Endovenous treatment of Klippel-Trenaunay Syndrome

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Klippel-Trenaunay syndrome (KTS), is a rare congenital malformation characterized by the triad of capillary malformations, atypical varicosities or venous malformations, and bony or soft tissue hypertrophy usually affecting one extremity. Combined with angiodysplasias, either in the form of aplasia/hypoplasia of deep veins and/or in the form of arteriovenous shunts, it is referred to as Parkes-Weber syndrome. The varicose veins will frequently be the dominant symptom, and it is emphasized that surgical treatment should not be instituted until the patient has been examined angiographically, with both phlebography and arteriography.

Servelle (Ann Surg. 1985) operated on 786 patients with Klippel and Trenaunay's syndrome and found that elongation of the impaired limb was invariably found while edema was present in 84%, varicose veins in 36%, and flat angiomata in 32%. Venography and surgical exploration demonstrated malformation of the deep veins involving the popliteal vein in 51%; superficial femoral vein, 16%; both popliteal and superficial femoral veins; 29%; iliac veins, three per cent; and lower vena cava, one per cent. Good clinical results have been achieved following the surgical release of these deep veins in the lower limb.

Gloviczki (Surgery, 1991) reported on 144 patients and found hemangioma 95%, varicosity 76%, and limb hypertrophy 93%. In 72% the disease was unilateral. Most patients did well with elastic compression only. Nine patients in the last decade underwent operation for a vascular malformation of the lower extremity. Seven had removed varicose veins or resected hemangioma of the lower extremity. Five who underwent resection of varicose veins improved. One deep venous reconstruction for atresia of the SFV, using the contralateral GSV had a patent graft at 6 months follow up. The authors noted that although patients with severe chronic venous insufficiency, disturbing cosmetic appearance, or complications of hemangioma may benefit from surgical treatment, detailed preoperative imaging of the extremity and pelvis with contrast venography is needed. Reconstruction for atresia or hypoplasia of the deep veins rarely needed.

We report a 16 year old female with KTS and bilateral lower extremity venous malformation. She had a normal deep system by venogram and duplex ultrasound (except for a duplicated left popliteal vein). There were bilateral embryonic lateral veins and facial, rectal and uterine hemangioma. She presented with pain and ulceration. In the past 2 years she has undergone endovenous laser ablation of 6 different axial veins in the office setting. This is the first case demonstrating endovenous therapy as a viable and safe alternative for a complex venous problem such as KTS.[3] Her pain improved from 9/10 to 5/10 after treatment. The lateral embryonic veins (bilateral) recanalized at 18 months. She has also undergone phlebectomy of lateral leg varicosities and ultrasound-guided sclerotherapy.

Endothermal venous ablation is applicable to more complex venous problems. Detailed work-up preoperatively is advised, and patients should be selected carefully.

References:

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