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CASE REPORT

Bone lengthening for correction of limb length discrepancy in a patient with Klippel-Trenaunay syndrome
A case report

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Klippel-Trenanay Syndrome (KTS) consists of a triad of cutaneous haemangioma, bone and soft tissue hypertrophy and venous varicosities (*4*). The aetiology of this syndrome is unknown but some authors have suggested that it results from a mesodermal abnormality that occurs during fetal development (*5*).

Bone hypertrophy usually results in limb length discrepancy, with the affected leg being the longer one. In most of the patients with limb length discrepancy due to KTS, operative treatment of the affected leg takes place before skeletal maturity.

CASE REPORT

A 21-year-old male with KTS presented for treatment of his leg length inequality. The patient showed an affected right side with aplasia of the great saphenous vein, cutaneous haemangioma extending to the right hemipelvis, back and abdomen, arteriovenous fistulae and leg hypertrophy. He had so far received no treatment for his limb length discrepancy.

Using a scanogram examination we found the right leg 8.5 cm longer, with the femur being 4.5 cm and the tibia 4 cm longer.

Lengthening of the healthy left limb was decided in order to avoid all the postoperative complications that might occur after operating on a KTS limb, and also because the patient came to us after skeletal maturity.

The femur was first lengthened. The operation was carried out using a combined technique proposed by Paley *et al* (*6*) : intramedullary nailing was performed using a Russell-Taylor nail (380/10 mm, Smith and Nephew Orthopaedics, Memphis, Tennessee) which was locked proximally. An external fixator was then applied (Orthofix hybrid external device, Bussolengo, Verona, Italy). The operation was completed with an osteotomy of the left femur using a Gigli saw (fig 1). Five days postoperatively, distraction commenced with 0.25 mm quarterly every day. After 47 days the desired lengthening of 4.5 cm was achieved.

Eight days later the patient entered the clinic and a second operation took place. The external fixator was removed from the femur, after distal locking of the intramedullary nail and an Orthofix LRS external fixator was applied to the tibia along with

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Fig. 1. — Radiograph of the left femur during lengthening. Intramedullary nail locked proximally.

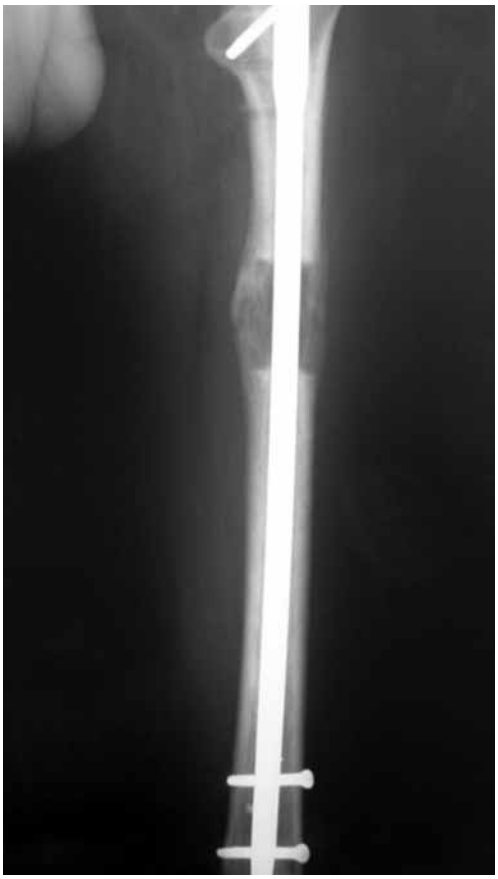

Fig. 2. — Radiograph of the left femur one month after distal locking of the nail and removal of the external fixator.

a syndesmotic screw in the distal tibiofibular joint. A 1-cm long segment from the fibula was excised after osteotomy, and operation was completed with a subperiosteal tibial osteotomy. Distraction commenced 5 days later with 0.25 mm quarterly every day and the desired length of 4 cm was achieved after 42 days. Both legs now had the same length (fig 2, 3).

The external device from the tibia was removed, 6 months after the second operation and the patient was allowed to walk freely. Regarding complications, stiffness of the left knee joint was treated by manipulation during the second operation and stiffness of the left ankle joint developed during tibia lengthening was treated with physiotherapy. No other complications were noted.

At the last follow-up, one year later, the limbs were equalised and the patient had full range of motion of the knee and ankle joints as well as full level activity (fig 4a, 4b, 5, 6)

DISCUSSION

KTS consists of a triad of cutaneous haemangioma, bone and soft tissue hypertrophy and venous varicosities (*5*). This syndrome often results in major malformations. Children with KTS almost always appear to have overgrowth of the leg secondary to the increased blood supply. They also have multiple arteriovenous fistulae which are not localised to any one portion of the lower extremity (*4*). Patients usually appear with the extremity warm, with many dilated superficial veins and cutaneous haemangiomas. The functional disabilities of limb-length inequality, secondary pelvic tilt, scoliosis and joint contractures contribute to

Fig. 3. — Radiograph of the left tibia. 4cm of lengthening. Satisfactory new bone formation at the distraction site.

increasing physical impairment. Sometimes the altered haemodynamics may result in severe cardiac failure, and amputation of the involved leg may be necessary to reduce the overload of the myocardium (*4*).

