Case Report

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An unusual variant of Klippel-Trenaunay syndrome

Rupesh Mandava^{1*}, Divya Ganesan², Rajeswaran Rangaswami¹, Venkata Sai¹, R. Radhakrishnan³

¹Department of Radiology, Sri Ramachandra University, Chennai, India

³Department of Vascular Surgery, Sri Ramachandra University, Chennai, India

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*Correspondence: Dr. Rupesh Mandava,

E-mail: rupeshmandava@gmail.com

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ABSTRACT

A twenty four year old male presented with complaints of pain and recurrent non healing ulcer over the medial aspect of the right leg. He had dilated and tortuous veins with mild hypertrophy of the right lower limb. Ultrasound and ascending lower limb venogram revealed absence of common femoral, superficial femoral and iliac veins on the right side. The popliteal vein was seen to continue as a medial venous channel, crossing in the pelvis to the left side and draining into the left common femoral vein. Klippel-Trenaunay syndrome with dilated medial venous channel draining into the opposite common femoral vein have so far not been described in the literature to our knowledge. This case is presented for its rarity.

Keywords: Varicose veins, Venogram, Ultrasound, Klippel-Trenaunay syndrome

INTRODUCTION

Varicose veins are dilated and tortuous superficial veins. It is more common in women than in men. Primary varicose veins develop due to defective valves, postural straining or from congenital absence of the valves in the deep venous system. Klippel-Trenaunay syndrome is one such presentation of primary varicose veins.

CASE REPORT

A twenty four year old male presented with complaints of pain and recurrent non healing ulcer over the medial aspect of the right leg and gradually increasing swelling of leg for the past two years. He also had history of dilated and tortuous veins in the right lower limb since childhood. On examination, there were dilated and tortuous superficial veins with port wine stain pigmentation all over the right lower limb. Mild

hypertrophy of the right lower limb was noted (Figure 1 A. B).

Colour Doppler imaging of the right lower limb revealed complete absence of the common femoral, superficial femoral and iliac veins (Figure 2 A). The popliteal vein was seen to continue as a medial venous channel in the thigh, crossing in the pelvis to the left side and draining into the left common femoral vein. This medial venous channel appeared dilated (Figure 2 B).

Ascending right lower limb venogram was performed which confirmed these findings in the form of absence of the common femoral, superficial femoral and iliac veins on the right side with popliteal vein continuing as a dilated medial venous channel and draining into the left common femoral vein (Figure 3 A-D).

²Department of Radiology, Sakthi Hospital and Research Centre, Chennai, India



Figure 1: Mild hypertrophy of the right lower limb was noted.

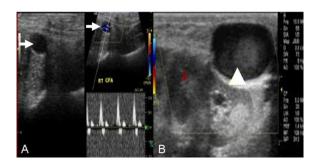


Figure 2: (A) Colour Doppler imaging of the right lower limb revealed complete absence of the common femoral, superficial femoral and iliac veins. (B) The popliteal veins was seen to continue as a medial venous channel in the thigh, crossing in the pelvis to the left side and draining into the left common femoral vein. This medial venous channel appeared dilated.



Figure 3: Ascending right lower limb venogram was performed which confirmed these findings in the form of absence of the common femoral, superficial femoral and iliac veins on the right side with popliteal vein continuing as a dilated medial venous channel and draining into the left common femoral vein.

A diagnosis of variant of Klippel-Trenaunay syndrome was made. The patient was managed conservatively with antibiotics, limb elevation and compression garments.

DISCUSSION

Klippel-Trenaunay is a syndrome triad which has an association of varicose veins; limb- length discrepancy and port- wine stains on the skin. It is otherwise called as Naevus vasculosus osteohypertrophicus. It is a sporadic non hereditary rare mesodermal abnormality. Klippel and Trenaunay described this triad and later Parke Weber included arteriovenous fistula. Both coined together and named the syndrome as Klippel-Trenaunay Weber syndrome. The superficial venous channel represents the fetal limb bud vein that has failed to regress. The tissue overgrowth is mainly secondary to impaired venous return. It is usually first seen in the childhood with ratio of male: female being 1:1. The lower limb is 10-15 times more commonly affected than the upper limb. Less than 5% of the patients are affected bilaterally.

Vascular malformations associated with Klippel-Trenaunay syndrome include capillary hemangioma,² which is visible from birth. Port wine staining of the skin appears an area on the skin appearing deep purple in colour with clear demarcation from the normal skin having clear margins.² Venous malformations and varicose veins may be present. Limb enlargement may be caused by the presence of lymphoedema. Sometimes, limb abnormalities like limb hypertrophy, macrodactyly, syndactyly, polydactyly or oligodactyly may be associated with Klippel-Trenaunay syndrome.² It may present with increase in girth of the limb as the only feature with bones being less affected than the soft tissues. The discrepancy in the limb length may present as a gait abnormality with the rare presentation as atrophy rather than hypertrophy.2 It may produce significant arthropathy.⁴ Involvement of the head may cause learning difficulties and macrocephaly.

Differential diagnoses include Neurofibromatosis type 1, Proteus syndrome, Macrodystrophia lipomatosis and congenital lymphatic obstruction.

In macrodystrophia lipomatosis overgrowth of the mesenchymal elements progressively with increase in the fibroadipose tissue disproportionately causes localized gigantism.

Proteus syndrome; ² a congenital neurocutaneous disorder shows presence of lipomas, hemangiomas, hamartomas, lymphangiomas with evidence of hyperkeratosis and thickening of the soles and the palms.

Neurofibromatosis type 1¹ is a genetic disorder characterized by skin lesions on skin such as neurofibromas, cafe aulait spots, freckles in the armpit and groin and plexiform neurofibromas.

Lymphoedema which causes causing chronic swelling ulcers characterizes congenital lymphatic obstruction.² The complications include hemorrhage, chronic venous dermatitis, venous ulcer, thrombophlebitis, cellulitis, pulmonary venous gait disturbance and thromboembolism, scoliosis, Kasabach-Merritt syndrome. Kasabach-Merritt syndrome presents with decreased hemoglobin, platelets and fibrinogen levels, with increased prothrombin time and fibrin degradation products. Erectile dysfunction sometimes may be seen in men due to disturbance of venous function.⁵ The treatment of Klippel-Trenaunay syndrome is unsatisfactory. There is no curative therapy. Ligation and stripping of varicose veins often leads to severe vascular complications. Elastic compression bandage, compression garments and intermittent pneumatic compression pumps can also be used. The complications of thrombophlebitis and cellulitis can be effectively treated by elevation of the limb, with the concomitant use of steroids, analgesics and antibiotics. During pregnancy, prior to surgery and in cases of recurrent thrombophlebitis prophylactic anticoagulants and aspirin can be used.³

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