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KLIPPEL-TRENAUNAY SYNDROME

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A 5-year-old girl presented with verrucous plaques with ulceration and bleeding over posterolateral aspect of right thigh and with multiple compressible subcutaneous swellings over the entire right lower limb with lengthening of the same limb.

Key Words : Angiokeratoma, Naevus flammeus, Clitoromegaly, Klippel-Trenaunay Syndrome

Introduction

The association of naevus flammeus on a limb with soft tissue swelling with or without bony overgrowth is generally termed the Klippel-Trenaunay syndrome. One or several macular telangiectatic vascular naevus of port-wine stain type are almost invariably present at birth. Usually the vascular lesions occur in lower limb than the other regions of the body.

Case Report

A 5-year-old girl presented with the symptoms of verrucous plaques over the posterolateral aspect of the right thigh of sizes 12 cm X 6 cm X 11 cm, certain areas over the plaque showed ulceration and occasional bleeding on minor trauma. In addition, patient had multiple compressible subcutaneous swellings over the entire right lower limb with lengthening of the right lower limb and with clitoromegaly. All these symptoms were present since birth (Fig.1).

Routine haematological investigation were normal. Radiological examinations of the right lower limb showed soft tissue shadow without any evidence of calcification. X-ray scalogram comparision of both the lower limbs showed increase in size of right femur by 1.2 cm, right tibia by 1.75 cm and the right febula 0.5 cm longer than the left side. Ultrasound

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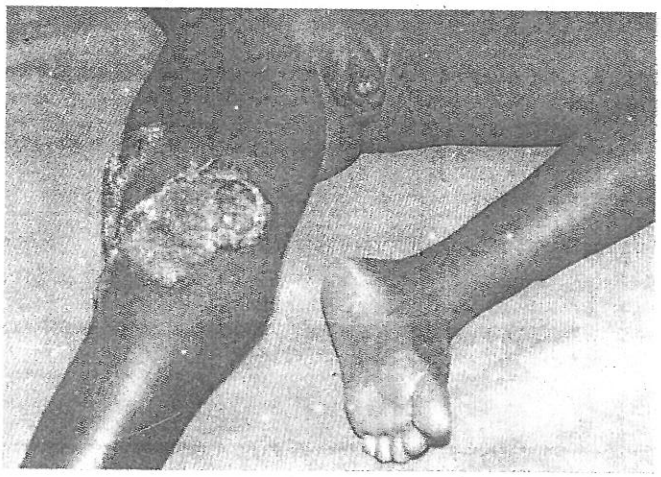

Fig. 1. Clinical picture showing multiple compressible sub-cutaneous swelling with clitoromegaly.

picture of the pelvis and abdomen were within normal limits. Venogram with 60% conray study of the right lower limb showed varicosity of the short saphenous and popliteal veins with incompetence of perforators. There was leaking and pooling of dye into the soft tissue swelling of the right thigh which extended upto the surface of the skin. Presence of contrast in the clitoral region were also seen (Fig.2).

Biochemical analysis and immunoglobulin assay and endocrine study were within normal limits. Histopathological picture of the verrucous plaque was compatible with angiokeratoma. Cytogenetic investigation proved karyotype 46XX.

Comments

Originally, the syndrome comprised of a triad, a port wine stain naevus extending the full lingth of a limb, venous varicosities of the

same limb and overgrowth of all the tissues of the affected limb. Later the term Klippel-Trenaunay syndrome is less specifically used for port-wine stain and increased limb size. A few year after Klippel and Trenaunay paper Parkes Weber described a syndrome which called haemangiectatic hypertrophy. It denote the presence of arteriovenous anastamosis the affected limb.

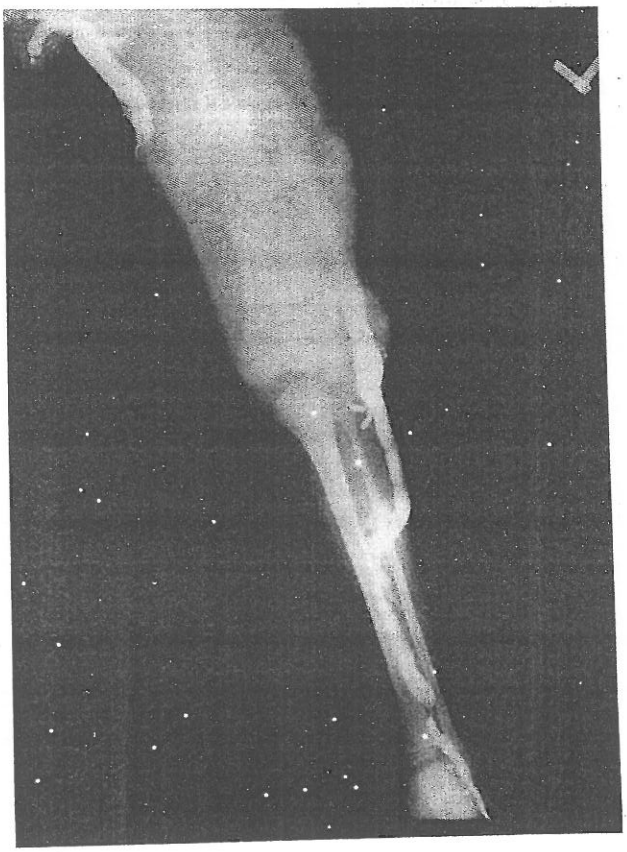

Fig. 2. Venogram with 60% Conray study of right lower limb showing varicosity of blood vessels.

So, this syndrome can be broadly classified into three.

1. Predominantly venous malformations,
2. Predominantly arteriovenous fistulae,
3. Predominantly mixed venous-lymphatic malformations.[1]

These lesions are often accompanied by blue red papules that are more angiomatous than telangiectatic.[2] This case is reported because of its rarity in presentation.

References

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