PERSISTENCE OF THE EMBRYONIC LATERAL MARGINAL VEIN: REPORT OF TWO CASES

Raúl Rojas Martinez, Pedro Puech-Leão, Paulo Motta Guimarães and Baptista Muraco Netto

RHCFAP/3054

ROJAS MARTINEZ R et al. - Persistence of the embryonic lateral marginal vein: report of two cases. **Rev. Hosp. Clín. Fac. Med. S. Paulo 56**(5):159-162, 2001.

Purpose: Congenital venous malformations of the lower limbs represent a particular challenge for the vascular surgeon. Persistence of fetal veins is a rare malformation, and the most common is the persistence of the lateral marginal vein usually observed in patients with Klippel-Trenaunnay Syndrome. The persistence of this embryonic vein as an isolated venous malformation without the other characteristics of the Klippel-Trenaunnay Syndrome has not yet been reported. This paper describes two cases.

Methods: Two patients, a 17-year-old male patient and a 16-year-old female, have had since their birth a large venous trunk in the lateral aspect of the right leg and thigh. The limbs underwent duplex scanning and phlebography. The surgical removal of the lateral marginal vein was performed.

Results: Surgical treatment resulted in very good functional and aesthetic results. Follow-up at 26 months showed no evidence of varicose vein recurrence.

Conclusions: To achieve good results, surgical intervention may be indicated in cases of orthopedic deformity, hemorrhage, symptomatic, and unaesthetic lesions.

DESCRIPTORS: **Klippel-Trenaunay Syndrome. Marginal vein. Congenital venous malformations. Angiodysplasias. Varicosities.**

Based on their anatomic and pathologic structure, congenital vascular malformations can be divided into 5 groups: capillary, arterial or venous when they have only one component, arteriovenous malformations that have arteriovenous fistulas, and complex lesions when different combinations of the previous 4 types occur. Congenital venous malformations of the lower limbs include: valvular agenesis, extrinsic constriction, hypoplasia, atresia or aplasia of veins, persistence of fetal veins, hypertrophy, tortuousities, and varicosities[1-3]. Clinical findings of venous malformations may vary significantly, from superficial lesions such as varicosities or circumscribed lesions to deep lesions that involve muscle and bone and can cause limb hypertrophy and deformity. Venous anomaly may occur as an isolated lesion, or is most often associated with complex vascular malformations like the Klippel-Trenaunnay Syndrome (KTS). It is important to distinguish persistence of fetal veins from primary varicosities and from varicosities secondary to arteriovenous fistula or deep venous thrombosis[3,4].

Persistence of fetal veins is a rare congenital malformation. The most common is the persistence of the lateral marginal vein, which is usually associated with complex congenital venous malformations such as KTS[1,4]. Its presence as an isolated lesion has not been previously reported. This article presents two cases of persistent embryonic lateral marginal vein as an isolated venous malformation without being part of KTS, treated surgically. Literature review did not reveal any citation of similar cases.

From the Department of Vascular Surgery, Hospital das Clínicas, Faculty of Medicine, University of São Paulo.

CASES REPORT

We treated two patients: one 17-year-old white man and one 16-year-old

white woman, complaining of varicosities, pain, and heaviness on the right lower limb. They had no history of venous thrombosis, and neither family had varicosity or venous malformations.

Physical examination revealed a large venous trunk existing since birth in the lateral aspect of the right lower limb from the ankle to the upper thigh. In the lateral aspect of the upper leg below the knee, the marginal vein had developed substantial collateral varicosity. One patient presented a large incompetent perforator vein that developed a venous aneurysm below the knee (Fig. 1). Limb pulse was palpable and normal with no audible bruit or palpable thrill.

Venous duplex scanning of the limbs showed a competent and normal deep venous system with competent saphenofemoral and saphenopopliteal junctions. Phlebography demonstrated incompetence of the abnormal lateral vein communicating to the common femoral vein, and to incompetent perforators veins below the knee. Reflux of saphenous junctions and perforator veins communicating to the saphenous system was not detected. The deep venous system of the extremity was phlebographically normal (Figs. 2 and 3).

