A Case of Klippel Trenaunay Syndrome and Review of Literature

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Abstract

Klippel-Trenaunay syndrome is a capillary-lymphatic-venous malformation
1. That is characterized by a triad of port-wine stain, varicose veins, and bony and soft tissue hypertrophy
2. High-flow high-shunt arteriovenous fistulae are characteristics of Parkes-Weber syndrome, a malformation that has significantly more hemodynamic complications. The cause of KTS remains obscure. Several theories exist like intrauterine injury to the sympathetic ganglia
3. deep vein abnormalities
4. mesodermal defect
5. mutated angiogenic factor gene
6. The management for the various components of this syndrome remains controversy. Here we present a 14 years old boy with KTS and discussed the management of often challenging components of this syndrome.

Case report

A 14 years old boy who was normal when born and till 7 yrs of age developed dilated tortuous vein in the left leg [Figure-1] when he started riding bicycle and playing games which gradually extended to mid thigh over this period of five years and is associated with pain (Visual analog score 8) while walking. Also complains of skin lesion [Figure-2] in the thigh and left buttok region and few lesions in the leg. Associated with abnormality of the gait because of increase in size and length of the affected limb. No consanguinous marriage among their parents.No h/o similar complaints among their family members.No h/o similar lesion present over the left gluteal region 10cm x 10cm in dimension, pink in colour, blanches on pressure. Similar lesion present in the thigh on the medial aspect on the lower one third. Venous doppler of left lower limb showed severe incompetence of the left saphenofemoral junction. The left saphenopopliteal junction and the proximal segment of the left popliteal veins not visualised and was replaced by multiple venous collaterals .Incompetent perforators are noted at the midcalf level and multiple tortuous superficial as well as intermuscular veins noted. Scanogram showed a limb length discrepancy of 2.8cm in left lower limb.[Figure-3].

Discussion

Klippel Trenaunay Syndrome is a rare cause of congenital varicose veins and is an example of capillary-lymphatic-venous malformation (CLVM). The capillary malformation (port-wine stain) is the most common cutaneous manifestation of KTS (7). This hemangioma has a distinct, linear border that respects the midline. Hemangioma is often noted on the lateral aspect of the limb. It is typically of the nevus flammeus type, but cavernous hemangiomas or lymphangiomas may also occur.

Atypical varicose veins or venous malformations may not always be evident at birth. They become more obvious as the child becomes ambulatory. Abnormalities of the superficial veins range from ectasia of small veins and varicosities to persistent embryologic veins like the lateral vein of the thigh and the sciatic vein(4). These embryologic veins frequently are large, tortuous, and dysplastic and may not contain valves. They often cause symptoms of fatigue and heaviness of the legs. Deep vein anomalies include aneurysmal dilatation, duplication, hypoplasia and aplasia, and external compression by fibrous bands or anomalous vessels(4)(5). The popliteal and superficial femoral veins are most commonly affected (4) The degree of venous hypertension in patients with outflow obstruction depends on the existent collateral channels. Hypertrophy is the most variable of the three cardinal features of KTS. In children with major limb length discrepancies serial scanograms are required to determine exact differences (to within 0.1 cm), to determine the rate of growth of the individual legs, to determine whether the difference is static or
progressing, and, finally, to determine proper timing of leg length equalization. The leg length discrepancy does not progress after physeal closure at skeletal maturity.

Careful evaluation by duplex-scanning to ensure normal anatomy of the deep vein system is important before removal of superficial dilated venous channels. Ascending and descending venography, and magnetic resonance angiography with gadolinium enhancement are useful techniques to assess the absence, presence, and extent of extremity, intra-abdominal, and pelvic involvement. In addition, MRI helps to delineate the extent of muscle and bone involvement when surgical procedures are planned.

The management of patients with KTS continues to be primarily nonoperative, but those patients with patent deep veins can be considered for excision of symptomatic varicose veins and VMs. In our patient the varicose vein is C2, E, F, A5, D, with aplasia of popliteal vein. Hence surgery is not attempted unless complication occurs where reconstruction of deep venous system can be considered. Patient managed with external compression with graduated compression stockings and garments. With six months followup pain has decreased in intensity from a VAS of 8 to 4. Since patient had osteohypertrophy scanogram was done which showed a LLD of 2.4cm. The indication for epiphysiodesis is a leg length discrepancy that exceeds 2.0 cm in the growing child. Patient was provided with heal inserts to the unaffected limb and advised to have serial scanogram every six months for proper timing of the epiphysiodesis. Laser treatment of the hemangioma can be effective in lightening the color of the port-wine stain. Currently, the flashlamp-pumped pulsed dye laser is the treatment of choice in vascular lesions. Typically, many treatments are required to achieve the desired effect.

All young patients with varicose veins should be examined thoroughly with a proper history. These rare conditions should always be kept in mind to prevent unnecessary surgeries as they are better managed conservatively. Unnecessary early surgery should be avoided as the chance of recurrence and worsening of symptoms are high. Management should be conservative and symptomatic. Because KTS is rare and the patient’s problems often complex, patients should receive multidisciplinary care.

Reference

2. Klippel M, Trénaunay P. Du noevus variqueux ostéohypertrophique. Arch Gen Med 1900;185:641-672
Illustrations

Illustration 1

Congenital varicose veins

Illustration 2

Port wine stain haemangioma
Illustration 3

Scanogram showing osteohypetrophy
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