Case Reports

Right Iliac Vein Agenesis, Varicosities, and Widespread Hemangiomas

Report of a Rare Case

Ali Kutsal, MD
Thomas D. Lampros, MD
Adnan Cobanoglu, MD

We present a probable variant of the Klippel-Trénaunay syndrome with the clinical features of capillary hemangiomas, varicosities, and agenesis of the right iliac venous system, but without limb hypertrophy. To our knowledge, this is the 1st such case reported in the medical literature. ***(Tex Heart Inst J 1999;26:149-51)***

In 1900, Klippel and Trénaunay[1] described a syndrome consisting of childhood limb varicosity, nevus, and soft-tissue and bone hypertrophy of an involved limb. Since then the definition has broadened to include a deep venous anomaly with an absence of clinically important arteriovenous fistulae.[2-7] We present a rare variant of Klippel-Trénaunay syndrome (KTS) consisting of combined agenesis of the right iliac veins, varicosities, and widespread hemangiomas, without limb hypertrophy.

Case Report

A 20-year-old white man was referred to our department at Mevki Hospital with varicose veins and a 4-month history of right-lower-extremity ulcerations due to venous stasis. The patient had a red-blue discoloration over the right half of his body, as well as a dilated vein across the pubic symphysis, present since early childhood, which had slowly been increasing in size.

Clinical Examination. On physical examination, we found that the patient's entire right leg, arm, trunk, and neck were covered with large capillary hemangiomas. Capillary hemangiomas also covered his left chest, back, groin, thigh, and foot (Fig. 1). Three new venous stasis ulcers and varicose veins were present on the inner right thigh. In addition, a large suprapubic vein, present since early childhood, was increasing in size and now measured 2 cm in diameter with blood flow from right to left. The distal pulses were intact, and no lower-extremity limb edema or hypertrophy was identified.

Diagnostic Procedures. Bilateral lower-extremity and percutaneous femoral venography were performed. The venous system in each leg was normal; however, the right iliac venous system was absent. Angiography showed a dilated suprapubic vein that crossed the pubic symphysis, with contrast medium passing from the right common femoral vein to the left common femoral vein (Fig. 2). The left iliac venous system and the inferior vena cava were enlarged due to the development of collateral vessels. Aortography revealed normal anatomy without an arteriovenous fistula. Plain films of the abdomen, pelvis, and lower extremities also yielded results within normal limits. There was no limb hypertrophy.

Discussion

Although many authors[8-10] have reported variations in the anatomic development of the inferior vena cava, few reports have described similar abnormalities of the iliac veins—especially agenesis.[11,12] Since the causes of such abnormalities are not clear, we present a discussion of the embryogenesis of the inferior vena cava and the iliac veins as an aid to understanding.

Key words: *Hemangioma; hypertrophy/etiology; iliac vein/abnormalities; Klippel-Trénaunay-Weber syndrome/physiopathology; leg/blood supply; neovascularization, pathologic; varicose veins/pathology*

From: *The Department of Cardiothoracic Surgery, Mevki Hospital, Ankara, Turkey; and The Division of Cardiopulmonary Surgery, Oregon Health Sciences University, Portland, Oregon 97201*

Address for reprints: *Adnan Cobanoglu, MD, Professor and Chief, Division of Cardiopulmonary Surgery, Oregon Health Sciences University, 3181 S.W. Sam Jackson Park Road, L353, Portland, OR 97201-3098*

Early in embryogenesis, the cardinal veins form the main venous drainage system of the human embryo. The anterior cardinal veins drain the cephalic portion of the embryo, and the posterior cardinal veins drain the remaining portion of the embryo.

