PRIMARY LYMPHEDEMA - DO WE HAVE STRATEGIES TO AVOID LATE DIAGNOSIS? - CASE REPORT

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Primary lymphedema is rare disease that implies developmental lymphatic abnormalities leading to the absorption failure of the interstitial fluid which is manifested by characteristic swelling affecting the extremities. There are limited data on PLE prevalence but it is estimated that affects 1.2 per 100,000 people under 20 years of age.

Often, there is prolonged time period between first lymphedema symptoms onset and clinical diagnosis. Prolonged diagnosis of PLE results in high number of undiagnosed and untreated individuals which leads to lifelong physical and psychological morbidity caused by limb deformities and tissue changes.

Diagnosis of PLE should be made on the basis of clinical examination to confirm physical, psychological and functional impact. Specific laboratory tests, lymphoscintigraphy, venous doppler ultrasound, magnetic resonance and genetic consultation may also be performed.

We hereby present a case of 46-year-old nurse with a long-standing history of swelling that affected both of her legs. Path from first symptoms onset which manifested as right ankle swelling when she was 15 until proper diagnosis at the age 35 was not easy and straightforward which led to the formation of non-pitting second stage leg lymphedema. During twenty-year period, she was seen by various specialist being left without proper diagnosis and treatment with low functionality. Leg venous doppler ultrasound was performed on several occasions, but although clinically visible, presence of lymphoedema was not mentioned in the findings. Clinical diagnosis of Meige-like

primary lymphoedema was finally made during 2019. based on clinical examination using The St George's Classification Algorithm. Diagnosis was confirmed in 2022. after lymphoscintigraphy was performed. After proper management lymphedema reduction was accomplished and patient is fully functional. Early diagnosis and proper management of lymphoedema is a key component for preventing complication and achieving high quality of life in patients with primary lymphoedema.