Surgical treatment in KTS is controversial. The limb hypertrophy is most often unilateral. The most common treatment of the leg length discrepancy, as referred in the international bibliography, is epiphysiodesis (*1-4, 5, 7, 9*). The operation must be performed at the age of 10-14 years, and the difference in length between the two legs must be over 2-2.5 cm. The timing of surgery is crucial, as an early epiphysiodesis may result in aa longer normal leg (*2*). For patients who have resched skeletal maturity, bone shortening can be attempted but consolidayion of union may be delayed (*7*).

Other treatments consist of venous freeing of the longer side and ligation of the popliteal vein on the shorter side (*8*), stripping of varicose veins (*2, 3*).

Fig. 4a. — Photograph of the patient at the last follow up. Limbs equalized.

Fig. 4b. — Photograph of the patient at the last follow up, demonstrating full range of motion of knee and ankle joints.

Any surgical procedure performed on the affected limb is associated with bleeding and a higher rate of wound complications than normal (*1, 4*).

Little attention has been given in the literature to the possibility of lengthening the healthy shorter leg. Operating on the healty limb is always a diffi-

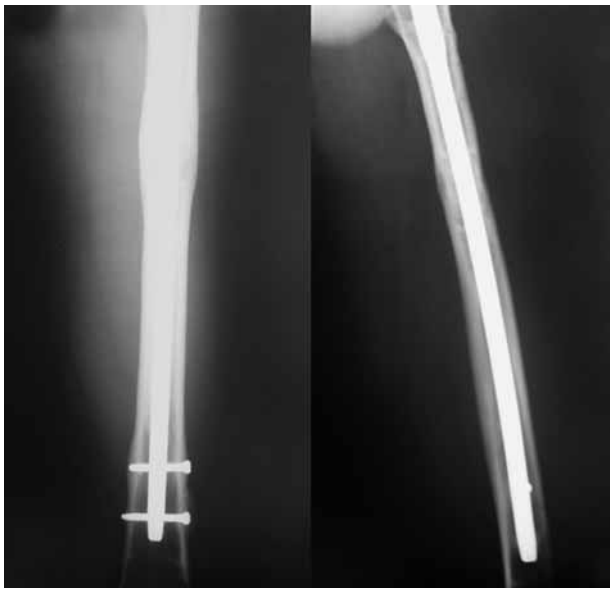

Fig. 5. — Radiograph of the left femur at the last follow-up.

cult decision and the patient must be informed and give formal consent.

We report such a case of a 21-year-old patient with untreated leg hypertrophy due to KTS, on whom we operated onr the healthy shorter limb, lengthening both segments (femur and tibia).

We believe the present case suggests that there is a treatment, with excellent results and minor complications, for untreated KTS leg hypertrophy even if the patient has passed the age of skeletal maturity. We also believe that it is a good choice even in adolescents, in order to avoid the high risk of morbidity from the operation on the affected leg.

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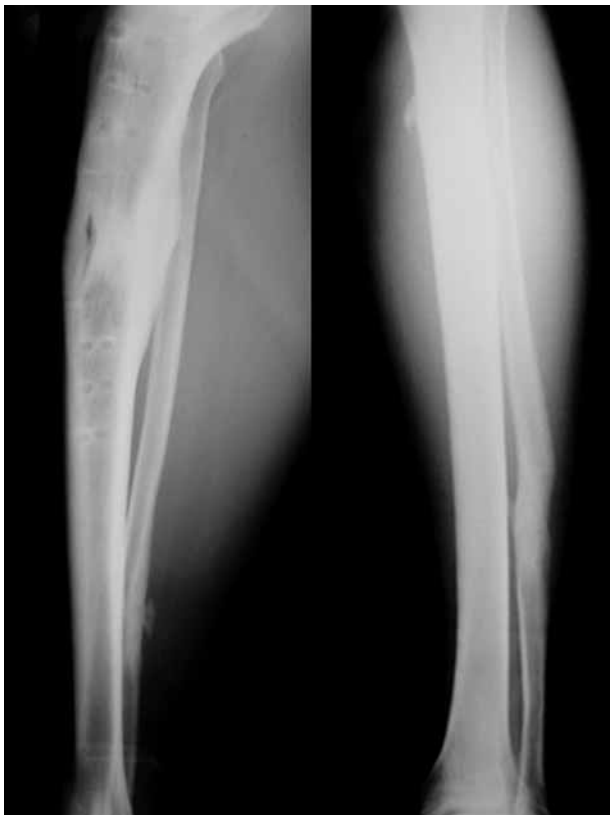

Fig. 6. — Radiograph of the left tibia at the last follow-up.

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