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Four cases of Klippel Trenaunay-Weber Syndrom seen at Jra Antananarivo University Hospital

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ABSTRACT: Objective: We report the management of four cases of Klippel Trenaunay syndrome (SKP). This syndrome is a malformative congenital association characterized by the clinical triad of: cutaneous hemangioma, arteriovenous fistula or venous varicosities; unilateral hypertrophy of soft and bone tissues. **Methods**: Retrospective and descriptive the study was performed in the cardiovascular surgery unit of the university teaching hospital / JRA Antananarivo. All cases of KlippelTrenaunay syndrome seen within cardiovascular surgery during the follow periode: June 2004 to October 2017 were included. **Results**: Four cases of Klippel Trenaunay Syndrome were collected during 12 years –studie periode-.
The mean age is of 14.6 year-old with extremes of [6; 32], a female preponderance with a sex ratio of 1.5. The association of limb pain and limb varicose represents the circumstance of discovery in 100% of including patients. The evolution was unfavorable marked by the occurrence of limbs pain in 100% of cases, the limitation of the movements, extension of the angiomas, the inequality of the limbs in 75% of cases, phlebitis in 50%of cases and the sequelae aesthetic (100%). There is no death.
Discussion and conclusion: Our four cases present at least the 2 diagnostic criteria. The etiology remains unknown and the incidence low. Bone hypertrophy causes for an inequality in limbs of average of 9 cm requiring orthopedic and / or surgery management.
A vein stripping is necessary in case of painful varicose veins.

Keywords: Vascular Malformation –Klippel Trenaunay Syndrome- inequality of limb.

Introduction

Klippel Trenaunay-Weber Syndrome is a rare congenital disorder (1 to 10/100000 births) [2] characterized by a classic triad [1]: cutaneous hemangioma, arteriovenous fistula or varicose veins, unilateral hypertrophy of soft and bone tissues. These three principal characteristics describe this syndrome but it could be related with other anomalies like lymphatic obstruction, distal limb lipodermatosclerosis, affectation of the abdomino-pelvic vasculature leading to varying degrees of vascular malformations involving the gastro intestinal system, genito-urinary and central nervous system.

The aim of this work is to report our four cases considering the peculiarities of a late management context

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Observation

CASE 1

A 10-year-old girl, first child of a sibship of four, with no particular medical history, was referred to our service for cutaneous hemangioma of the left hand evolving since birth. At 8 years, her last fingers and ulnar rim had increased in volume and becoming varicose.

Clinical examination reports an numbness of the 5th finger. On this finger, there are varicose veins, a cutaneous hemangioma in the form of a punctiform wine stain next to the 5th metacarpus.

On the other hand, the left hand was hypertrophic and one heard a thrill at the hypothenar level.

The biological examination reveals a PAL> 5N. Standard X-ray of the left arm showed hypertrophy of the soft parts with bones of normal volume and density.

Doppler ultrasonography of the left upper limb showed a capillary-like cubito- cubital arteriovenous fistula with low-resistance flow.

The surgical procedure consisted in removing and surgically clipping the arteriovenous shunt at the level of the upper palmar dorsal arch.

The suites were simple.

The child discharged after six days of hospitalization

Active physiotherapy of the hand allowed progressive deflation observed at day 45 and day 60.

CASE 2

An eleven-year-old girl, the youngest of four siblings, who had no particular medical story, was referred to our department for an increasing volume of the right lower limb, and a gait disturbance. She had a blue spot on her right lower limb since her birth, a gait disorder from the 18th month of life, a pain of the right foot exaggerated by the station prolonged, resistant to usual analgesics. Clinic al examination rapports that lateral aspect of the right thigh was strewn with stains of wine.

Planar angiomas were localized on the anterolateral side of this thigh and multiple blackish vesicles in the right leg. We noted an inequality concerning length of the right lower limb of 4cm, with hypertrophy at the level 1/3 middle right thigh of 9cm, a varicose dilatation of the superficial veins of the right lower limb, a flessum of 30° and an articular deformity at walking (Figure 01). X-ray of the right lower limb showed hypertrophy of soft tissue and bone tissue. Doppler ultrasound of the right lower limb found ectasia of the short saphenous vein and the right tibial vein. Patient had benefited from an anticoagulant, an analgesic, an antibiotic prophylaxis, an elastic compression, an orthopedic shoe wear, an active physiotherapy of the right lower limb, and from a crossectomy of the short saphenous vein.

Patient outecomes was unfavorable with persistent difference between the two limbs and a functional impotence of the right lower limb. CASE 3

CASE 3

A 32-year-old woman with a medical history of malformation of the right big toe already treated surgically.

The patient is referred for painful venous dilatation of the external face of the lower left limb. Clinical examination revealed a left upper limb hypertrophied with a difference of 2cm compared to the contralateral limb and painful venous dilatation on palpation.

We note at the level of the big right toe a postoperative scar.

X-ray of the lower left limb showed an hypertrophy of the soft tissues and bones.

Doppler ultrasound test found ectasia of the great saphenous vein and an ovarian varix with a diameter of 18 mm.

Thus, the patient had benefited from a ligation and section of the ovarian varix and a wearing of compression stockings.

