

A Rare Clinical Presentation of Lower Extremity Edema

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BACKGROUND

- Parkes Weber syndrome (PWS) is a rare disorder with an unknown incidence and prevalence.
- Klippel-Trenaunay Syndrome (KTS) is a complex congenital disorder with a triad of capillary malformation, venous malformation, and limb overgrowth (1). PWS is KTS with arteriovenous malformations (AVMs).
- PWS is associated with RASA1 mutations(2).
- Management requires a multi-disciplinary approach at a facility with experience in congenital vascular anomalies.
- Management goal should be quality of life improvement and preventing complications.
- Embolization and surgical resection leads to clinical improvement (3).

Image 1



CASE PRESENTATION

- 79 yo WM presents to the vascular medicine clinic with cc of LE swelling. Patient reports being told in the past that he has “arterial venous connections” and opted for conservative management. Now having worsening LE swelling, pain, and nodule enlargement (Image 1).
- Venous duplex showed highly pulsatile flow and reflux from the sapheno-femoral junction to the mid calf.
- MRI showed extensive venous malformation and AVF in the medial left thigh and multiple focal masses.
- Cyanoacrylate adhesive attempted, only could close distal GSV.
- Patient was referred for angiograms and consideration of embolization, surgical resection.

DISCUSSION

- Venous and lymphatic malformations have a 1% prevalence, 40% of which involve the LE (4).
- High-flow malformations can lead to compression neuropathy, soft tissue ulceration, bleeding, arterial steal, high output heart failure
- Diagnosis can be made clinically with imaging to confirm.
- Multi-disciplinary approach at a facility with experience in congenital malformations is recommended.

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