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Case report
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Klippel-Trenaunay syndrome in a patient presenting with venous stasis ulcer

Venöz staz ülseri olan bir hastada Klippel Trenaunay sendromu

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Abstract

Klippel-Trenaunay syndrome is a congenital capillary-venous vascular malformation characterized by malformation of the capillaries, soft tissue and bone hypertrophy and atypical varicosities. Klippel-Trenaunay syndrome often affects the lower extremity but can also involve arms, trunk, head, neck, abdominal and pelvic organs, and central nervous system. Deep venous system anomalies are important in directing the treatment of these cases. In cases of deep vein hypoplasia, marginal vein can be resected since hypoplastic deep veins can spontaneously dilate to almost normal dimensions after marginal vein resection. In cases of deep venous aplasia, the embryonal vein can be the main drainage vein of the leg and the resection is unlikely. In this article, we present a case of a 20-year-old male with Klippel-Trenaunay syndrome who had right leg diameter increase, port-wine stain appearance, and venous ulcer.
Keywords: Klippel-Trenaunay syndrome, Port-wine stain, Venous ulcer, Limb hypertrophy

Öz

Klippel-Trenaunay sendromu kapiller malformasyon, yumuşak doku ve kemik hipertrofisi ve atipik varikoziteler ile karakterize edilen konjenital bir kapiler venöz vasküler malformasyondur. Klippel-Trenaunay sendromu genellikle alt ekstremiteyi etkiler; ancak kollar, gövde, baş, boyun, abdominal ve pelvik organlar ve santral sinir sistemini de etkileyebilir. Derin venöz sistem anomalileri, bu vakaların tedavisinin yönlendirilmesinde önemlidir. Derin ven hipoplazisi olan olgularda marjinal ven rezeke edilebilir çünkü rezeksiyondan sonra hipoplastik derin venler kendiliğinden dilate olurlar. Derin venöz aplazisi olan olgularda ise embriyonal ven bacağın ana drenaj veni olabilir ve rezeksiyonu olası değildir. Bu yazıda, sağ bacak çapında artış, port-wine lekesi görünümü ve venöz ülseri bulunan, Klippel-Trenaunay sendromu olan 20 yaşında erkek olgu sunulmuştur.
Anahtar kelimeler: Klippel-Trenaunay sendromu, Şarap lekesi görünümü, Venöz ülser, Bacak hipertrofisi

Introduction

Klippel-Trenaunay syndrome (KTS) was described as a syndrome characterized by limbic capillary nevi, leg hypertrophy and varicose veins [1]. With the addition of significant arteriovenous fistulas, Klippel-Trenaunay-Weber syndrome was also described later [2]. KTS often affects the lower extremity [1] but also can involve arms, trunk, head, neck [3], abdominal and pelvic organs [4,5] and central nervous system [6]. The ethio-pathogenesis of KTS has not been fully elucidated. Although it is mostly seen as sporadic, it has been reported to be inherited, autosomal dominant and originating from the genetic defects [7, 8]. Syndrome equally affects men and women [9]. Treatment of KTS is usually conservative, but surgical procedures for complications are required [10,11].

Case presentation

A 20-year-old male patient was referred to our clinic due to recurrent venous ulceration and swollen right leg. Clinical examination further revealed port wine stain of the right lower extremity covering an area from right foot to hip. The right ankle had a 1 cm venous ulcer scar in the lateral upper part (Figure 1). Both the port wine stain and increased right leg diameter were present since childhood (Figure 2). There was no functional loss. There was no clinical evidence of AV macrofistula.

