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A rare case of primary lymphedema tarda in an elderly Filipino: Diagnostic and management considerations

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Abstract

Introduction: Primary lymphedema is a benign condition resulting from malformations in the lymphatic vessels or nodes, which may include aplasia, hypoplasia, or hyperplasia of the lymphatic system. It is classified into three types based on the age of onset: congenital lymphedema, lymphedema praecox, and lymphedema tarda. Data on primary lymphedema is limited, and this case represents the second documented instance in the Philippines.

Case description: We present a 62-year-old Filipino male with a 16-year history of progressively worsening unilateral lower extremity edema. The patient has a history of hypertension and stroke but no history of cancer, surgery, or trauma to the right leg. Initial considerations included more common causes of extremity edema. The diagnosis of primary lymphedema tarda was confirmed through lymphoscintigraphy.

Conclusion: Primary lymphedema tarda is a rare cause of lymphedema. Early recognition and understanding of its clinical manifestations are crucial for timely intervention and to mitigate potential disability.

Keywords: Lymphedema; Primary lymphedema; Lymphedema tarda; Edema.

Introduction

Primary lymphedema is a benign condition that is generally caused by malformation of the lymph vessels or lymph nodes. The lymphatic vasculature of affected patients may have aplasia, hypoplasia or hyperplasia. It is classified into 3 types based on the age of onset: congenital lymphedema (clinically apparent at or near birth); lymphedena praecox (apparent after birth to 35 years old; typically, peripubertal years); and lymphedema tarda (apparent after the age of 35). To date there are only a few cases of lymphedema tarda reported, and this is the second case reported in the Philippines [1].

Case report

A 62-year-old Filipino male presented in the clinic with a 16 year history of unilateral leg swelling. Sixteen (16) years prior to consultation, he had an onset of swelling of the right leg but no associated tenderness, warmth nor history of trauma, insect bite. Progressive increase in unilateral leg swelling over the course of five (5) years which prompted him to consult physician wherein he was given Furosemide 40 mg/tab once a day to be taken thrice a week (Monday – Wednesday – Friday) and Spironolactone 40 mg/tab once a day on the remaining days of the week; which provided relief but offered no complete

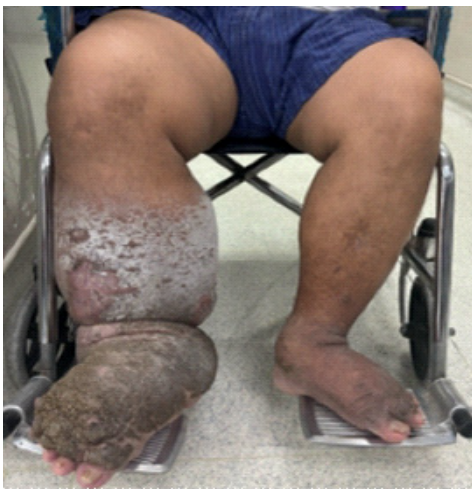

Figure 1: Patient's lower extremities on physical examination with gross swelling, skin thickening, scaly plaques, and woody appearance.

resolution of the edema. He was subsequently lost to follow-up. In the interim, he had progressive increase in unilateral leg swelling for which he intermittently takes Furosemide 40 mg/tab once a day with minimal improvement of the symptom. A week prior to consult, he had a continuing increase in swelling and he noted an open wound on the anterolateral aspect of the right leg. Eventually, this wound developed surrounding erythema, tenderness and minimal bleeding but no purulent discharge. Persistence of symptoms prompted consultation with our institution.

The patient's past medical history include hypertension, chronic kidney disease Stage IIIB and stroke. He had no history of surgery, no family history of cancer or with the same presentation of unilateral extremity swelling. Physical examination was significant with enlarged right lower extremity with a diameter of 73 cm on right thigh, 66 cm on right calf, 48 cm on right ankle and 40 cm on right dorsum, in comparison to the left lower extremity with a diameter of 58 cm on left thigh, 48 cm left calf, 27 cm on left ankle and 22 cm of left dorsum. The consistency of his right leg was firm, with skin thickening, scaly plaques, and a woody appearance. There was a dry wound approximately 1.0 x 1.0 cm on the anterolateral aspect of the right leg with surrounding erythema and tenderness, with normal skin temperature, and a positive stemmer sign– a physical examination finding used to diagnose lymphedema (Figure 1).

