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Management for Gait Disturbance and Foot Pain in a Patient with Klippel-Trenaunay-Weber Syndrome : A case report

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Abstract

Background : Klippel-Trenaunay-Weber syndrome (KTS) is a rare congenital medical condition characterized by complex vascular malformation. KTS consists of a classic triad of capillary malformation (hemangioma), venous malformations and bone or soft tissue hypertrophy causing limb asymmetry. The aim of this report is to describe management for gait disturbance and foot pain in a Patient with KTS using custom-made total contact insole.

Case presentation : A 32-year-old man with KTS presented with a 3-year history of gait disturbance on hard surface due to right first toe pain and Achilles tendon tightness. The patient had soft tissue hypertrophy, varicose veins and port-wine stains over the right lower limb associated with KTS. True leg length discrepancy was 2 cm. We prescribed custom-made total contact insole to protect his deformed foot and correct leg length discrepancy. The insole of right side included wedge shaped heel lift and the insole of left side included full length lift to add extra support on unaffected side. Also, we provided compression stocking and physiotherapy including manual lymphatic drainage for lymphedema and stretching exercise for tightness in right lower extremity. At 3 years follow-up, postural alignment including pelvic obliquity was improved using a custom-made total contact insole. The degree of scoliosis and foot pain were also reduced.

Conclusion : An individualized and multidisciplinary approach is essential regarding the complexity of comorbidities in patients with KTS. For patients with KTS, orthotic management should be considered to prevent and correct deformities related to KTS. Active orthotic management, compression stocking and physiotherapy can enhance the quality of life and function in patients.

Key Words : deformity, Klippel-Trenaunay-Weber syndrome, leg length discrepancy, orthosis, vascular malformation

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Ⅰ. Introduction

1. Research background and needs

Klippel-Trenaunay-Weber syndrome (KTS) is a congenital medical condition characterized by complex vascular malformation. KTS consists of a classic triad of capillary malformation (hemangioma), venous malformations and bone or soft tissue hypertrophy causing limb asymmetry (Oduber et al., 2008). Klippel and Trenaunay first described the classic triad of cutaneous hemangioma, hemihypertrophy and varivose veins in 1900 (Klippel & Trenaunay, 1900). Later, Weber reported cases with arteriovenous fistulae (Weber, 1907). Many orthopedic problems associated with KTS including limb hypertrophy and atrophy, limb-length discrepancies, polydactylia, ulcerations, neuropathies, congenital dislocation of hip joint, scoliosis, spina bifida have been reported (Carvell & Chopin, 1984; McGrory et al., 1991). The regional distribution of abnormalities varies as follows: lower limb, 74.00 % of cases, upper limb 8.00 % of cases: combination, 18.00 % of cases, and unilateral involvement, 72.00 % of cases. All components of the affected limb, including muscle, bone and connective tissues are hypertrophied. Clinical presentation of KTS is variable from negligible limb hypertrophy to life-threatening complications (Alwalid et al., 2018).

Up to now, literatures about KTS usually have focused on diagnostic procedures. KTS is associated with significant comorbidities such as pain, edema, ulcerations and pruritis (Wang et al., 2017). However, no consensus has been achieved for its proper treatment and management until now (Asghar et al., 2020). The treatment in KTS has been described as a conservative treatment for lymphedema and surgical treatment of varicose veins, venous malformations or lymphedema.

2. Research purpose

The aim of this report is to describe management for gait disturbance and foot pain in a Patient with KTS using custom-made total contact insole.

Ⅱ. Case presentation

A 35-year-old man was diagnosed with Klippel-Trenaunay-Weber syndrome and presented with unilateral lower limb involvement. He got Achilles tendon lengthening operation on his right leg 17 years ago. He presented with a 3-year history of gait disturbance on hard surface due to right first toe pain and Achilles tendon tightness. On physical examination, port-wine stains, varicose veins and hypertrophy of soft tissues were spread over the right lower limb, and deformity was noted at his right toes [Fig 1 (a)]. Diffuse muscle atrophy on thigh, calf, and pretibial part of right lower leg was shown, and range of motion in his right knee joint was limited by 105 °. True leg length discrepancy measured from the anterior superior iliac spine to the tip of medial malleolus between the longer (right) and shorter (left) legs was 2 ㎝. Severe soft tissue hypertrophy ensuing toes deformity in the affected side was also observed in foot x-ray [Fig 1 (b)]. Vascular malformation and soft tissue hypertrophy Right lower leg MRI revealed extensive engorged vascular structures suggested low flow type vascular malformation was noted [Fig 1 (c)]. Lymphoscintigraphy with 99 mTc-phytate, 148 MBq (4 mCi), demonstrated abnormal lymphatic vessels in right lower extremity and right buttock was found in the early phase imaging and no uptake in right inguinal lymph node was found in the delay phase imaging. It suggested there was combined lymphatic malformation [Fig 1 (d)]. Spine x-ray revealed that right hip joint was inclined to the left side (pelvic obliquity of 6.9 °) in standing position, inducing a scoliosis, with a Cobb's angle of 23 ° [Fig 2 (a)].