Patient outecomes was favorable with pain

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CASE 4

A 21-year-old man was referred to our service for varicose veins of the left forearm that had been evolving since he was 8 years old, particularly painful at night with limitation of flexion and extension.
A hypertrophy of the left forearm evolving since childhood is mentioned
clinical examination reveals a left forearm increased in volume.
We note a circumferential difference of 1cm and a difference in length of 9 mm with respect to the right forearm.
We note also dilation of the superficial veins of the left forearm, observable under the skin and painful on palpation.
The movements of the elbow were limited: flexion at 30 °, extension at 10 °.
X-Ray of the left forearm shows an hypertrophy of the soft and bone tissues.
The Doppler ultrasonic of this limb found varicose bundles of the arm.
The patient received an analgesic, an elastic compression, and an active physiotherapy of the left arm.
The patient outecomes was unfavorable with persistent limitation of movements.

DISCUSSION

KTS is a malformation associating a clinico-radiological triad. Our 4 cases had at least 2 diagnostic criteria. KTS' incidence is low and the average age of discovery is 9 years [3], with a especially female predominance [4] That is consistent with our cases. However, Charlene and Al report the same prevalence for both sexes [9].
The localization of the growth abnormality in KTS varies considerably: It can be localized at the level of the hemibody, at the level of a limb, or part of a limb [10]. In rare cases, growth asymmetry increases progressively and will localized mainly at the level of the big toes or at the level of the feet [10, 11,12].
KTS' etiology is still unknown [3].
However, Its management should be multidisciplinary both medical and surgical.
It will focus on a correction of functional and aesthetic disorders and a prevention of complications.
The hemangioma should be treated by laser, the arteriovenous fistula treated by a ligature-section, varicose veins by stripping or cross-ectomy, the length inequality of the two limbs by an epiphysiodesis of the joint [5].
The result of surgical treatment is controversial; it is optimal if it is performed before puberty.
This is confirmed by the successfull results observed by Servelle and al on more than 700 operated patients [6].
However, this attitude is considered difficult and ineffective by Mayo Clinic which reports only 40% of success [7].
However, sclerotherapy [7] and and interventional radiographic embolization are an alternative to surgery, which is sclerotherapy [7].
An annual check should be performed according to the clinic.
In the majority of cases, there was worsening in the absence of treatment [8].

CONCLUSION

KTS is a rare condition that requires multidisciplinary management.
The lack of technical and financial resources makes its management difficult in our country.
Neonatal screening and regular monitoring is essential to avoid disabling complications.

CONFLICT OF INTEREST

There is no conflict of interest for this manuscript.

Table 1: Clinico-radiological parameters

	Case n°1	Case n°2	Case n°3	Case n°4
AVF and /or varicose	+	+	+	+
Angioma	+	+	-	-
Hypertrophy of soft tissue	Dc : 5mm	IL : 4cm Dc : 9cm	IL : 0 Dc : 2cm	IL : 9mm Dc : 1cm
Standard Radiography: HTO	-	+	+	+
Echo-doppler : AVF/Varicose	+	+	+	+
Evolution	Favorable	Unfavorable	Favorable	Unfavorable

Dc = circumferential difference; **IL** = inequality of length; **HTO** = bone hypertrophy; **HTM** = soft tissue hypertrophy; **AVF** = Arterio-venous Fistulas

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Figures

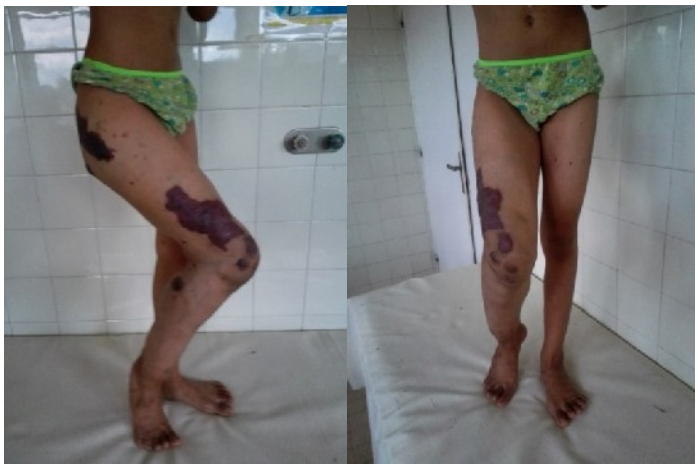

Figure 1: SKP on right lower limb in an 11-year-old girl.

Figure 2: SKP on left lower limb in a 32-year-old patient.

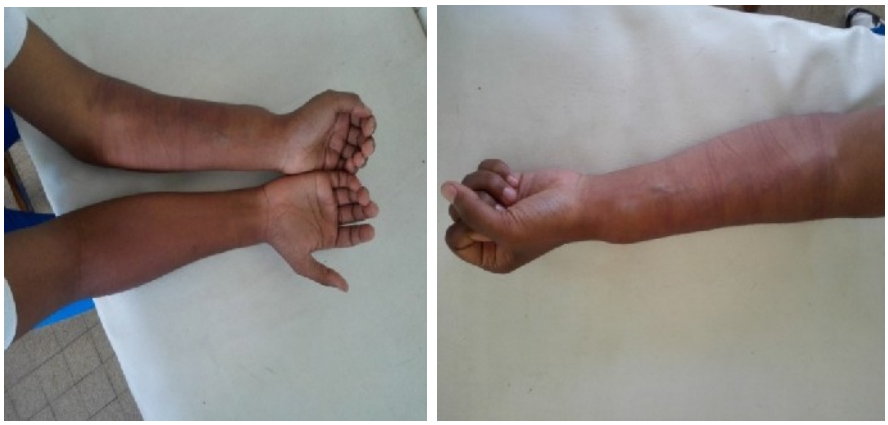

Figure 3: SKP on upper left limb in a 21-year-old man

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