SKIN HEMANGIOMAS WITH LIMB HYPERTROPHY

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A 9-month-old male child had large irregular compressible bluish purple hemangiomas and port vein stains in the right gluteal region and right lower limb laterally with asymmetric hypertrophy of right lower limb (Figure 1). Blood investigations were normal. Viscera, bones, spinal canal, gastrointestinal and genitourinary tracts were not involved. Ultrasound showed numerous subcutaneous cystic lesions.

Figure 1: Bluish purple hemangiomas and port vein stains in the right gluteal region and right lower limb laterally with asymmetric hypertrophy of right lower limb.

What is the diagnosis?

Klippel-Trenaunay syndrome. It is a sporadic rare cutaneous vascular disorder characterized by a triad of a port-wine stain, varicose veins, and hemangioma along with bony or soft tissue hypertrophy of an extremity associated with life-threatening complications.\(^1,2\) Complications include limb-length discrepancy leading to impaired gait and pain, thromboembolism, bleeding, venous insufficiency, and soft-tissue infection.\(^3\) A primary mesodermal abnormality in fetal development leads to persistence of microscopic arteriovenous communications.\(^1\) The differential diagnosis are Parkes Weber syndrome, Proteus syndrome, Macrodyostrophia lipomatosa, Beckwith-Wiedemann syndrome, neurofibromatosis, soft tissue sarcomas, and lymphangioma.\(^4\) Ultrasound-guided foam sclerotherapy is the state of the art new treatment to potentially close many large vascular malformations.\(^5\) Compression therapies and surgical debulking can be considered.\(^6\)

Compliance with Ethical Standards

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Conflict of Interest: None

References:


\(^5\) Smith PC. Chronic venous disease treated by ultrasound guided foam sclerotherapy. Eur J Vasc Endovasc Surg. 2006; 32:577-583