The patient was started on Diosmin + Hesperidin 500 mg/tab, one tablet twice a day for CVI and was referred to a Lymphedema Specialist. Following decongestive lymphatic therapy, the patient demonstrated gradual clinical improvement in lymphedema.

Discussion

Differential diagnosis

In a patient presenting with unilateral lower extremity edema, infection is a common cause (eg. cellulitis) which usually presents with localised pain, erythema, swelling and heat. In our patient, he had a history of wound which could be the portal of entry of the infection, accompanied by surrounding erythema and tenderness. However, there was no purulent discharge on the wound, with normal skin temperature. Moreover, cellulitis will present acutely which is in contrast in this case.

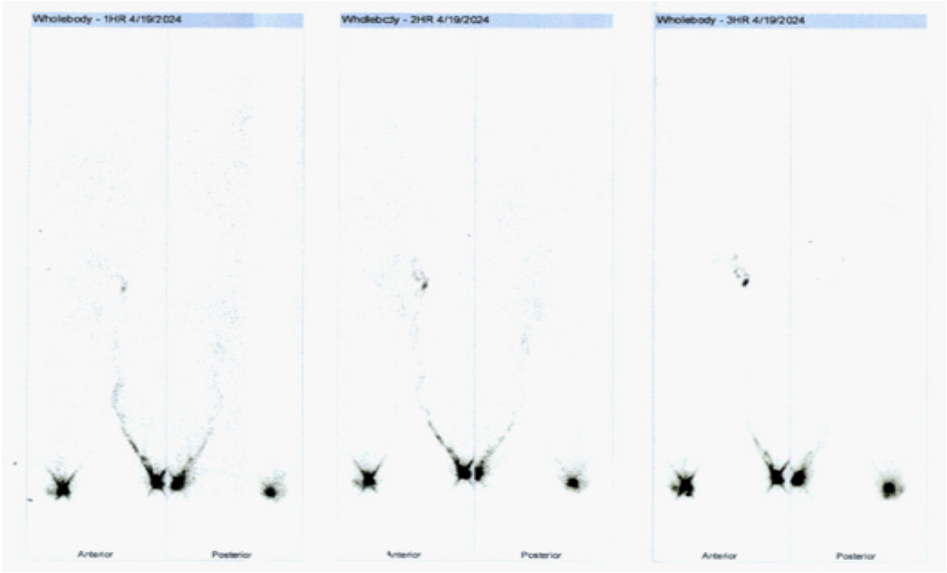

Figure 2: Lymphoscintigraphy results. There was prompt radiocolloid migration from the injection site in the left lower extremity, with visualisation of the left deep lymphatic tract and left inguinal and iliac lymph nodes. Meanwhile, the right lower extremity showed absent radiocolloid migration from the injection site. Early and delayed whole body planar and SPECT-CT images exhibited further tracer accumulation in the left inguinal and left iliac lymph nodes. Meanwhile, the right lower extremity still showed absent radiocolloid migration from the injection site. The right inguinal and iliac lymph nodes were not visualised throughout the entire study.

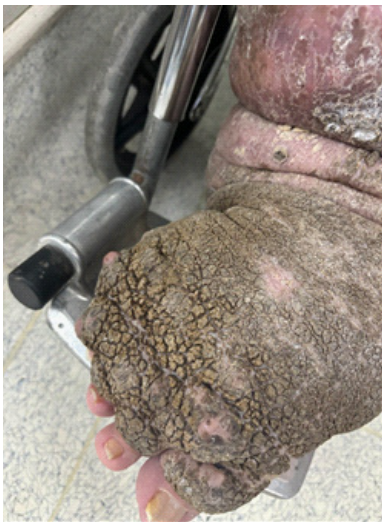

Figure 3: Cobblestone skin texture of the dorsal aspect of the right lower extremity of the patient.