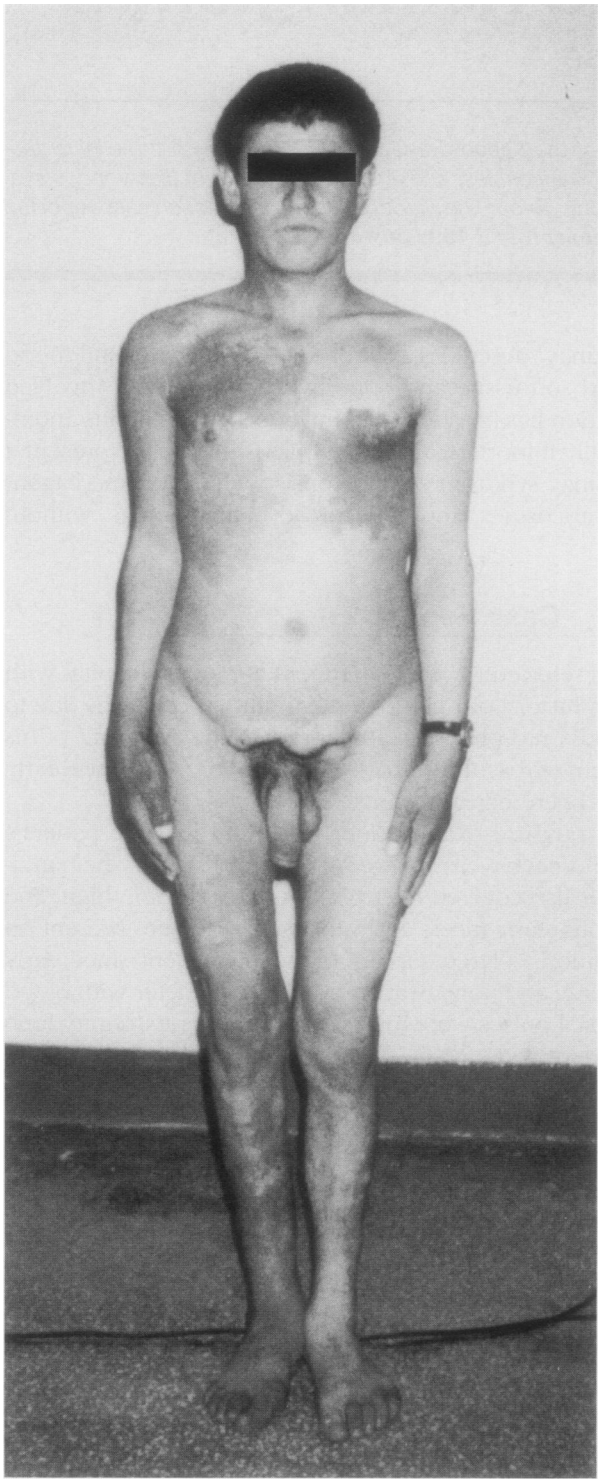

Fig. 1 *Photograph shows capillary hemangioma covering approximately 60% of the patient's body. In addition, there is a dilated suprapubic vein.*

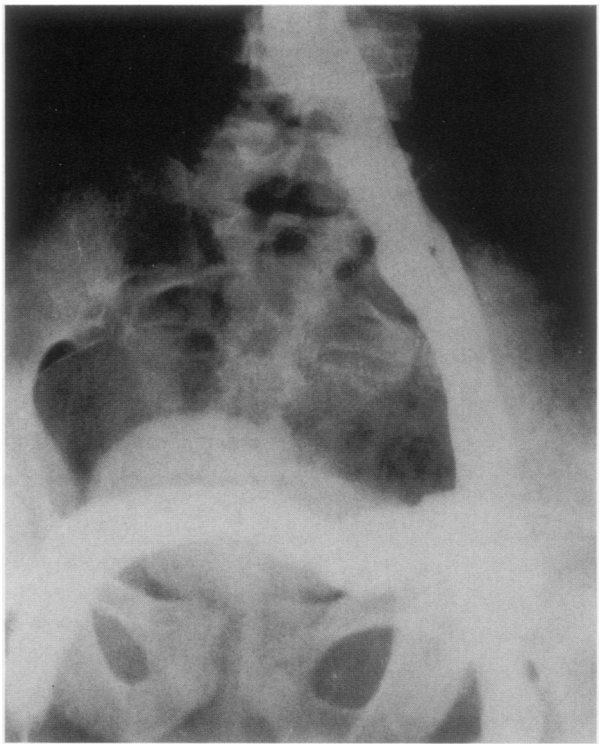

Fig. 2 *Angiogram shows the dilated suprapubic vein crossing the symphysis pubis in the right-to-left direction.*

During weeks 5 to 7, the sacrocardinal veins provide drainage of the right and left lower extremities. The vena caval system arises from an anastomosis between the left and right sides of the cardinal system in such a manner that the blood from the left side is channeled to the right side. The right sacrocardinal vein finally becomes the sacrocardinal segment of the inferior vena cava, and the anastomosis between the sacrocardinal veins becomes the left common iliac vein.[13] A failure in the development of these veins or the anastomosis may account for the agenesis of the right iliac venous system, as seen in our patient.

The original description of KTS in 1900 consisted of limb varicosity, hemangioma, and hypertrophy.[1] Although the exact origin of KTS remains unclear, some authors[14,15] have hypothesized that this clinical triad is the result of chronic venous hypertension produced by a deep venous anomaly. Others[16] have hypothesized that a mesodermal abnormality during fetal development is the primary cause. Involvement is usually unilateral but may include a 2nd lower or upper extremity.[3] Trunk involvement has also been described.[17]

Varicosities may develop secondary to a deep venous abnormality of the involved limb,[14-15] which might account for the common distribution of varicose veins along the lateral aspect of the thigh.[2-4] Most varicosities occur early in infancy or childhood[1]

and may be the major source of venous outflow to the involved extremity.[6] Varicosities are almost always visible on a venogram but are clinically apparent in only 39% to 79% of patients, due to substantial amounts of edema in the involved extremity.[2-7] Moreover, surgical resection of varicosities without identification of an associated deep venous abnormality may aggravate symptoms.[6] These points reinforce the need for venography before surgical intervention.