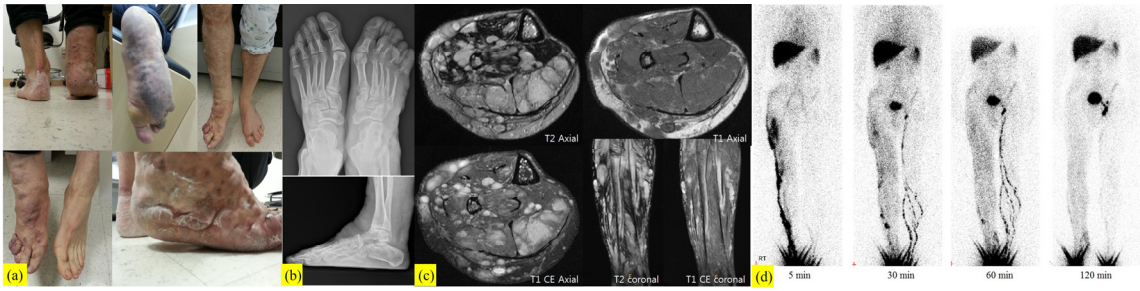

Fig 1. Right lower leg deformity related to vascular malformation and lymphedema

(a) Photographs of affected lower leg showing port-wine stains, varicose veins and hypertrophy of soft tissues
(b) Foot x-ray showing soft tissue hypertrophy and deformity
(c) MRI shows extensive dilated vascular structures in the right lower leg involving subcutaneous layer, and multiple muscle compartments, and intraosseous involvement of tibia and fibula. There is no flow void and pulsation artifact, suggesting a low flow type vascular malformation
(d) Lymphoscintigraphy of the both lower extremities was obtained at 5, 30, 60, and 120 minutes after intradermal injection of 148 MBq/0.4 mL 99mTc-phytate. The early phase image showed abnormal lymphatic vessels in right lower extremity and right buttock and the delayed phase image revealed no uptake of the right inguinal lymph nodes, suggesting the lymphatic malformation in right lower extremity and right buttock

The custom-made total contact insoles (Biomechanics, Goyang, South Korea) were prescribed to correct leg length discrepancy and inclined right hip joint. It was designed to protect his deformed foot and prevent further damage. Because the shape of the left and the right foot were very different, the insole was made separately to suit the shape and characteristics of each foot. Soft tissue hypertrophy was spread on the dorsum, sole and toes,