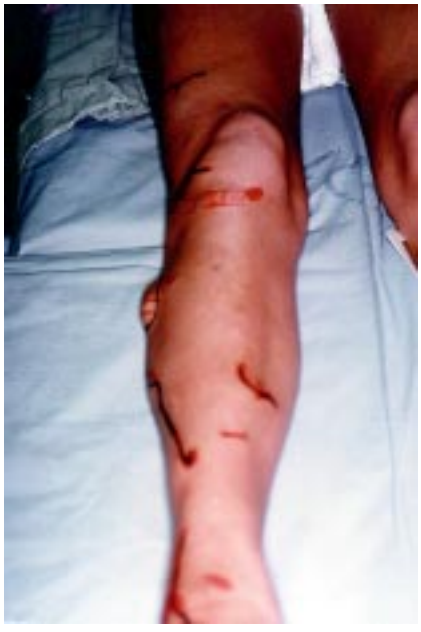
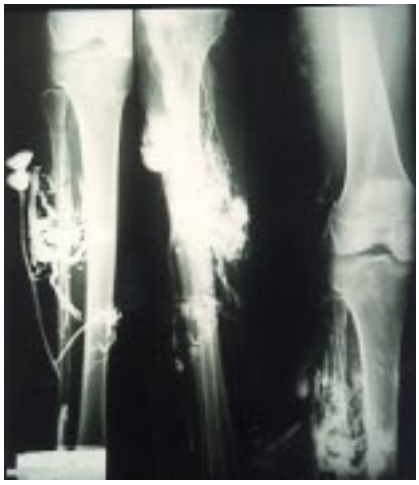
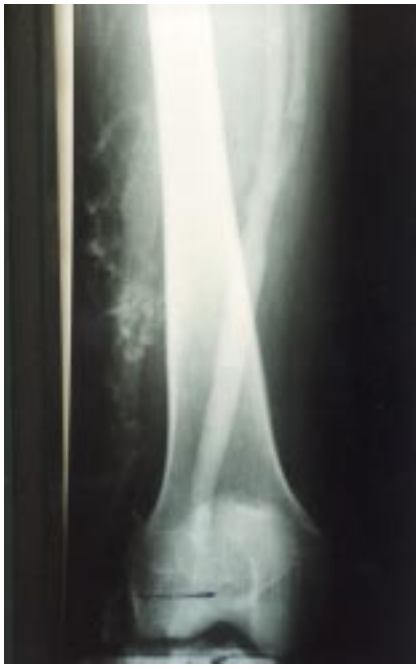


Fig. 1 - Preoperative design on the skin of persistent of the lateral marginal vein showing the typical feature of a large lateral venous channel.

Surgical removal of the lateral marginal vein and its tributaries was performed through small transverse incisions in the skin and suture of the perforating veins behind the fascia without the use of the vein stripper. The ends of the lateral marginal vein in the ankle and thigh were tied (Fig. 4). Except for the absence of the valves, the removed vein was macroscopically similar to primary varicosity, with a dilated and thick wall.





Figs. 2 and 3 - Phebography showing a large lateral vein marginal arising from varicosities within the ankle and running up the lateral aspect of the leg and thigh. The presence of perforator veins and normal deep venous system allows surgical removal of the vein.

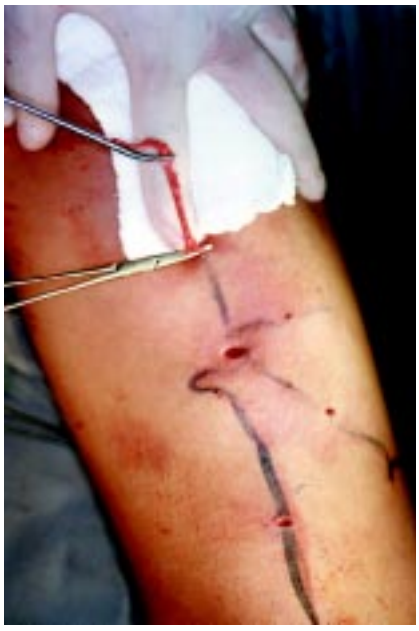


Fig. 4 - Surgical removal of the lateral marginal vein through small excisions in the skin with suture of perforating veins.

Surgical removal of the lateral marginal vein was performed with very good functional and aesthetic results. The patients recovered well and were discharged the day after surgery with elastic stockings and physiotherapy exercises. They returned to normal activities two weeks after surgery. The mean duration of follow-up was 26 months. There was no clinical or physical evidence of recurrence of varicose veins; the saphenous vein system remains normal.

DISCUSSION

In 1900, the French physicians Klippel and Trenaunnay described a clinical syndrome consisting of congenital varicose veins, cutaneous hemangiomas, and hypertrophy of the same extremity[6]. The Klippel-Trenaunnay Syndrome (KTS) is the most common complex congenital venous malformation. Deep vein apla-

sia or hypoplasia and a persistent valveless lateral channel are frequently involved. The precise etiology of congenital venous malformations is unknown. The mechanism and time of embryonic damage are uncertain, and no hereditary factors have been confirmed. De Takats and other authors have suggested that the anatomic-pathologic changes seen in angiodysplasias can be explained by aberrations in vasculogenesis occurring in specific stages of embryonic development[1,2,7,8].

