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KLIPPEL TRENAUNAY WEBER SYNDROME – A CASE REPORT OF THIS POLYVALENT ANGIODYSPLASIA

Radiodiagnosis

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ABSTRACT

The syndrome of klippel and Trenaunay is a venous angiodysplasia characterised by the classical clinical triad of vascular naevus , varicosities and limb hypertrophy.Its closest differential is Parkes-Weber syndrome which is a mixed high flow arteriovenous shunting anomaly.Diagnosis is purely clinical . Appropriate multidisciplinary approach of management is required as disease affects multiple organs.

KEYWORDS

Klippel trenaunay weber syndrome , limb hypertrophy , naevus

CLINICAL HISTORY

A 4 years old female patient presented to Surgery OPD at New civil Hospital, Surat with chief complaint of dilated veins over outer aspect of left lower limb since birth . She reported that dilatation over the limb increased during walking and standing but subsided on lying down with limb raised . Cutaneous pigmentation was found over left knee.

There was no H/o ulceration , walking difficulty , pain in legs , paresthesia or cellulitis.

On local examination : Varicose veins were found over lateral aspect of left lower limb extending from above the ankle to the upper thigh.

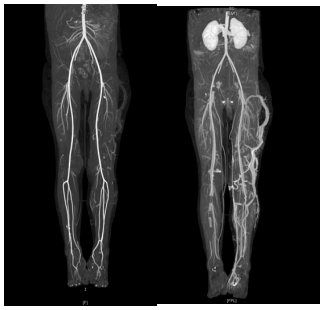
Test results :

Brodie Trendelenburg : SFJ competence
Pratts test :competent deep venous system
Trendelenburg test : competent perforators
Color Doppler of the patient revealed multiple varicosities all over the leg & lower thigh region, few showing thrombus formation within competent deep venous system. Mid thigh & medial leg perforators could be visualized & found normal in size & flow. Sapheno-femoral & sapheno-popliteal junctions were patent

Investigation work-up : CT-Angiography and Venography of both lower limbs was advised . Continous 1 mm axial sections CT-scan from D10 vertebra upto ankle joint immediately after contrast injection . Special reformation CT-angiography of abdominal aorta , aortic bifurcation , iliac , femoral and popliteal artery was done .

FINDINGS :

- There are multiple venous collateral channels noted in left lower limb , i.e. in left thigh , left leg and left foot.
- A large dilated venous collateral channel arising from left common femoral vein with avg diameter measuring 10mm and running along the lateral aspect of left lower limb. It shows multiple communication with other venous collaterals in left lower limb .(S/o Multiple venous malformations)
- Thickened subcutaneous tissue and muscular hypertrophy noted involving left lower limb .



A&B: CT- lower limb angiography coronal image showing Large dilated venous collateral channel ,arising from left common femoral vein and Multiple communications between dilated collateral channel and other channels.

DISCUSSION :

Klippel-Tranuanay Syndrome is a rare sporadic disease characterized by clinical triad of capillary malformation; soft tissue and bony hypertrophy; and atypical varicosity.

KTS is most commonly a sporadic event. Several theories have been proposed, including

Servelle's theory of a primary obstruction of the venous system resulting in venous hypertension and therefore development of abnormal venous pathways and tissue overgrowth. Failure of regression of the lateral limb bud vein is another mechanism. Alteration of the tight balance between angiogenesis and vasculogenesis, which is controlled by numerous genes, among other theories. Berry et al in 1998 speculated that in KTS there is an alteration in vascular remodelling, perhaps at the level of altered angiopoietin-2 antagonism.

The classic clinical triad includes capillary malformations (port wine stain), a usually longer and larger extremity because of soft tissue and bone hypertrophy atypical mostly lateral superficial varicosity.

Patients with at least two of the three cardinal features have been classified as having an incomplete form of KT syndrome.

Deep vein anomalies like venous hypoplasia to frank aneurysm, valve hypoplasia to avalvulia have been decribed.Lymphatic malformations have also been prevalent.

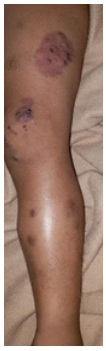
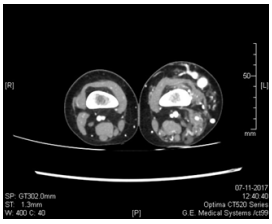
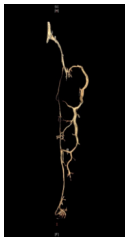
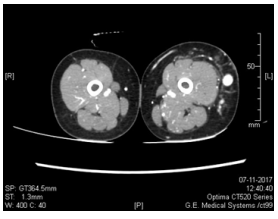
The venous malformations frequently present as persistence of embryonic veins, of which the lateral marginal vein (the vein of Servelle) has been the most typical findingfound in 68-80% of patients.This vein originates from the lateral aspect of the foot and courses upwards along the lateral border of the leg. The vein is usually thick walled and strong, it is located immediately under the skin and it is incompetent along its entire length due to the absence of venous valves.Varicose veins and venous malformations can involve abdominal and pelvic organs.

Genitourinary manifestations may present as intrapelvic and retroperitoneal vascular malformations and affect the penis, scrotum, vagina or vulva, and bladder.

Bleeding from capillary or venous malformations or persistent embryonic veins may occur through defects in the skin or mucosa, or the patient can have intramuscular or retroperitoneal haematoma, haematuria, rectal bleeding, intracerebral or intraspinal haemorrhage.
GI tract involvement may be more common in KTS than previously believed because most of cases remain unrecognized without overt

symptoms. Large cutaneous lesions may sequester platelets leading to Kasabach-Merritt syndrome, a type of consumptive coagulopathy. Rarely, patients with KTS can have intraosseous vascular malformations.

The bony abnormalities may affect all bones in an extremity or limited to one or two bones. Single limb involvement is found in 80-85% patients. Apart from hypertrophy or sometimes hypotrophy of bones other deformities includes macrodactyly, syndactyly, split hand deformity, phalangeal agenesis and dislocation of hip joint.Soft tissue hypertrophy may be limited to a localized mass on the back or chest, or it can be diffuse involving an entire arm or leg.

C: Naevus lesions in the same patient
D: 3D Volume rendered CT-angiography image showing large dilated 10-mm collateral arising from left common femoral vein
E & F : Axial CT image showing a large collateral(big arrow) and multiple collaterals in left thigh and leg(small arrow)

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