Chronic venous insufficiency (CVI) is another differential diagnosis that we considered in this case. Our patient previously worked as a kitchen staff, which required him to stand for long periods of time which is a risk factor in developing CVI. The venous duplex scan revealed deep and superficial vein valve refluxes in the bilateral lower extremities confirming the diagnosis of CVI. However, it is important to note that CVI could also lead to a secondary lymphedema, known as the "Phlebolymphedema" which is characterised by large collecting lymphatic vessels or nodes that have been damaged, compressed, or severed due to an underlying cause [4]. In here, there is activation of endothelium and inflammation causing the venous hypertension and valvular destruction and edema, this also causes lymphatic system overload causing a dual system dysfunction of the lymphatic pump and dermal lymphatic backflow which causes the lymphedema then again causes the secondary skin changes, the edema also compromises the microarterial perfusion causing leg fatigue, heaviness [6]. In this case, the lymphoscintigraphic findings of the patient was not compatible with Phlebolymphedema.

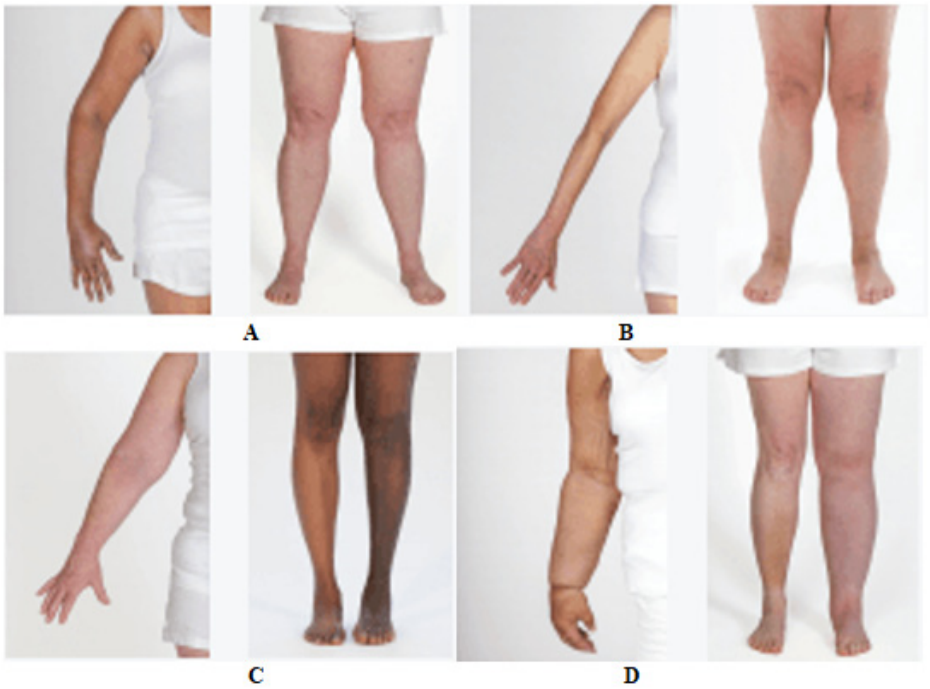

Figure 4: Lymphedema Staging. **(A)** Stage 0 is the latent or subclinical stage wherein Swelling is not yet evident despite impaired lymph transport. **(B)** Stage 1 is described as the early accumulation of fluid in protein content which subsides with limb elevation. Pitting edema may occur. **(C)** Stage 2 is when limb elevation alone rarely reduces tissue swelling. **(D)** Stage 3 is when pitting is absent, with trophic skin changes such as acanthosis, alterations in skin character and thickness. Source: Lymphoedema Management | What is Lymphoedema | Stage of Lymphoedema. (n.d.). https://www.lymphaticdrainage.co.nz/lymphoedema

Additionally, deep vein thrombosis (DVT) should always be ruled out in patients presenting with unilateral lower extremity because it can lead to life-threatening complications such as pulmonary embolism. However, given the temporal profile and physical examination findings in this case, DVT is an unlikely diagnosis.

Lastly, given the temporal profile of progressive sixteen (16) year history of unilateral lower extremity edema, lymphedema should be considered. Lymphoscintigraphy of the bilateral lower extremities confirmed severe lymphatic obstruction with absent radiocolloid migration from the injection site. Early and delayed SPECT-CT images showed further tracer accumulation in the left inguinal and left iliac lymph nodes, while the right lower extremity still showed absent radiocolloid migration from the injection site. These lymphoscintigraphic findings established the diagnosis of primary lymphedema.

