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Short communication

An arch-shaped suprapubic (aberrant) cross-femoral vein with aplasia of left common iliac vein: A very rare presentation of Klippel Trenaunay Syndrome

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ABSTRACT

An 8-year-old boy presented with a hugely dilated arch-shaped venous channel over the suprapubic area. Examination revealed that the child had features of Klippel Trenaunay syndrome. USG – colour Doppler study showed absent left external iliac vein in its normal position. Instead the left femoral vein continued over the suprapubic region in the subcutaneous plane to join its counterpart on the opposite side. It carried the venous drainage of the major part of the left lower limb to the inferior vena cava through femoral vein and external iliac vein of opposite side.

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1. Introduction

Klippel Trenaunay syndrome, described first in 1900, is a non-heritable condition. It comprises of the triad of cutaneous vascular malformation especially port-wine stain, bony and soft tissue hypertrophy and venous malformation namely varicosities involving one/both legs or deep venous system anomaly alongwith lymphatic malformation .[1] Parkes Weber Syndrome is a condition when there is association of arterio-venous malformation which may involve vital organs like brain and heart causing more morbidities.

The anomaly usually presents at birth and gradually involves a limb (usually lower) as well as portions of trunk and face. Bony and soft tissue overgrowth of the limb or body parts at times causes difficulty and discrepancy which requires intervention.

Though there are many publications related to this syndrome we have here a very rare presentation of the disease.

2. Case report

An 8 year old boy walked into our OPD with his worried mother to know the future consequences of a transverse swelling over his lower abdomen which was gradually increasing in size (Fig. 1).

He suffered no other significant problem and was good in study. He was born in a Muslim family, of non-consanguineous marriage. There was no family history of similar illness.

On examination, we found a transverse, arch shaped, non-pulsatile, venous swelling of 5×1 cm dimension, situated about 2.5 cm above the symphysis pubis, covered by skin with a bluish-pink hue. At either ends it entered deep into the abdominal cavity. There was port-wine stain involving left side of the face, ear, left arm and forearm including hand, left half of trunk along with left leg. The left arm and leg showed overgrowth both in its length and circumference. Left leg was 1 cm longer than the right. Left lower limb circumference at mid- thigh level was 0.8 cm more and at mid-leg level 0.4 cm more than the right sided measurements. The left mid arm circumference was 0.4 cm more than right but the length of both the forearms were almost same. No facial asymmetry was noticed.

The vitals were stable. There was no rash or bleeding manifestation. Cardiovascular and respiratory system examinations were within normal limits. Head circumference was normal (51 cm) and neurological examination revealed no neurodeficit. Abdominal examination was normal without any organomegaly.

On investigation, blood report showed- total leucocyte count of 8800/cmm, differential count with polymorphs 57%, lymphocytes 37%, eosinophil 05%, monocyte 01%, platelet count-1,80,000/cmm, hemoglobin of 12 gm%, and ESR–22 mm/h. Chest X-ray was normal. X-rays of the bones of left arm and leg showed widening, 0.3 cm more in the middle part than that of right side.

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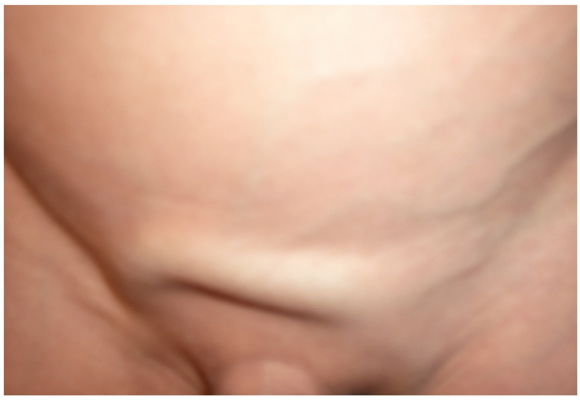

Fig. 1. Suprapubic veinous arch (aberrant) cross-femoral vein.

USG study of the abdomen was normal and showed no organomegaly.

The USG- color Doppler examination revealed one dilated arch-shaped venous channel (aberrant) joining proximal end of left femoral vein with the same vein of right side. The right femoral vein continued into the abdomen as external iliac vein and ultimately as inferior vena cava. There was no external iliac vein in left side and left common iliac vein was not formed in the absence of left external iliac vein. Left internal iliac vein drained into the inferior vena cava. Mild dilatation of deep femoral vein tributaries and moderate dilatation of the mid part of right sided external iliac vein were noted (Fig. 2).

The left great saphenous vein drained in the left-end of suprapubic arch-shaped (aberrant) vein. Another superficial vein which ran along the medial aspect of left thigh up to the knee and had at least three tributaries below knee, drained into the above said arch. No evidence of dilatation or reflux noted in great/short saphenous system. Superficial femoral, popliteal and paired calf veins were within normal limits. Finally the arch-shaped aberrant suprapubic cross femoral vein appeared to be the only major drainage channel for the left lower limb and there was no evidence of thickening or tortuosity/varicosity of any vein in the left lower limb.

