Lengthening of the normal tibia in a patient with hemihypertrophy caused by Klippel-Trenaunay-Weber syndrome: a case report

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ABSTRACT

We report a case of Klippel-Trenaunay-Weber syndrome in a 31-year-old woman who presented with hypertrophy of the left leg. She had severe osteoarthritic changes in the left hip joint secondary to the lack of acetabular coverage of the femoral head as the result of lateral inclination of the pelvis owing to leg-length discrepancy of 4 cm. The centre-edge angle (coverage ratio of the acetabulum to the femoral head) was improved from 15º to 33º after a foot lift. She underwent osteotomy and lengthening of the normal contralateral tibia using a Taylor spatial frame. Hip arthroplasty could be avoided as osteoarthritic changes of the hip joint had improved.

Key words: external fixators; Ilizarov technique; Klippel-Trenaunay-Weber syndrome; leg length inequality

INTRODUCTION

Klippel-Trenaunay-Weber syndrome is a congenital disorder characterised by capillary malformation (hemangioma), venous malformation (arteriovenous fistula), and/or soft tissue and/or bony hypertrophy of the limb that causes limb length discrepancy. Stricter criteria for Klippel-Trenaunay-Weber syndrome comprise 2 major features: vascular malformation (capillary, venous, arteriovenous, and lymphatic malformations) and disturbed growth of bone and/or soft tissues (hypertrophy and infrequently hypotrophy). There is no racial and gender predilection for the condition. Familial occurrence is rare. Its pathogenesis and association with genetic defects is not well understood. It may be caused by mesodermal abnormality during foetal development, leading to the maintenance of microscopic arteriovenous communications in the limb bud. An increase of blood flow may lead to nevus formation, superficial varices, and hypertrophy of the juxtaepiphysial cartilage.

We report one such case in a patient with severe osteoarthritic changes in her left hip joint because of insufficient acetabular coverage of the femoral head secondary to lateral inclination of the pelvis caused by a leg-length discrepancy. She underwent lengthening
of the normal contralateral tibia using a Taylor spatial frame.

CASE REPORT

In June 2004, a 31-year-old woman presented with Klippel-Trenaunay-Weber syndrome leading to hypertrophy of the left lower leg and limping. Her complaint was severe left hip joint pain. Venous varicosities were spread over the lower limb, and a hemangioma (3 cm in diameter) was noted at the dorsum of her foot (Fig. 1). The skin temperature on the affected side was 4°C higher. The affected hip joint was tender and swollen, and the range of motion was limited (flexion, 90°; abduction, 5°; external rotation, 30°; internal rotation, 0°). Her upper extremities had no discrepancy, and she was not obese (body mass index, 23.7 kg/m²).

Radiographs revealed that the length of the right lower limb was 65 cm and that of the left was 69 cm (Fig. 2). Leg-length discrepancy mainly occurred in the tibia (3 cm difference). Mechanical lines (from the centre of the femoral head to the centre of the ankle) of both lower limbs passed through the centres of the knee. In a standing position, the left hip joint inclined to the right side, with a centre-edge angle of 15°, which reduced the acetabular coverage of the femoral head. The inclination of the pelvis induced a reversible scoliosis, with a Cobb angle of 13° (Fig. 2). End-stage osteoarthritic changes were noted in her left hip.

Figure 1  A 31-year-old woman with Klippel-Trenaunay-Weber syndrome showing (a) hypertrophy of the left leg and limb-length discrepancy, (b) venous varicosities on the left lower limb, and (c) a hemangioma (3 cm in diameter) at the dorsum of the foot.

Figure 2  (a) The length of the right leg is 65 cm and that of the left is 69 cm. Leg-length discrepancy is 1 cm at the femur and 3 cm at the tibia. (b) The inclination of the pelvis induces reversible scoliosis, with a Cobb angle of 13° between T9 and L5.
joint, in which cystic changes at the acetabulum and narrowing of the joint space were apparent (Fig. 3). A foot lift of 4 cm on the right side made the pelvis horizontal, and the centre-edge angle was improved to 33° (Fig. 3).

Surgical intervention was necessary because of left hip joint pain and end-stage osteoarthritis. Simple acute shortening of the affected limb might induce infection or massive bleeding, because of the varices, arteriovenous fistulae, and lymphatic abnormality. Moreover, the patient did not wish to become shorter.

Therefore, lengthening of the normal right tibia was performed using the Taylor spatial frame (Fig. 4). The lengthening period was 107 days, and the external fixator was applied for 750 days until bone union. The amount of lengthening achieved was 4 cm; the distraction index was 26.8 days/cm, and the external fixation index was 187.5 days/cm. The proximal half pin and the distal ring broke at month 8 (4 months after the lengthening was completed). Reconstruction under anaesthesia was conducted. There was no other severe complication.
At the 59-month follow-up, the hip joint pain had disappeared; the joint space had enlarged; and the centre-edge angle had improved to 35° (Fig. 5). Hip arthroplasty could be avoided as osteoarthritic changes of the hip joint had improved.

DISCUSSION

Acquired leg length discrepancy can be caused by fracture, infection, epiphyseal injury, tumour, and coxa magna after open reduction of developmental dysplasia of the hip. Congenital causes for leg length discrepancy include several types of hemihypertrophy (asymmetry between the right and left sides of the body more than the normal variation). They include idiopathic hemihypertrophy (associated with intra-abdominal malignancy such as Wilms tumour and hepatoblastoma), Beckwith-Wiedemann syndrome, Klippel-Trenaunay-Weber syndrome, proteus syndrome, melorheostosis, and neurofibromatosis. When a greater discrepancy is noted or anticipated with continued growth, epiphysiodesis or tibial osteotomy should be considered. The lower femoral, upper tibial, and upper fibular epiphyses are the most common sites of overgrowth. The rate of overgrowth in Klippel-Trenaunay-Weber syndrome is unpredictable, but limb length discrepancy rarely increases after age 12 years. Degenerative arthropathy secondary to the Klippel-Trenaunay-Weber syndrome sometimes presents similarly to haemophilic syndromes with frequent haemarthroses.

In our patient, surgery on the healthy leg and long duration of treatment were disadvantageous. The external fixation index was longer than usual, owing to breakage of the proximal half pin and the distal ring during bone maturation. The broken point on the ring was anterior, close to the hole, and likely to be caused by ductile stress. Instability might have damaged the regenerated callus bone. Nonetheless, full weight bearing was preferable during the maturation period, and therefore ductile stress was unavoidable.

The Taylor spatial frame enables easy modification of the correction and requires only minimally invasive surgery. Slight translation and angulation usually occur during a prolonged correction, but can be corrected simultaneously by controlling the length of the 6 struts. These are advantages of the Taylor spatial frame over the Ilizarov external fixator, which requires staged correction. The Taylor spatial frame is more accurate because the struts are calibrated so every turn provides an equal amount of movement, whereas the Ilizarov strut adjustment requires subjective interpretation.
REFERENCES