant of macrodactyly combined with an enormous excess of surrounding muscle fiber separating fatty tissue. A similar case of a hand in a 6-year-old Ovambo girl, without any function, due to a foot-like congenital macrodactyly with syndactyly, operated on in the same institution, was reported by Lauschke (1988).

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