

# A Rare Lateral Embryonic Varicose Vein Surgery in Klippel Trenauny Syndrome with Brief Review of the Literature

Fatih Ada<sup>1</sup>, Arif Aydın<sup>2</sup>, Sadık Volkan Emren<sup>3</sup>, Ersin Çelik<sup>1</sup>

## ABSTRACT

**Introduction:** Varicose vein surgery is usually performed great saphenous vein and small saphenous vein, which is rarely performed lateral embryonic vein and other vascular malformations. Klippel trenauny syndrome includes; varicose vein, port-wine skin pigmentation and limb hypertrophy triad. Arteriovenous fistula had added next time in this syndrome and it was called klippel trenauny weber syndrome.

**Case report:** We present a male adolescent patient varicose vein surgery with klippel trenauny syndrome and review of the literature.

**Conclusion:** Preoperative physical examination and diagnostic tests are very important for intraoperative approach in Klippel Trenauny syndorme. Symptomatic treatment should be given presence of deep venous aplasia or insufficiency.

**Keywords:** Klippel-Trenaunay Syndrome, Varicose Veins, Venous Malformation

## INTRODUCTION

Klippel Trenaunay syndrome (KTS) is a rare congenital anomaly characterized by capillary malformations, varicosities, soft tissue and bone hypertrophy.<sup>1</sup> KTS was described by Klippel Trenaunay in 1900 and which was called 'naevus vasculosus osteohipertrophicus'. Etiopathogenesis is not elucidated fully. There is no gender difference in the prevalence of disease and which occur at birth or in early infancy. Deep venous thrombosis and pulmonary embolism can occur in this syndrome therefore early diagnosis and treatment very important. We present a male adolescent patient varicose vein surgery with klippel trenauny syndrome and review of the literature.

## CASE REPORT

A 15 years old adolescent male patient had admitted the our clinic with left leg pain and swelling. Varicose veins at lateral side of left leg, port wine stain at anteromedial side of left leg and left food hypertrophy were not seen in examination (Figure 1). Duplex ultrasonography showed the varicose lateral embryogenic vein (LEV) and no insufficiency at saphenofemoral junction and deep venous system. Deep venous system was completely normal, there was no aplasia or hypoplasia. LEV's connection was found and marked with duplex ultrasoundography preoperatively. After written informed consent obtained from the patient, preoperative preparation was completed. Under the spinal anesthesia, groin incision was performed 4-5 cm below the inguinal ligamentum. Then LEV and saphenofemoral junction connection were found. LEV was connected the greater saphenous vein just 2-3 cm under the saphenofemoral junction and which was ligated and removed by a stripper in two stage (Figure 2). Mini phlebectomies were performed the other small varicose veins.

Pathological samples were taken on excrescence tissue at the left foot dorsum. Patient was discharged completely healthy with compression stockings on the second day after his admission. The patient has not any complain 3 months later the operation. There was no varicose veins and swelling on left leg and duplex ultrasonography was completely normal (Figure 3).

## DISCUSSION

Klippel trenauny syndrome is a rare vascular anomaly disease. KTS typical triad are cutaneous capillary malformation of a limb, venous or lymphatic malformations, and disturbed growth of soft tissue or bone.<sup>1</sup> Typical skin lesions are similarly to the port wine fleck cutaneous hemangioma. This cutaneous hemangioma separated by a precise line of demarcation from the normal skin. Typical port wine skin pigmentation was at anteromedial side of left leg in our case. Hemihypertrophy holds unilateral extremity usually.<sup>2</sup> There was not observe hemihypertrophy in our case but an excrescence tissue was observed at the left foot dorsum. KTS diagnosis is defined clinically. The etiology of the disease is not known clearly. However, LEV is known as a residue embryological vein tissue.<sup>3</sup> Many different names are used to describe for LEV in the literature such as lateral marginal vein, persistent sciatic vein, lateral venous anomaly, forgotten vein of angiologists, vein of servelle, klippel-trenaunay vein, marginal vein, embryonic vein.<sup>4</sup> LEV starts from the foot dorsolateral and tends as tortuous to the cranial and the drainage often continue to lateral branches of deep femoral vein or the internal iliac vein. LEV is seen %68-80 in KTS patient and which is function as a collateral brunch in deep venous system aplasia. The absence of valves causes venous reflux and venous hypertension ultimately it causes varicosities.<sup>5-7</sup> Baskerville et al. reported LEV drained into external iliac vein (%5), popliteal vein (%11), greater saphenous vein (%14), superficial femoral vein (%17), deep femoral vein (%20), gluteal veins (%33) in phlebography. Venous system aplasia or hypoplasia can occur in KTS. Venous system aplasia or hypoplasia ratio found %18 in same study and Rendo et al. found %22 in KTS.<sup>8,9</sup> Our patient also had a large LEV at lateral side of left leg which was began cruris to continue