The congenital vascular anomalies could be comprised essentially of just one type of vessel anomaly: capillary, arterial, or venous anomaly. In evaluating the venous dysplasias, we have to separate two basic features: phleboangioma and phlebectasia. Phleboangioma is essentially a venous hamartoma with varying amounts of capillary and cavernous elements. These lesions are usually found in the subcutaneous tissue and muscles and can produce symptoms of heaviness or pain. Phlebectasia, in contrast to phleboangioma, is a congenital dilatation of a large, superficial venous trunk that dilates progressively over time. Usually, the muscular and deep veins are not dilated, but may be hypoplastic or aplastic. It is especially noted that in this disorder, the greater saphenous vein is often not involved in these congenital varicosities[2,3]. More often, the varicosity is a lateral vein running from the ankle up across the knee and thigh toward the greater trochanter and then into the inguinal or gluteal area. This phlebectasia is related to the congenital persistence of the lateral marginal vein, also called a "lateral venous anomaly" by Dodd and Cockett or a "lateral marginal vein of Servelle"[5,9]. This relationship suggests the persistence of the embryonic dorsal or sciatic vein system that normally should have involuted around the tenth to twelfth week of intrauterine life[2,3,12]. In many cases, the lateral marginal vein develops incompetent collateral and perforator veins that form a large varicose mass. This persistent vein, usually thick-walled, may or may not be valveless, and is not always visible. The termination of the lateral vein is variable, may be single or multiple, and is best determined by phlebography. Common sites of termination are the common or profunda femoral vein, the external or internal iliac vein, popliteal vein, greater saphenous vein, and inferior vena cava[1]. Baskerville et al. performed phlebography in 36 cases of KTS and demonstrated that the lateral marginal vein drained into the popliteal vein (11%), superficial femoral vein (17%), deep femoral vein (20%), external iliac vein (5%), greater saphenous vein (14%), and the internal iliac artery through the gluteal veins (33%)[10]. Our two patients showed a large varicose trunk in the lateral aspect of the limb that began in the ankle and ran up to the upper thigh with a varicose mass and incompetent perforators below the knee without communication with the greater saphenous vein. In the two cases, the vein drained into the common femoral vein.

This persistent vein is often associated with hypoplasia or aplasia of some parts of the deep venous system of the extremity, and its presence should always alert to this possibility; careful phlebography should be carried out before surgical removal of the lateral marginal vein is considered[1,3,5,11,12]. The persistent vein can occur alone, or more often, is associated with KTS or with other angiodysplasias, such as the Servelle-Martorell syndrome or the Kasabach-Merrit Syndrome[1,3,5,10,11]. There was no history suggestive of genetic transmission. The most common concomitant congenital anomalies were digital agenesis, congenital hip dislocation, coxa vara, atresia of the ear canal, spina bifida, syndactylia, and clinodactylia[3,10-12]. The two patients underwent duplex scanning and phlebography that revealed a normal and competent deep venous system. Neither presented skin nevus, limb hypertrophy, or another congenital anomaly.

Servelle demonstrated on 768 patients with KTS, who had undergone surgery the presence of varicose veins in 36%, the majority with persistence of the lateral marginal vein[5]. Gloviczki et al. demonstrated in 40 patients with KTS varicosities in 25 cases, with persistent lateral marginal vein in 7 cases[12]. Baskerville et al. reported that in 49 patients with KTS, the lateral vein was present in 68%, with incompetent perforator veins below the knee in 45% of the cases[10]. In our institution, in a series of 180 patients with KTS, 17 patients (9%) had persistence of the lateral marginal vein (not published).

Symptoms derived from the persistent lateral vein may be related to the resulting orthopedic deformity or to the changes due to venous stasis such as edema, pain, eczema, dermatitis, induration of the skin and subcutaneous tissue, ulceration, and thrombophlebitis. Phleboliths can develop from repeated episodes of local thrombosis[3]. In a study of 40 cases of KTS, the lateral marginal vein presented with hemorrhage in 22% and thrombophlebitis in 16%[10]. Our two patients complained of "burning" pain and heaviness on the right lower limb that was prominent with orthostatism. The persistent vein was prominent, painful, and dilated progressively over time.