Lymphedema

Lymphedema develops when the production of lymphatic fluid exceeds the transport capacity of the lymphatic vasculature [4]. This causes lymphatic insufficiency that results in accumulation of macromolecular proteins normally transported by lymph such as hyaluronan, lipoproteins, tissue antigens and cellular metabolites [11,12]. The lymphatic vessels also transport immune cells, hence this tissue lymph stasis causes an inflammatory response leading to increased cytokines, chemo-

kines that further decreases lymphatic function and causes tissue remodelling by collagen deposition and connective tissue overgrowth [12]. These events lead to fibrosis which manifests as skin thickening, loss of elasticity, cobblestone skin texture (Figure 3), peau d'orange and stemmer sign [3].

Lymphedema staging

Lymphedema can be staged clinically based on the severity of the swelling (Figure 4). Correctly determining the stage of lymphedema is important to appropriately manage our patients.

Stage 0 is te latent or subclinical stage wherein swelling is not yet evident despite impaired lymph transport. Stage 1 is described as the early accumulation of fluid in protein content which subsides with limb elevation. At this stage, pitting edema may occur. Meanwhile, Stage 2 is when limb elevation alone rarely reduces tissue swelling. The limb may not pit as excess subcutaneous fat and fibrosis develop. Finally, Stage 3 is when pitting is absent, with trophic skin changes such as acanthosis, alterations in skin character and thickness [5]. This is most likely the stage of the patient we presented in this paper.

Classification of lymphedema

Lymphedema is classified as primary or secondary. Primary lymphedema has aberrant lymphatic anatomy or function that may include lack of lymphatic capillaries or valve formation, lymphatic hyperplasia, malformations, leaky blood vessels, or impaired pumping. This is classified into three (3) types based on the age of onset: 1. congenital lymphedema which is clinically apparent at or near birth, 2. lymphedema praecox, apparent after birth to 35 years old and typically affects individuals in peripubertal years; and 3. lymphedema tarda, commonly occurs after the age of 35 years [4,12]. For this case, primary lymphedema tarda is the most likely cause since our patient's symptoms started at the age of 46 years.

Primary lymphedema is rare and affects 1 in 1,000,000 individuals while secondary lymphedema affects 1 in 1,000 individuals [8]. Moreover, A study done regarding lymphedema by a tertiary center in the Philippines showed that the majority of the lymphedema patients have secondary lymphedema and only 8% had primary lymphedema [9].

On the other hand, secondary lymphedema is more common than the primary one and it is usually due to insults, infection or obstruction to the lymphatic system [4]. Worldwide, the most common cause of secondary lymphedema is Filariasis, however in developed countries, the most prevalent etiology is malignancy and treatment related to malignancy [8]. According to Timbol et,al (2014), there are 7.7 filariasis cases per 1,000 population in the endemic provinces in the philippines. However, this patient's travel history did not have any case of filariasis [1], and he had no chronic exposure to activities associated with filariasis infection. Blood film microscopy for filaria was done twice for this patient which yielded both negative results.

Diagnosis

Since there are a number of possible causes of the lymphedema, lymphoscintigraphy is crucial to further delineate the cause of the patient's unilateral extremity edema [4,7]. Lymphscintigraphy is a special type of nuclear imaging that provides pictures of the lymphatic system, which transports fluid throughout your immune system. It involves the injection of radiolabeled macromolecules such as sulphur colloid or albumin into the distal subcutaneous tissue of the affected extremity. The progression of radionuclide through the lymphatic system is followed by radioscintigraphic camera [4]. Correlating it to the results of the patient's lymphoscintigraphy wherein there is nondemonstration of radiocolloid migration from the injection site and non visualisation of the right inguinal and iliac lymph nodes, which establishes the diagnosis of primary lymphedema tarda.

Management

Primary lymphedema requires a holistic and individualised approach to management [4]. Primary lymphedema may be treated conservatively or surgically. One of the options for conservative management include decongestive lymphatic therapy with the goal of reducing the edematous limb volume, ensuring optimal health and functional integrity of the skin [4]. Decongestive lymphatic therapy involves manual lymph drainage, compression bandages and garments, exercises and skin care [4]. Surgical treatments are offered to patients with Stage III lymphedema and may include lymphovenous anastomosis, vascularized surgical transfer of the lymph node and palliative reduction procedures [4]. Unfortunately, as of writing, these advanced surgical management of lymphedema is not yet available in the Philippines. Hence, despite having Stage III lymphedema, our patient of interest in this case report was not able to undergo these procedures. Instead, he was referred to a Lymphedema Specialist to undergo decongestive lymphatic drainage.