Hemangiomas are vascular tumors with large numbers of mast cells and a high rate of endothelial cell turnover.[18] In cases of KTS, hemangiomas are typically located on the involved extremity[2-7] but may extend to the trunk,[2,12,17] to other extremities,[2] or both. Capillary hemangiomas with telangiectasis of the superficial dermal vessels are the most common.[19]

Limb hypertrophy is present in 87.5% to 100% of patients diagnosed with KTS.[2-5] At present, it remains unclear whether the above-normal skeletal growth is caused by venous hyperemia due to a deep venous anomaly,[14,15] or if it is caused by an inborn error in tissue development.[16] Although microscopic arteriovenous connections may be present with KTS, they are clinically inconsequential—unlike those in Parkes-Weber syndrome.[9]

A deep venous anomaly is most often associated with the popliteal vein.[5,20] Agenesis and hypoplasia of the iliac vein has been reported,[2,4,5,12] but its relative frequency remains low. Our patient's clinical diagnosis differed from typical KTS, primarily because of a lack of limb hypertrophy. This lack has previously been reported in the literature in association with KTS. To our knowledge, however, this is the 1st report in the medical literature of a large capillary hemangioma, varicosities, and agenesis of the right iliac venous system, all in the absence of limb hypertrophy.

References

1. Klippel M, Trénaunay P. Du naevus variquex osteo hypertrophique. [French] Arch Gen Med (Paris) 1900;3:641-72.
2. Baskerville PA, Ackroyd JS, Lea Thomas M, Browse NL. The Klippel-Trénaunay syndrome: clinical, radiological and haemodynamic features and management. Br J Surg 1985;72: 232-6.
3. Gloviczki P, Hollier LH, Telander RL, Kaufman B, Bianco AJ, Stickler GB. Surgical implications of Klippel-Trénaunay syndrome. Ann Surg 1983;197:353-62.
4. Gloviczki P, Stanson AW, Stickler GB, Johnson CM, Toomey BJ, Meland NB, et al. Klippel-Trénaunay syndrome: the risks and benefits of vascular interventions. Surgery 1991;110:469-79.
5. Servelle M. Klippel and Trénaunay's syndrome: 768 operated cases. Ann Surg 1985;201:365-73.
6. Lindenauer SM. The Klippel-Trenaunay syndrome: varicosity, hypertrophy and hemangioma with no arteriovenous fistula. Ann Surg 1965;162:303-14.
7. Weber FP. Haemangiectatic hypertrophy of limbs—congenital phlebarteriectasis and so-called congenital varicose veins. Br J Child Dis 1918;15:13-7.
8. Edwards EA. Clinical anatomy of lesser variations of the inferior vena cava; and a proposal for classifying the anomalies of this vessel. Angiology 1951;2:85-99.
9. Effler DB, Greer AE, Sifer EC. Anomaly of the vena cava inferior. Report of a fatality after ligation. JAMA 1978;146:1321-2. [This reference is in error and could not be corrected by press time.]
10. Katona I, Vadasz Y. Rare developmental anomaly of inferior vena cava in adult. Acta Morphol 1951;1:281-7.
11. Lotz PR, Seeger JF. Normal variations in iliac venous anatomy. AJR Am J Roentgenol 1982;138:735-8.
12. Thomas ML, Posniak HV. Agenesis of the iliac veins. J Cardiovasc Surg (Torino) 1984;25:64-6.
13. Nicholson CP, Gloviczki P. Embryology and development of the vascular system. In: White RA, Hollier LH, editors. Vascular surgery: basic science and clinical correlations. Philadelphia: JB Lippincott Company, 1994:6-9.
14. Servelle M, Zolotas E, Soulié J, Andrieux J, Cornu C. Syndrome de Klippel et Trénaunay. Malformations des veines iliaques, fémorale et poplitée. [French] Arch Mal Coeur Vaiss 1965;58:1187-97.
15. Van der Molen HR. Maladie de Klippel-Trénaunay et grosses jambes. [French] Societe Francaise Phlebologie 1968;2:187-97.
16. Baskerville PA, Ackroyd JS, Browse NL. The etiology of the Klippel-Trénaunay syndrome. Ann Surg 1985;202:624-7.
17. Telander RL, Kaufman BH, Gloviczki P, Stickler GB, Hollier LH. Prognosis and management of lesions of the trunk in children with Klippel-Trénaunay syndrome. J Pediatr Surg 1984;19:417-22.
18. Mulliken JB, Zetter BR, Folkman J. In vitro characteristics of endothelium from hemangiomas and vascular malformations. Surgery 1982;92:348-53.
19. Pack GT, Miller TR. Hemangiomas. Classification, diagnosis and treatment. Angiology 1950;1:405-26.
20. Taheri SA, Williams J, Boman L, Pisano S. Superficial femoral vein transposition in Klippel-Trénaunay syndrome. J Pediatr Surg 1989;24:494-6.