The diagnosis is essentially clinical and obvious on inspection, represented by a large venous channel presented since birth on the lateral aspect of the extremity. The most frequent error that is made in diagnosis is assuming that a large dilated venous mass represents an arteriovenous malformation. Differentiation can often be made by clinical examination, such as auscultation of a bruit or by Nicoladoni-Branham's test. Other investigations include duplex scanning and arteriography. Phlebography is mandatory before the surgical

removal of the lateral marginal vein is considered. Phlebography identifies anomalies of the deep vein system, and it helps to localize incompetent perforating veins. If aplasia or hypoplasia of the deep venous system is present, an extensive varicectomy should not be performed because these varicose veins serve as substitute channels for the obstructed deep vein[1,3,10,12].

The persistence of the embryonic lateral marginal vein may occur as an isolated venous malformation without association with other congenital venous syndromes and is very rare. The best method of treatment in these cases depends on the extent of the lesion, the severity of the symptoms, and the clinical condition of the patient. We suggest surgical intervention in cases of orthopedic deformity, hemorrhage, symptomatic, or unaesthetic lesions.

RESUMO

RHCFAP/3054

ROJAS MARTINEZ R e col. - Persistência da veia marginal do embrião: relato de dois casos. **Rev. Hosp. Clín. Fac. Med. S. Paulo 56**(5):159-162, 2001.

Objetivo: As anomalias venosas congênitas representam um desafio especial para o cirurgião vascular. A persistência de veias fetais é uma malformação rara, sendo o exemplo mais comum a persistência da veia marginal embrionária na face lateral do membro inferior em pacientes com hemangiodisplasias complexas, particularmente a Síndrome de Klippel-Trenaunnay. Seu aparecimento como defeito vascular congênito único é raríssimo. Mostramos 2 destes casos.

Método: Dois pacientes, com 17 e 16 anos sendo um do sexo masculino, apresentavam cordão varicoso, único e exuberante na face lateral da coxa e perna direita, presentes desde o nascimento. Os membros foram avaliados com mapeamento Duplex e flebografia. Foi realizado tratamento cirúrgico com ressecção total da veia marginal.

Resultado: Ambos pacientes evoluíram satisfatoriamente. O seguimento médio foi de 26 meses, sem evidência de recorrência das varizes.

Conclusão: A terapêutica nestes casos depende do estado clínico e extensão da lesão. As indicações para o tratamento cirúrgico incluem: sangramento, lesões tróficas, dor, edema e deformidade estética, obtendo-se bons resultados.

DESCRITORES: **Síndrome de Klippel-Trenaunnay. Veia Marginal. Malformação venosa congênita. Hemangiodisplasia . Varizes.**

REFERENCES

1. MULLIKEN J & YOUNG A - Combined Vascular Malformations. In: MULLIKEN J. **Vascular Birthmarks: Hemangiomas and Malformations**. Philadelphia, Saunders, 1988. p. 159, 256-58.

2. MALAN & PUGLIONISI A - Congenital angiodysplasias of the extremities. In: Generalities and classification, venous dysplasias. **J Cardiovasc Surg,** 1964; **5**:87-130.

3. HOLLIER L, BERGAN J & YAO JST - Chapter 21: Surgical treatment of congenital venous malformations. In: BERGAN J. **Surgery of the veins**. Orlando, Grune, 1985. p. 275-283.

4. MYERS T & JANES J - Comprehensive surgical treatment of cavernous hemangiomas of the lower extremity. **Surgery,** 1955; **27**(2): 184-197.

5. SERVELLE M - Klippel and Trénaunay's Syndrome. **Ann Surg,** 1985; **201**(3): 365-372.

6. KLIPPEL M & TRENAUNAY P - Du naevus variquex osteohypertrophique. **Arch Gen Med** (Paris) 1900; **3**:641-672.

7. DETAKATS G - Vascular anomalies of the extremities: Report of 5 cases. **Surg Gynecol Obstet** 1932; **55**:227-237.

8. VOLLMAR J - Angiodysplasie und skeletsystem. **Chirurg,** 1976; **47**(4): 205-213.

9. DODD H & COCKETT FB - **The Pathology and Surgery of the Veins of the Lower Limbs**. 2nd. ed. New York, Churchill, 1976. p. 187-190.

10. BASKERVILLE PA et al. - The Klippel-Trenaunay Syndrome: clinical, radiological and hemodynamic features and management. **Br J Surg** 1985;**72**: 232-236.

11. VOLLMAR J & VOSS EU - Vena marginalis lateralis persistens - die vergessene vene der angiologeu. **VASA** 1979; **8**: 192-202.

12. GLOVICZKI P & HOLLIER LH - Surgical implications of Klippel-Trenaunay Syndrome. **Ann Surg** 1983; **197**(3): 353- 362.

Received for publication on April 20, 2001.