Advances in Clinical and Medical Research

Genesis-ACMR-1(1)-04
Volume 1 | Issue 1
Open Access
ISSN: 2583-2778

Klippel-Trenaunay of the Child: Case Report

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Citation: Ziani J, Baybay H, Dahouki S, Eloudi S, Douhi Z, et al. (2020) Klippel-Trenaunay of the Child: Case Report. Adv Clin Med Res. 1(1):1-4.

Received: May 1, 2020 | **Published:** May 18, 2020

Abstract

A multidisciplinary approach for the treatment and prevention of complications is possible for KTS, including pediatrician, internist, phlebologist, orthopedic surgeons, plastic surgeons and vascular surgeons, a radiologist. The absolute indications for treatment are hemorrhages, infections, acute thromboembolism or refractory ulcers. The management of KTS has been largely conservative and a specific treatment for this syndrome is unfortunately lacking. The goal is to relieve symptoms with conservative treatment. Conservative treatment involves compression garments. Compression therapy has been the mainstay of conservative therapy in the form of an elastic garment or a compression bandage. This is beneficial in the management of lymphedema and chronic venous insufficiency. A certain number of minimally invasive procedures have been adopted for the management of venous anomalies. It includes pulsed dye laser treatment. Sclerotherapy has been used successfully but can cause nerve and skin damage.

Keywords

Klippel-trenaunay syndrome; Child; Case report

Introduction

Klippel-Tranuanay Syndrome (KTS) is a rare sporadic disease characterized by a clinical trial of capillary, soft tissue and bone enlargement malformations; and atypical varicose veins. Although this syndrome was first described more than a hundred years ago, the exact incidence has not yet been estimated. Management of this syndrome includes careful diagnosis, prevention and treatment of complications. We report a pediatric case of KTS.

Observation

A 2 year old child born from an inbreeding marriage which was presented to us with a family history of father epilepsy under undocumented treatment who presents from the age of birth erythematous layers with an inequality of the limbs, with clinical examination : purplish erythematous planar angiomas, poorly limited, with a crumbled edge in geographic map partially fading at vitro pressure (Figure 1), sitting opposite the dorsal spine and at the level of the sacral region with an extension reaching up to the member and the face anterior part of the trunk. An inequality of the upper and lower limbs with a circumferential enlargement of the two limbs and of the left hemi-body compared to the right hemi-body (Figure 2).

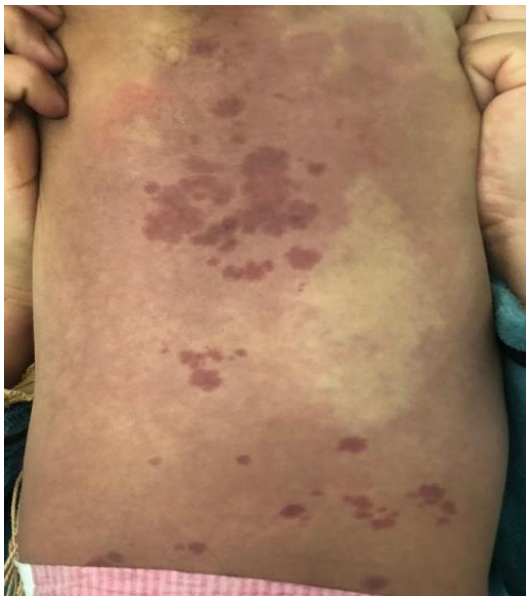

Figure 1: Purplish erythematous planar angiomas, poorly limited with a crumbled edge.

DOI: https://doi.org/10.52793/ACMR.2020.1(1)-04

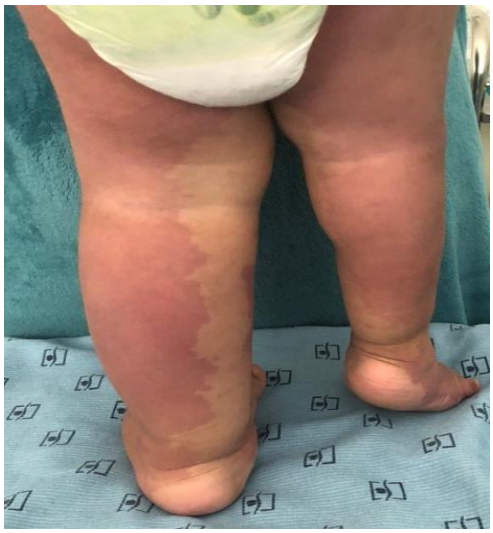

Figure 2: Inequality of the lower limbs with a circumferential widening of both limbs and the left hemi-body compared to the right hemi-body.

The rest of the somatic examination is unremarkable. The child benefited from a radiation measurement of the limbs objectifying an inequality of more than 2cm, a doppler with regard to plane angiomas not objectifying venous anomalies and an MRI of the cerebral medulla returned without particularities. The child was put on compression therapy with elastic restraint and was taken care of by orthopedic surgeons for his uneven limbs and connected with vascular surgeons.

Discussion

KTS is a rare congenital peripheral vascular disease diagnosed at birth, infancy or childhood. Its annual incidence varies between 1 for 40,000 inhabitants and 1 for 20,000 inhabitants and. affects men and women equally, and without racial dominance [1]. KTS occurs on a hemicody with thickening of the limb and swelling and increased skin temperature and uneven limb length [2]. There may be cardiac damage secondary to a decrease in peripheral resistance and an increase in cardiac output as well as modifications such as conjunctival telangiectasia, renal varices and encephalopathy [3]. The pathogenesis of this syndrome is poorly understood. It is not associated with a hereditary familial model with a genetic history or chromosomal abnormalities and occurs sporadically although authors have reported an autosomal dominant hereditary pattern [4]. Another theory suggests a paradoxical model of transmission where this syndrome is produced by a single genomic modification which is lethal [5]. In KTS, varicose veins are birth defects and bone and soft tissue hypertrophy are important consequences. The association of vascular anomalies and lymphatic damage are the cause of bluish or purplish skin lesions.

Radiological investigations are of interest to define and characterize the KTS. Angiography is considered important to assess the local extent of the disease as well as to guide if there is an operative gesture. MRI is recommended, it clearly highlights bone changes in blood vessels. Although it is not specific and

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there is a deficiency in distinguishing KTS from other diseases with vascular involvement. The final diagnosis rests on the clinic with the help of radiological investigations.

Conclusion

The exact incidence and pathogenesis remain unknown. The diagnosis is purely clinical. Management should be rough and provided with lifelong follow-up. An appropriate multidisciplinary approach to management is necessary since the disease affects several organs.

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DOI: https://doi.org/10.52793/ACMR.2020.1(1)-04