KTS was first described in 1900 by the French physicians Klippel and Trenaunay [3] and was classified by You in 1983 into 5 levels of severity [4]. There is no documented gender or racial predilection but cases of similar demographic [6]. Complete pattern of KTS has not yet been demonstrated but alterations in the VG5Q gene on chromosome 5 may result in the vascular defects. In some patients the mutation of chromosomes 8q22.3 and 14q13 have been observed. Life expectancy depends on the severity of the malformation.

Suspected patients should be evaluated with imaging studies in order to differentiate hemangiomas from vascular malformations: CT and MRI should be used for visualizing the extent of the lesions and the depth of tissue infiltration [7].

Additional imaging studies can be used to improve the diagnosis (color-doppler ultrasound, radiography, ascending phlebogram with or without contrast material).

The complicated lesions are the most common causes of debilitating pain in patients with KTS. Frequent complications related to venous abnormalities are: chronic venous insufficiency, cellulitis, infections, superficial thrombophlebitis and deep vein thrombosis [8].

Other complication, described by Maari et al. [9] in 2004, were: leg length discrepancy, cellulitis, severe pain, thrombophlebitis, pulmonary embolus, coagulopathy, mental retardation, necessity of limb amputation, gastrointestinal bleeding and death.

Patients with KTS should receive multidisciplinary medical care. Treatment of KTS patients has consisted mainly of conservative medical management, including compressive stockings and anti-inflammatory medications for pain relief. Operative treatment has been controversial and surgery on the superficial venous system has been reserved for patients with intact deep systems only. Patients with patent deep veins can be considered for excision of symptomatic varicose veins and VMs venous malformations [10].

Frasier et al. [11] describe an alternative treatment of superficial venous varices utilizing endovenous thermal...
ablation via radiofrequency (RFA) complimented by ultrasound guided sclerotherapy in KTS patients as a minimally invasive and potentially efficacious treatment. Open technique or subfascial endoscopic perforator surgery are also described for incompetent perforator veins surgery.

Excision of varicosities is often incomplete and VMs may recur in 50% of patients. For this reason the preferred treatment in uncomplicated cases is, in first instance, conservative approach [10].

2 Case Report

We present a case of a 77-year old woman that was referred to our Emergency Department in December 2014 because of intense pain due to ulcers of the lower limbs and asthenia. Her medical history was significant for diabetes, arterial hypertension, chronic gastropathy and previous cholecystectomy, hysterectomy and left hip prosthesis. The patient was seen by our department 5 months prior in order to evaluate varicose veins (Fig. 1). She was affected by Klippel-Trenaunay Syndrome (diagnosed 30 years before). The patient's right shoulder displayed a port wine stain and a slight deformity of the right leg; the right leg of the patient was longer than the other one from birth. We proposed another screening with color-doppler ultrasound, ascending phlebogram and vascular-RM to confirm the diagnosis of KTS but the patient refused it. From then, we have not had any news of patient up to the access at the Emergency Department.

On admission, the patient presented with dirty bandages, upon removal of the dressings, multiple ulcerations were present with larval overlay that consumed the entire leg extending proximally to the pubis. The ulcers were clean. We cleansed the leg in a solution of povidone-iodine and physiological solution for 15 minutes. Than we rinse the leg with plenty of water. The procedure was repeated three times and parasites were completely removed.

Due to the patient’s obvious failure to thrive, she was admitted to the Medicine Department. She was treated with broad-spectrum antibiotics and supportive therapy. Larvae were analyzed and were classified in: houseflies, cheeseflies and sarcophagaflies.

One day after admission, we observed additional larval presence in her wounds. We repeated our cleaning regimen performed one day prior. Complete drainage required 10 days of treatment. No sign of bacterial infection was observed. Currently the patient has been assigned to a social worker and she has agreed to perform the complete screening for KTS to confirm the diagnosis.

Ethical approval: The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent committee.

Informed consent: Informed consent has been obtained from all individuals included in this study.

3 Discussion

KTS can be diagnosed on the basis of any 2 of 3 features: cutaneous capillary malformations, soft tissue or bony hypertrophy and varicose veins [12].

You [4] in 1983 described 4 levels of severity in KTS respect to varicose veins: in the I Class the features are venous dysplasia and phlebectasic dysplasia; the peculiarity of II Class is Arterial dysplasia. In the III Class described arterial and associated venous dysplasias, phlebectasia without arteriovenous shunts and angiodysloaisas with shunt (Kllippel-Treaunay-Weber syndrome). The most serious Class is the IV with mixed angiodysplasias (atypical KTS) [4].

Our patient belongs to the first class. The diagnosis more over are those made in children who have a deformity of a limb; in our case the diagnosis was made when the patient was 40 years old.

After diagnosis, the patient had not been followed by the specialist and no therapies was performed.

Some authors described an association with parent but without a statistical significance [13]. In our case we had no information in regards to her family medical history. Ulcerations associated with diabetes are the most common cause of foot ulcers and approximately 15% of individulas with diabetes will develop foot ulceration during their lifetime [14]. In literature there is no evidence of association between diabetes and KTS but diabetes is a worsening factor in development of venous ulcers. Usually, the chronic wound microenvironment is complex and contaminated by more than one species of bacteria. In our report the patient had a colonization of the bandage of her leg from fly eggs. In a second time, the development of larvae have colonized ulcerating tissue. Finally, necrotic tissue allowed the larvae propagate. Bacteria produce biofilm as a protective mechanism which subsequently leads to an increase in an resistance from their hosts and their defenses, antiseptics, and topical or systemic antibiotics [15]. In our study, we propose that the larvae were beneficial to the integrity of the patient’s wounds. A complete debridement of necrosis and devitalized
and infected tissue was carried out by the larvae. The beneficial effects of using larvae in wounds were first noticed by Ambrose Paré in 1557 [16]. The first clinical application of larval therapy was performed by Zacharias and Jones during the American Civil War [17].

Scientists first postulated that the debriding action of larvae was due to their mechanical wriggling using a pair of mandibles/hooks for movement and attachment [18].

Recently Chamers et al. described three proteolytic enzyme classes have been identified in the maggot excretions that can degrade extracellular matrix components, including laminin and fibronectin [19-25].

Also in our case the maggots were the responsible factor of complete debridement of necrosis and sovrainfected tissue. After this event the patient is back to being completely asymptomatic.

Usually the patients with KTS are successfully treated with surgery or radiofrequency ablation of incompetent veins; probably, in our case, because of asintomaticity after conservative treatment and because patients' history. Unusually, in our case, the therapy of infective complication of venous ulcers in KTS, was larvae.

4 Conclusion

KTS is a rare syndrome that involves multiple clinical presentations. The most complex cases require a surgical therapy while simple cases only a conservative therapy. In the case of complications due to venous ulcers linked to the syndrome, classic loco-regional treatment and constant monitoring is preferred.

The report we analyzed is certainly a strange case of superinfection and therapy at the same time in a patient with complicated venous ulcerations in KTS.

Conflict of interest statement: Authors state no conflict of interest

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