The MRI angiography of brain was normal.

3. Discussion

Klippel Trenaunay syndrome with its triad components namely port-wine stain, bony and soft tissue hypertrophy alongwith venous and lymphatic abnormalities, are not much uncommon. There are reports showing aplasia of right common and external iliac veins, bilateral external iliac veins, formation of inferior vena cava by the union of right common iliac vein and left internal iliac vein and congenital absence of entire inferior vena cava resulting in distal pressure and varicosity of vein –though rare, features of klippel Treanaunay Syndrome were not found with these venous malformations.[2–5] Our case of Klippel Trenaunay syndrome along with aplasia of left external iliac vein leading to a hugely dilated aberrant suprapubic vein, which is actually a compensatory drainage route of the left lower limb – a 'natural Palma-Dale bypass' due to the iliac venous occlusion, is being reported as an extremely rare presentation.

In the present, otherwise active child, there is a definite risk of unwarranted catastrophe due to big size and superficial position (under the skin only) of the suprapubic aberrant cross-femoral vein. We counseled the parents for extra protection of the suprapubic area by means of a pad of cotton/linen or elastic

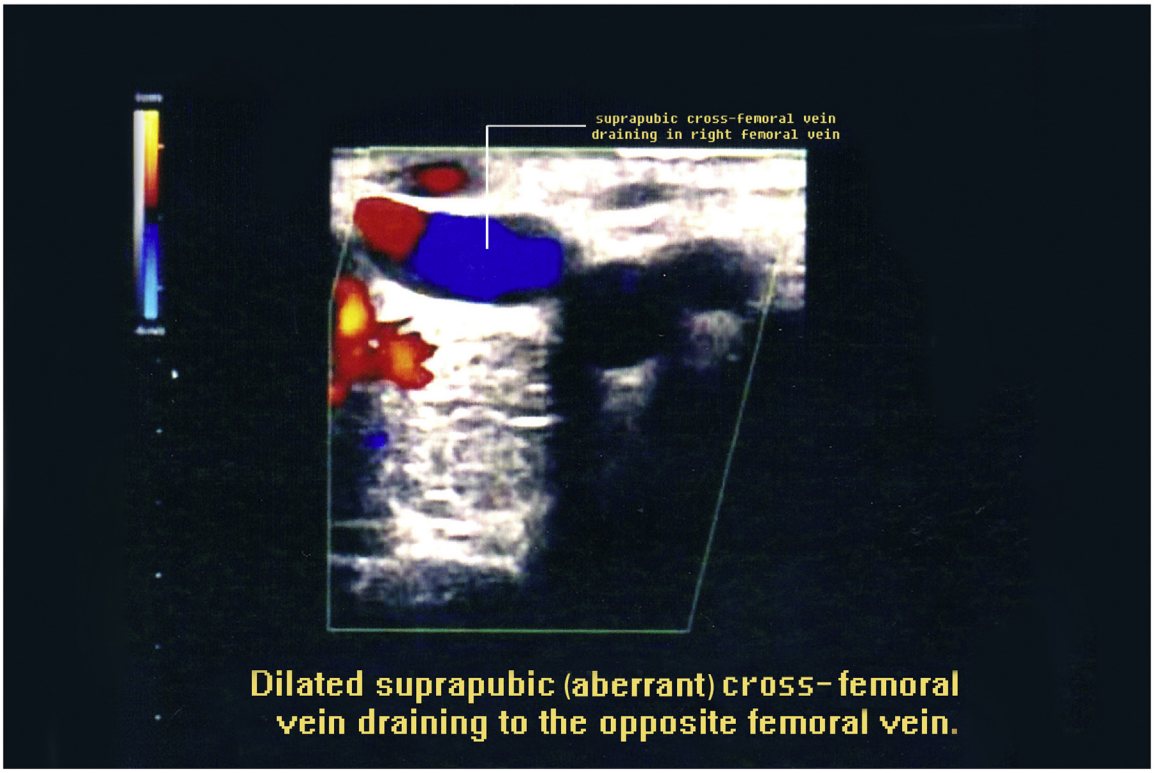

Fig. 2. Dilated suprapubic (aberrant) cross-femoral vein draining to opposite femoral vein.

coverings. Extra care should also be taken for any future surgery (retroperitoneal/laparoscopic) by lower abdominal approach. Venous system in the left lower limb may develop varicosity in near future.

Orthopedic opinion was taken and parents were explained about non-requirement of any intervention for the limb length difference at present, as it was causing no clinical tilting or scoliosis. Moreover, the criteria for epiphysiodesis namely leg length discrepancy of >2 cm or >2.5 cm was not fulfilled.[6,7] Management of Klippel Trenaunay Syndrome is primarily conservative. The necessity for follow-up for development of vascular bone syndrome/angio-osteodysplasia has been emphasized. Mortality depends on severity of the disease and postoperative complications in case surgical intervention needed.

There is no feature of any other system involvement.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Acknowledgement

None.

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