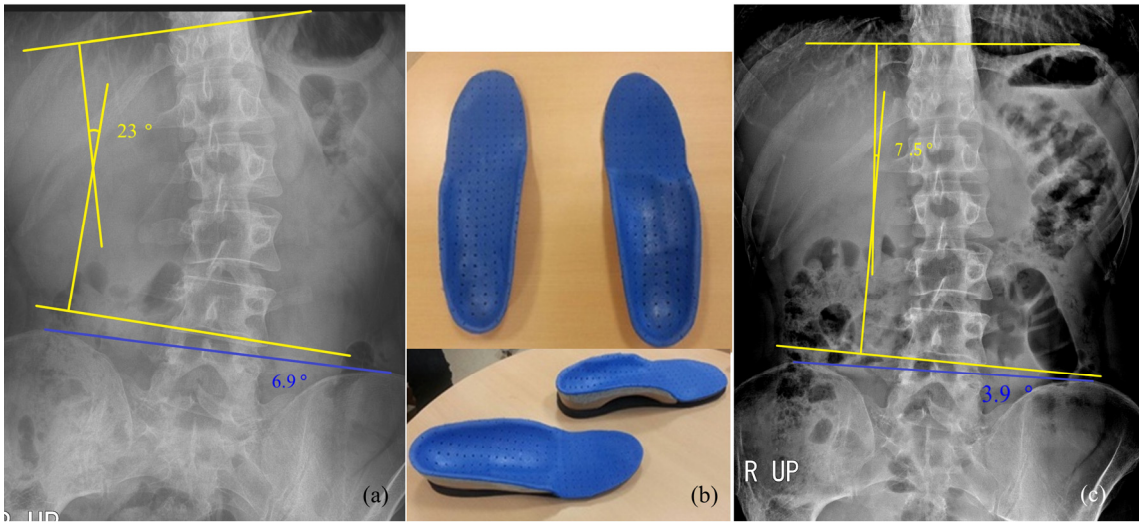

Fig 2. Total contact insoles for postural alignment

(a) L-S spine AP image showing inclined right hip joint (pelvic obliquity of 6.9 °) and scoliosis with a Cobb's angle of 23 °
(b) Custom-made total contact insoles
(c) L-spine AP image showing improved pelvic obliquity (3.9 °) and scoliosis with a Cobb's angle of 7.5 ° after using a custom-made total contact insoles

especially the 2nd, 3rd, 4th, 5th toes. The contour of the sole of the foot was uneven. Hypertrophy of the forefoot and midfoot was more severe than the rearfoot. The insole of right side included wedge shaped 1.2 ㎝-height heel lift because heel contact did not occur when walking due to right foot deformity. The insole of left side included full length lift (0.2 ㎝-height at toe, 0.6 ㎝-height at metatarsal, 1.0 ㎝-height at heel) to add extra support on unaffected side [Fig 2 (b)]. The orthotic materials in both total insoles were polyethylene (including Plastazote), ethyl vinyl acetate (EVA) and polyurethane (including PORON□). It was recommended to wear the insole for about an hour on the first day after fitting, then increase the wearing time by 30 minutes a day from the next day, and progress to full-time wear when wearing half of the day became comfortable. He wore the brace about 10 hours a day.

It improved postural alignment and gait pattern of the patient. At 3 years follow-up, postural alignment including pelvic obliquity was improved using a custom-made total contact insole. The degree of scoliosis and foot pain were also reduced [Fig 2 (c)]. L-spine AP image showing improved pelvic obliquity (3.9 °) and scoliosis with a Cobb's angle of 7.5 ° after using a custom-made total contact insoles.

Ⅲ. Discussion

KTS is a complex vascular malformation syndrome with multi-system involvement. This is a rare syndrome, with a wide spectrum of clinical findings that can manifest during infancy and can progress throughout childhood and adults. The prognosis of KTS is related to the severity of vascular malformations. The mainstay is symptomatic care with medical management, with only a few cases needed surgical intervention. Due to the varying presentations and severity of disease, treatment strategies need to be individualized for every patient with KTS. Also, a multidisciplinary approach is essential regarding the complexity of comorbidities. In the present case, we provided compression stocking and physiotherapy including stretching exercise for tightness in right lower extremity. During follow-up period, cellulitis and skin ulceration was treated antibiotics. Unlike the present case, if patients with KTS have symptomatic varicose vain or localized arteriovenous malformations, surgical removal can be considered. Also, endovascular surgery can be necessary for the hemodynamic alterations related to arteriovenous malformations (Pandey et al., 2019).

Leg length discrepancy is commonly observed in patients with KTS. It is secondary to underlying soft tissue growth but can be associated with long bone hypertrophy. The management of leg length discrepancy depends upon the severity. Although severe difference in leg length more than 5 cm should be managed by surgical approaches including shortening, lengthening and epiphysiodesis, mild leg length discrepancy cam be resolve only with orthosis including insole and shoe lift. Conservative treatment including insoles, shoe lift and orthosis is typically used to patients with leg length discrepancy between 2 cm and 5 cm. Especially, leg length discrepancy of up to 2 cm can be corrected with heel wedge insoles. Orthosis can help reduce the difference in leg length and the pain that can occur when a deformed foot is wearing ready-made shoes (Vogt et al., 2020). Even though neutralization of the leg length discrepancy by insole could not correct the whole lumbar curvature, leg length equalization could reduce some compensatory lumbar scoliosis (Raczkowski et al., 2010).

Ⅳ. Conclusion

Patients with KTS commonly suffer from foot pain. leg length discrepancy, and gait disturbance. More interests on pain and quality of life are needed to care the patient. Using the orthosis should be considered in management of

patients with KTS. The custom-made moulded total contact insoles can help patients with KTS to reduce foot pain, leg length discrepancy and improved gait function. An individualized and multidisciplinary approach including orthosis, compression stocking and physiotherapy is essential for patients with KTS

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