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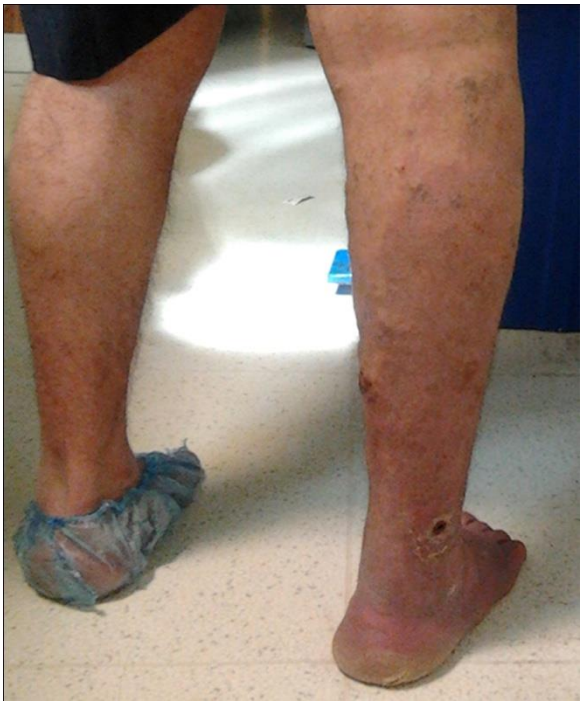

Figure 1: Venous ulcer in the lateral upper part of the right ankle of the patient

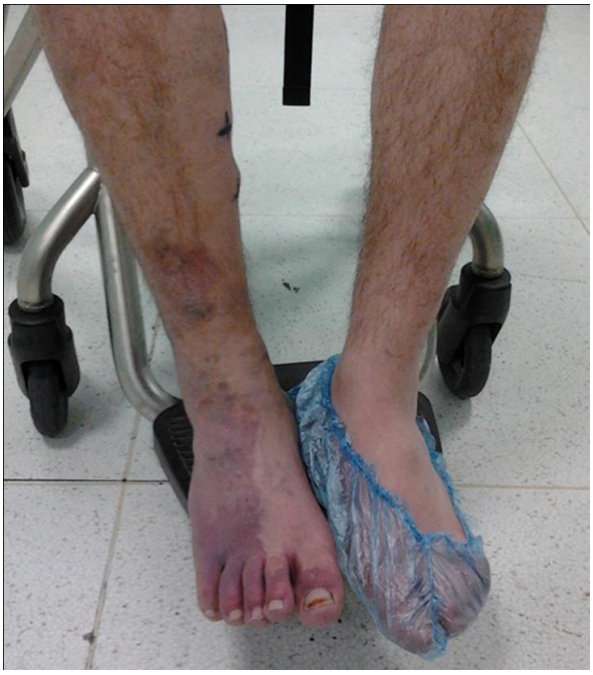

Figure 2: Port-wine stain and right limb hypertrophy of right leg of the patient

Doppler ultrasonography (USG) showed perforating venules 7.2 mm and 6.2 mm in diameter and dilated superficial varicose veins associated with those were observed at the Cockett levels 2 and 3 with reflux flow with proximal compression. The right vena saphena magna diameter was 5.7 mm and there was no reflux in the right saphenofemoral junction. There was no deep venous insufficiency or deep venous thrombus.

Decision for surgical repair was made after appropriate informed consent. Two perforating venules were ligated surgically. At 3rd month follow up, complete healing of the chronic venous ulcer was observed.

Discussion

KTS is a rare form of capillary-venous malformation [12]. Klippel-Trenaunay-Weber syndrome should be considered in cases with significant arteriovenous malformation due to poor prognosis and different treatment approaches [13].

Clinical features occur at birth or during childhood [9,14,15]. KTS is clinically characterized by capillary malformation, soft tissue/bone hypertrophy and atypical varicosities (often lateral superficial) are cardinal features. Two cardinal features are enough for the diagnosis [1,16].

Deep venous anomalies such as hypoplasia, atresia, aneurysmal dilatation, valve hypoplasia and lymphedema can also be identified [12,14]. In the study conducted by Browse et al., hypoplasia/aplasia of the deep venous system was detected in 18% of patients with KTS [17]. In the study reported by Berry et al, deep venous hypoplasia was detected in 35 of 79 patients [3]. Deep venous malformations frequently associated with the preservation of embryonic venues such as lateral marginal vein (vein of Servelle) and sciatic veins [10]. Persistence of lateral marginal vein was found in 68% of KTS cases [11]. The most common lymphatic anomaly is lymphatic aplasia and hypoplasia, which results in lymphedema [11].

