VASCULAR MALFORMATIONS ORIGINAL PAPER

Hemodynamics in Klippel-Trenaunay Syndrome

R Mattassi[1], W Pozzoli[1]

[1]Center for Vascular Malformation "Stefan Belov". Clinical Institute Humanitas "Mater Domini". Castellanza (Varese), Italy

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DOI: 10.24019/jtavr.48 - Corresponding author: Prof. Raul Mattassi, raulmattassi@gmail.com

Abstract Klippel-Trenaunay syndrome (KTS) is a complex vascular malformation that involves extensively the lower limb and may be combined with lymphatic and capillary defects. Very little has been published about hemodynamic of this disease. In this paper, we studied with duplex scan the hemodynamics in a group of 52 patients affected by KTS. In a former study, we recognized the following vascular venous defects: dysplastic venous areas infiltrating tissues, dysplastic superficial veins, hypoplasia and aplasia of deep veins, presence of marginal vein and presence of sciatic vein . The study demonstrates several hemodynamic defects, like venostasis and reflux in superficial dysplastic veins, blood stasis in infiltrating, mainly intramuscular, venous malformations, reflux and venostasis in large marginal veins, no main reflux in sciatic vein and dilatation of calf veins with some stasis but no reflux. Duplex scan study is mandatory in KTS but need to be an oriented exam in order to find out main defects existing in KTS. Standard examination, like in varicose veins, is not a correct study as it may ignore some main defects.

Keywords Klippel-Trenaunay syndrome, vein aplasia, marginal vein, venous malformations, intramuscular venous malformations

Introduction

Klippel Trenaunay Syndrome (KTS) has been described in 1900 as a congenital vascular disease of the lower limbs characterized by a triad of clinical signs: cutaneous nevus, limb hypertrophy and superficial abnormal veins[1]. At that time, only clinical descriptions were possible, as no diagnostic tools were available. In more recent times, several publications about the disease were available but a great majority regards single case descriptions[2,3]. Systematic analysis of hemodynamics in a series of patients affected by KTS has rarely been performed. A recent review of the literature shows that very often diagnosis is performed by MR and venography rather than by duplex scan[4]. However, a recent single study demonstrates the diagnostic efficacy of color Doppler in KTS[5]

Hemodynamic of a dysplastic venous system depends on the anatomical anomalies of veins. Knowledge of the possible anomalies that may exist is necessary for a correct hemodynamic analysis of KTS. In a former study we got that data on 46 cases[6]. Based on that knowledge, we analyzed with duplex scan a group of patients affected by KTS in order to find out flow anomalies. A main problem in studies about KTS is that the definition is controversial, as the role in the combination of venous and lymphatic and even AVM is not clearly indicated in many papers. Even the name of the defect is still today unclear, as the wrong term of Klippel-Trenaunay-Weber (a type of "combination" of Klippel-Trenaunay and Parkes Weber syndrome, which is meaningless) is often used. For that reason, strict selection criteria of patients to include in this study has been adopted, according to a consensus of experts[7]

Material and methods

A group of 52 patients, observed between 2011 and 2017 were included in this study. Inclusion criteria were:

- diffuse vascular disease of the whole limb: limited defects only of the thigh or of the calf were excluded as they do not meet the criteria of a diffuse, extended defect
- absence of artero-venous defects. First selection was done by clinical criteria; a duplex scan that rule out AVM was then also performed
- presence of venous anomalies. Patients without that criteria, like pure lymphatic dysplasia were excluded from that study

Types of vascular defects were recorded by duplex scan and MR. Hemodynamic defects in each case were studied by duplex scan.

