A RARE CASE OF KLIPPEL-TRENAUNAY SYNDROME IN 16-YEAR-OLD BOY

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INTRODUCTION

Klippel-Trenaunay syndrome is a rare congenital disorder that was first described by Klippel and Trenaunay in 1900. It characterized by clinical triad include vascular malformations, varicose veins, also soft tissue and bony hypertrophy. Radiology imaging was used to evaluate disorder characteristics and the possible complications, therefore can help determine the management of this disease.

CASE REPORT

A 16-year-old boy came to hospital with a lump in the scrotum to right dorsum pedis which appear since birth. A venography examination showed vascular venous malformations, vascular calcification and soft tissue swelling of the right femur-genu-cruris-pedis region. The lower extremities MSCT revealed a multiple solid lesions accompanied by calcification of the veins and smaller size of the right femur-tibia-fibula bone, so there was possibility of Klippel-Trenaunay syndrome.

Figure 1. Clinical Manifestation

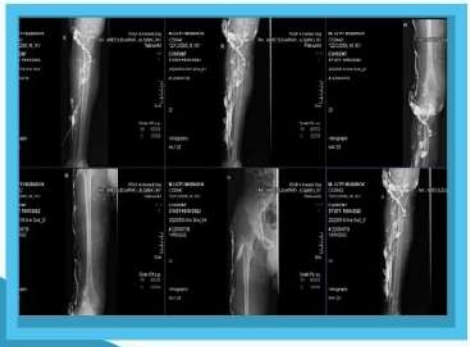

Figure 2. Venografi

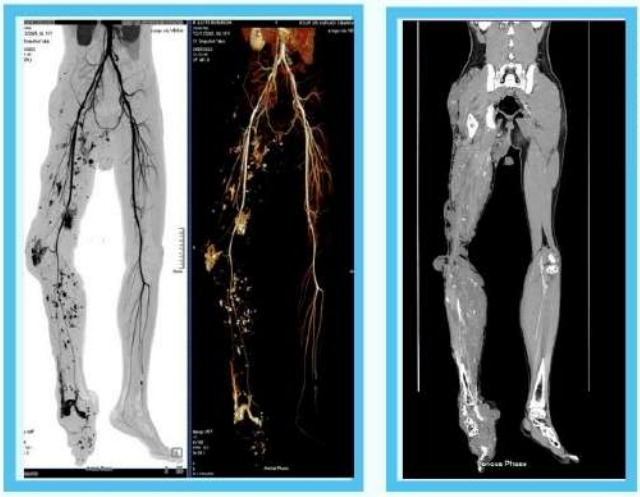

Figure 3. CT Angiography of The Lower Extremities

DISCUSSION

The diagnosis of Klippel-Trenaunay Syndrome usually determined clinically in the presence of at least 2 from 3 clinical triad symptoms such as cutaneous capillary malformations with port-wine naevi appearance; hypertrophy of soft tissues or bones in the extremities; and varicose veins or venous malformations with abnormal distribution. Radiology examinations that can be done such as conventional radiography, ultrasound, arteriography and venography, MSCT and MRI. Those examination were required to confirm the diagnosis and to evaluate the development of the disease, as well as ascertain possible complications that may arise.

CONCLUSION

Klippel-Trenaunay syndrome is a rare congenital disorder with unknown ethiology. Clinical diagnosis is determined based on clinical symptoms triad and radiology examinations are used to help establish the diagnosis, also to evaluate the disease. Excision surgery was performed in this patient and the final diagnosis was mixed-type vascular malformations. Considering that this disease is a congenital disorder, prospective observation of this patient was necessary,.

KEYWORDS

Klippel-Trenaunay Syndrome; Vascular Malformations

REFERENCE

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