In KTS, macular vascular nevus, lymphangioma, longer extremity, swollen extremity, venous varicosities, pain, thrombophlebitis, dislocation of joints, ulcers, gangrene of the affected extremity can be seen [18]. KTS can cause stasis dermatitis, cellulitis, thrombosis, coagulopathy and pulmonary embolism [3,4,9]. KTS may cause macrocephaly, deep venous hypoplasia in the jugular system and varicosities in the facial veins [3]. Intracerebral hemorrhage may develop due to intracerebral involvement [12]. Congenital bone anomalies such as syndactyly, spina bifida occulta, coxa vara can be seen in the KTS [15]. Capillary malformations can be identified in the gastrointestinal tract, liver, spleen, bladder, kidney, lung and heart [9]. Kasabach-Merritt syndrome was found in 45% of cases and high-output heart failure in 13% of cases [18].

Chronic venous hypertension during childhood may be a cause of leg growth [10,11]. Extreme enlargement of the involved extremity may be in the form of bone elongation or soft tissue hypertrophy [3,9]. Significant leg extension discrepancy is defined and can require orthopedic intervention [3].

KTS is a clinical diagnosis. Doppler USG is used for evaluation of venous structures and assesses patency [4]. Computed tomography, magnetic resonance imaging, lymphangiography, sigmoidoscopy and etc. are used for detection of multi-systemic involvement (pelvic and abdominal visceral involvement, intestinal capillary malformations etc) [4,5,10,15,19].

In KTS, treatment is usually conservative. Surgical treatment is recommended for complications [10,11]. In patients with deep vein thrombosis, inferior vena cava filtration or anticoagulant therapy is used to prevent venous thromboembolism [12]. Varicose veins caused by venous insufficiency are treated with compression stockings and lymphatic edema with intermittent pneumatic compression pumps. Surgery is considered for cases where skin ulcers are persistent and cause recurrent bleeding [11].

Deep venous system anomalies are important in directing the treatment of these cases [4,14,19,20]. In cases of deep vein hypoplasia, marginal vein can be resected since hypoplastic deep veins can spontaneously dilate to almost normal

dimensions after marginal vein resection. In cases of deep venous aplasia, the embryonal vein can be the main drainage vein of the leg and resection is unlikely [11,21]. In cases with arteriovenous fistula, venous stasis increases in vessels causing valvular atresia. In these cases, venous skeletonisation may be performed by completely tying up the arterial inflow vessel [21]. Catheter-directed embolization using coils, particulates, or sclerotherapy may be performed in small arteriovenous fistulas [3]. Foam sclerotherapy and endovenous radiofrequency ablation may be performed for perforating venous insufficiency [22].

Osteotomy, epiphysiodesis, and epiphyseal stapling may be performed if a leg is overgrown [23]. Amputation may be required when leg sizes are large enough to affect daily activities [11,15]. Reducing operations may be performed for soft tissues [18].

The diagnosis of KTS was established clinically in our case (venous insufficiency, port wine stain and increased extremity diameter). Surgical repair was seen fit due to the non-healing ulcer. Awareness of the particular syndrome led us to conduct a thorough examination to rule out other abnormalities such as absence of certain venous structures and AV malformations which would cause other complications. Also the surgical approach was also tailored to fit the situation, with specific identifications of venules leading to the leg ulcer and ligation of those.

KTS should be considered for patients with treatment resistant venous insufficiency and exhibiting other characteristics of the syndrome (port wine stain etc.) Careful evaluation of the venous system should be made to refrain from unnecessary or harmful surgical interventions. Once diagnosed, KTS patients should undergo multi-systemic screening to detect further vascular malformations that may be life endangering.

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