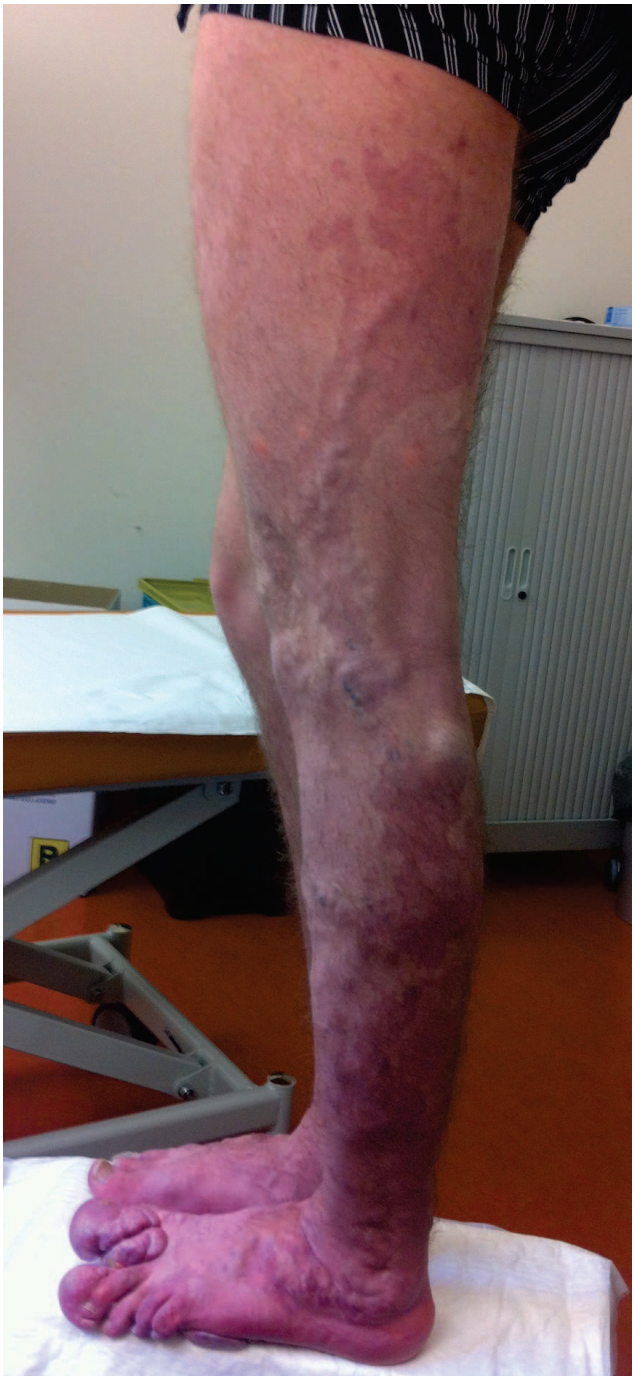

Figure 1 - Dilated abnormal superficial lateral veins in KTS

Result

In a former study on vascular anomalies in KTS, the following anatomic defects in 46 cases were recognized[6]:

- superficial dysplastic veins, like varicose veins but with a dysplastic wall (100%)
- deep infiltrating dysplastic veins (41%)
- marginal vein (30%)
- deep vein aplasia (19%)
- deep vein hypoplasia (19%)
- sciatic vein (7%)

Based on that list of anomalies, we analyzed our cases of KTS by duplex scan study, looking for those defects and trying to find out hemodynamic anomalies.

The following hemodynamic anomalies were observed:

- 1) Reflux in deep main veins (femoral and popliteal vein): 22 cases of 52 (42%).
- 2) Venostasis (blood stagnation in standing position) in superficial abnormal veins. These veins were often placed on the lateral edge of the limb, extending in some cases proximally until gluteus. Wide or extremely wide (up to 1 cm diameter) lateral perforating veins were sometimes recognizable. Locations were variable; lateral side of the calf (in several cases a huge, lateral perforating vein were sited just below the peroneal epiphysis), lateral side of the thigh and gluteus laterally but also on the posterior surface. These veins were not considered part of a marginal vein because there were no main longitudinal lateral located vessel that drain blood from distally to proximal, but only varicose like vessels with an oblique or transversal course. Significant reflux were recognized through these perforating veins with stasis on superficial dilated veins. All cases had dysplastic superficial veins with reflux (fig 1).
- 3) Stasis in deep infiltrating intramuscular venous malformations. Areas of sponge-like, dysplastic venous areas infiltrating muscles were common in these cases. The hemodynamic study demonstrates venous stasis in these areas, mainly in standing position, in which these defects filled with blood and enlarged, compressing the tissues and provoking discomfort. 23 cases (44%) (fig. 2).

Anatomic defect	n°(%)	Hemodynamic defect	n°(%)
Dysplastic superficial veins	52 (100%)	Reflux	52
Intramuscular dysplastic veins	23 (44%)	Venous stasis	23
Marginal vein	15 (29%)	Reflux	15
Sciatic vein	4 (8%)	Slight stasis, no reflux	4
Normal deep veins	40 (78%)	Reflux	22 (42%)
Dilated calf veins	8 (16%)	Venous stasis	8

Table I - Vein anomalies and hemodynamic defects in 52 cases of Klippel-Trenaunay syndrome

