Angiomyomatous Hamartoma: A Rare Cause of Lymphedema

Heidi Chen, MD[†]; Nasim A. Chowdhury, MD[‡]

[†] NewYork-Presbyterian Hospital – Columbia and Cornell, Physical Medicine and Rehabilitation, New York, NY

[‡] Weill Cornell Medical Center, Physical Medicine and Rehabilitation, New York, NY

CASE PRESENTATION

A 42-year-old male with no prior medical history presented to our outpatient clinic with a ten-year history of right thigh swelling that progressed from the size of a golf-ball to a watermelon. Prior work-up included normal blood work and ultrasound without evidence of deep vein thrombosis; no additional imaging was pursued.

Physical Exam: non-pitting edema, skin tightening, large flaking skin folds, and positive stemmer's sign. (figure 1)

Diagnostics:

CT abdomen and pelvis: adenopathy in right inguinal and iliac lymph nodes with significant lymphedema (figure 2)

Lymphoscintigraphy: delayed lymphatic drainage in right lower extremity with dermal backflow between the knee and the ankle (figure 3)

PET-CT: right inguinal and femoral hypermetabolic lymphadenopathy

Inguinal lymph node biopsy: angiomyomatous hamartoma

Treatment: tumor excision and lymphedema therapy for complete decongestive therapy

DISCUSSION

Lymphedema is limb swelling from lymphatic fluid accumulation due to imbalance in the rate of lymph production and lymph removal.

-Lymphedema grading on scale from I to III

-Treatment includes complete decongestive therapy with manual lymphatic drainage, compression garments, and decongestive exercises

Angiomyomatous hamartoma is an extremely rare and benign vascular tumor

-Preferentially affects inguinal lymph nodes and consist of fibrous tissue, irregular blood vessels, and adipocytes

-Diagnosis is made by histology and other imaging

-Definitive treatment involves complete excision of the lesion

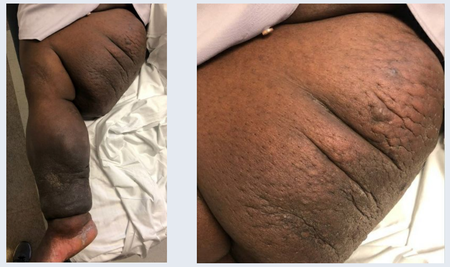

Figure1. Left: Entire RLE with significant lymphedema. Right: Close-up of R medial thigh. Permission obtained from patient to display images.

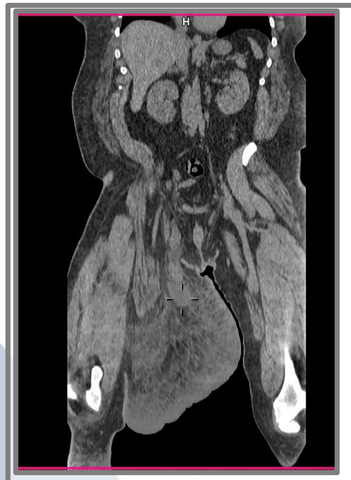

Figure 2. CT Abd/Pelvis: R inguinal lymphadenopathy and lymphedema

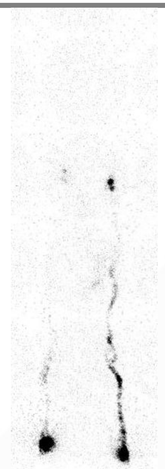

Figure 3. Lymphoscintigraphy: Anterior view 45 min post tracer injection

CONCLUSION

Additional imaging should be considered in cases of unexplained lymphedema. Angiomyomatous hamartoma is a rare, benign vascular tumor that should be considered on the differential for benign and malignant lymph node tumors which may present as lymphedema.

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Weill Cornell Medicine

NewYork-Presbyterian Rehabilitation Medicine

Columbia University Vagelos College of Physicians and Surgeons