Managing lymphedema tarda can be challenging. However, recent advancements in treatment offer renewed hope for patients. A number of studies have been done which include lymphangiogenic factors, anti-inflammatory agents and anti-fibrotic therapies were proposed to be used in conjunction with current surgical approaches but evidence is still lacking to establish their roles in the clinical setting and patient management [14]. Studies regarding the use of enzyme-based treatments such as proteolytic enzymes to break down fibrotic tissue in patients with lymphedema is ongoing [15]. Another promising treatment approach involves the use of nanoparticles injected into the lymphatic vessel to enhance vessel function and restore pumping pressure. However, further research is needed to clarify how this method can be effectively combined with existing therapies to prevent or treat lymphedema [16].

Conclusion

Lymphedema may be primary or secondary, with each type having its own classification. Primary lymphedema is diagnosed by excluding other causes, as secondary lymphedema is more prevalent and typically requires more intensive treatment. Accurate diagnosis often involves a series of tests to ensure the condition is not secondary. Managing lymphedema requires close collaboration between the patient and physician, addressing both the physical and psychological impacts of the disease. Decongestive lymphatic therapy (DLT) is a highly effective treatment for primary lymphedema tarda, focusing on techniques such as manual lymphatic drainage, compression therapy, exercise, and skincare to alleviate symptoms and improve quality of life.

Conflict of interest: The authors declare no conflict of interest relevant to this article.

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Appendix

Appendix 1: Laboratory results of the patient.

Table 1: Result of the complete blood count.

	Normal range	Result
Hemoglobin	14 - 18	17.2
Hematocrit	0.40 - 0.54	0.51
White Blood Cell Count	4 - 11	15.2
Red Blood Cell Count	5.0 - 6.4	6
Platelet Count	150 - 450	`91
MCV	80.0 - 96.0	89.4
MCH	27.0 - 31.0	31.6
MCHC	32.0 - 36.0	35.4
Segmenters	50 - 70	74
Lymphocyte	20 - 40	14
Monocyte	2 - 5	2
Eosinophil	2 - 4	1

Appendix B: Result of the Arterial and Venous Duplex Scan of the Lower Extremities of the patient.

Arterial Duplex Scan

Comments: B mode imaging showed plaque formation in all arterial segments of the bilateral lower extremities.

Table 2: Result of the basic metabolic panel.

	Normal range	Result
BUN	2.86-8.21	25
Creatinine	59-104	164
Sodium	136-145	141
Potassium	3.5-5.1	4.4

Normal peak systolic velocities with multiphasis waveform pattern in all arterial segments insonated.

Normal pressures and indices in the left posterior tibial and dorsalis pedis arteries.

Normal photoplethysmographs, pressures and indices in all digits of the left lower extremity.

Conclusion:

Lower extremity arterial disease, atherosclerotic with insignificant stenosis of less than 50% in all arterial segments of the bilateral lower extremities.

Normal left ankle brachial index.

Normal left toe brachial index.

Note: pressures and indices in the right lower extremity were

not taken due to presence of elephantiasis (technical difficulty).

Venous Duplex Scan

Comments:

All deep and superficial venous segments of the bilateral lower extremities are fully compressible with full color display.

Conclusion:

No evidence of deep and superficial venous thrombosis in the bilateral lower extremities.

Deep vein valve reflux in the right femoral; left popliteal veins.

Superficial vein valve reflux in the right small saphenous vein.

Incompetent right sapheno-popliteal and left sapheno-femoral junctions.

Tissue edema in the bilateral ankle and dorsal and right calf areas.

Appendix C: Lymphoscintigraphy result of the patient.

Scintigraphic Findings:

There was prompt radiocolloid migration from the injection site in the left lower extremity, with visualization of the left deep lymphatic tract and left inguinal and iliac lymph nodes. Meanwhile, the right lower extremity showed absent radiocolloid migration from the injection site.

Early and delayed whole body planar and SPECT-CT images exhibited further tracer accumulation in the left inguinal and left iliac lymph nodes. Meanwhile, the right lower extremity still showed absent radiocolloid migration from the injection site. The right inguinal and iliac lymph nodes were not visualized throughout the entire study.

Interpretation:

1.Evidence of severe lymphatic obstruction in the right lower extremity.

2. Patent deep lymphatic tract in the left lower extremity.