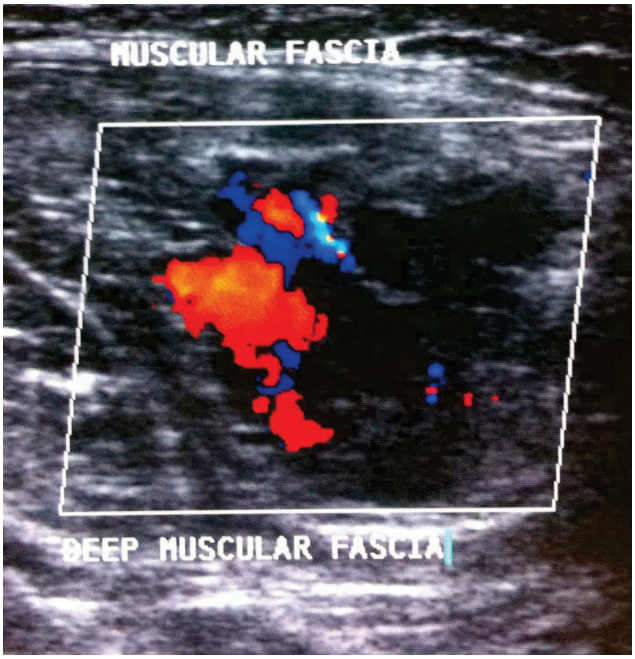

Figure 2 - Duplex scan picture of area of dysplastic veins inside muscle

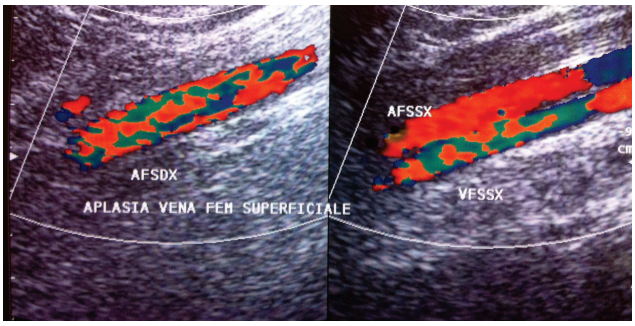

Figure 3 - Duplex scan of femoral vessels demonstrating aplasia of right femoral vein: only the artery is visible (on the left in the picture). Compare with the contralateral side (on the right of the picture) where both vessels are clearly visible.

- 4) Reflux in marginal vein. These abnormal, lateral veins, always demonstrate reflux. We couldn't find valves in all cases analyzed. In some small marginal veins, reflux was less evident, but always existed. All 4 four types described by Weber were recognized[8].
- 5) A sciatic vein was found in 4 cases. This was a large, deep, intramuscular, posterior thigh vein connecting with popliteal vein through several branches which ends proximally in several smaller veins, sometimes connecting with branches of deep femoral vein. In all 4 cases no marginal vein, which is located superficially, was found. Even if this vein was large in all cases, no reflux could be demonstrated but only relative flow stasis.
- 6) Calf veins were dilated in several cases; no reflux but only stasis were noticed.
- 7) Deep vein aplasia and hypoplasia had hemodynamic anomalies related to the adjunctive defects, like superficial dysplastic veins, marginal vein and deep infiltrating intramuscular malformations (fig. 3)

Venous anomalies and hemodynamic defects recognized in this study are summarized in Table I:

Discussion

KTS is a combination of venous defects (sometimes also of lymphatic anomalies) in different forms, originating different hemodynamic anomalies. That means that there is not a "standard" hemodynamic condition in KTS but it varies according to the anomalies existing. These flow anomalies explain symptoms.

Recognition of the defects by duplex scan is required for a correct, symptoms oriented, treatment strategy. However, for a complete duplex study in KTS, knowledge of the different types of anomalies that may exist is required. The standard study of deep reflux and of condition of

the saphenous veins, commonly performed, is absolutely incomplete. Search of marginal vein, abnormal perforating veins, hypoplasia or aplasia of deep veins, flow in abnormal lateral veins (which are different from marginal vein – see above), study of soleal veins and search of sciatic vein are necessary for a complete hemodynamic study o KTS.

The different types of anatomical and hemodynamic defects explains symptoms. Main discomforts were due to *superficial dysplastic veins*, especially if these were extremely large and with reflux through huge perforating veins, sited mainly laterally on the limb. These veins, which are different from marginal vein because of their course (not vertical on the lateral edge of the limb by variable and often transversal), represent a truncular defect because of their primitive vascular structure (thin walls with a high tendency to bleed during surgery).

Another cause of severe discomfort with pain was the *infiltrating dysplastic venous malformations, sited mainly inside muscles*. These sponge like vascular areas are cause of pain due to blood stasis and compression on surrounding tissues and also because of chronic thrombosis inside. These mechanism is demonstrated by permanent increased D-dimer. In case of extended malformation a local intravascular coagulopathy (LIC) may develop with consumption of coagulation factors and lowering of fibrinogen (fig 1).

Marginal vein may be also a cause of discomfort due to stasis distally, especially if the vessel is of large size; small marginal veins may have less stasis and can be also symptom free.

Sciatic vein may be even very large; however, no sciatic vein related symptoms were noticed in our cases. Probably the fact that in all our cases no main reflux were noticed because these veins had no main high connections with reflux. However, cases with a main, direct connection to main pelvic venous system has been described; these cases may be symptomatic[9].

Some sense of heaviness could be related with cases with *abnormal dilated calf veins*. However, coexistence of other cause of stasis, like superficial abnormal veins, intramuscular malformations and marginal vein made it difficult to distinguish origin of symptoms.

Conclusion

KTS is a complex, mainly venous, vascular malformation that may create significant discomfort due to hemodynamic anomalies. Duplex scan is the first, most important, diagnostic tool. To study KTS with duplex scan it is mandatory to know which anomalies may exist in this disease in order to perform a specific exam searching for the possible venous defects described before. That means also to study, for example, the muscles and to look for abnormal vessels inside, as also to search marginal vein on the lateral edge of the limb and of sciatic vein in the posterior thigh. A standard venous duplex scan is not sufficient to study hemodynamics of KTS.

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