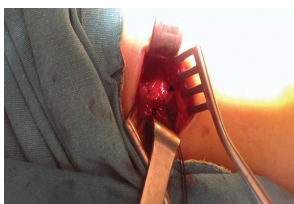
<sup>1</sup>Department of Cardiovascular Surgery, <sup>2</sup>Department of Plastic, Reconstructive and Aesthetic Surgery, <sup>3</sup>Department of Cardiology, Afyonkarahisar State Hospital, Afyonkarahisar, Turkey

**Corresponding author:** Fatih Ada, Orhangazi Mah. Nedim Helvacioğlu Bulvarı No:73 Afyonkarahi.

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**Figure-1:** Preoperative appearance of port wine skin pigmentation, lateral embryonic vein and varicose veins; **Figure-3:** Postoperative appearance, 3 months later the operation.



**Figure-2:** A: Lateral embryonic vein B: Greater saphenous vein

thigh. There were a lot of varicose veins drained to described vein. Duplex ultrasonography showed, LEV was drained to greater saphenous vein. International society for the study of vascular anomalies (ISSVA) was classified the KTS, as a vascular malformations associated with other anomalies (Capillary malformations+ Venous malformations+/- Lymphatic malformations+ Limb overgrowth).<sup>10</sup> Additionally, megalencephaly on craniofacial region, mental retardation, polysyndactyly on the fingers can occur in this syndrome. Some complications such as, hypercoagulability, thrombosis and pulmonary embolism may develop. Incidence of pulmonary embolism is between 14-22% in KTS.<sup>11</sup> Bone, fat and muscle hypertrophy can be demonstrated on computed tomography and magnetic resonance imaging. Computed tomography angiography and magnetic resonance angiography can be displayed venous system along the extremity simultaneously. Especially, deep venous system and collaterals should be evaluated for surgery or percutaneous treatment planned patients. In our case, LEV was drained to the greater saphenous vein and deep venous system was completely normal on duplex ultrasonography so that we did not need an advanced study. Main treatment include of compression stocking and venoactive agents in KTS. Surgery performed unusually in this syndrome because of deep venous insufficiency or hypoplasia/aplasia (Compleat or incompleat) were seen usually. Deep venous system aplasia is the absolute contraindication for the surgery. Soker et al. reported a case popliteal vein aplasia with KTS patient who was treated with compression stockings and venoactive drugs for the symptoms.<sup>7</sup> Only severe symptomatic patients were operated in Malgor et al studies.<sup>12</sup> In the normal deep venous system, LEV's complete resection is the most effective method. We operated the patient for the left leg pain and swelling complaint. LEV was ligated and removed by a stripper in the operation. Pathological specimens were reported as a normal

muscle tissue by the pathologist.

## CONCLUSION

Duplex ultrasonography, MR angiography and CT angiography tests very important before the surgery for evaluate the deep venous system. LEV should not be removed when the presence of deep venous system aplasia owing to it provide the collateral venous return. Symptomatic treatment should be kept in mind with mild symptomatic patients.

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