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RESEARCH ARTICLE

PRIMARY CONGENITAL LYMPHEDEMA WITH ATYPICAL UNILATERAL PRESENTATION: A REVIEW OF THE LITERATURE AND CASE ILLUSTRATION

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ABSTRACT

Background: Primary congenital lymphedema (PCL) characteristically presents as symmetrical, bilateral lower extremity edema. Atypical unilateral phenotypes, particularly those featuring discordant digit involvement across upper and lower limbs, are exceedingly rare clinical entities presenting unique diagnostic and management challenges. **Case Presentation:** We present a 23-year-old female with an atypical unilateral PCL variant, characterized by asymmetric localized swelling of the left upper and lower extremities that has been present since birth. Distinctively, the edema involved the upper limb digits while sparing the lower limb digits. The chronic course was complicated by recurrent non-healing ulcers starting at age 18, culminating in wet gangrene and subsequent amputation of the left fifth digit at age 23. Recent evaluation revealed recurrent left-hand ulcers. Magnetic resonance imaging (MRI) of both extremities revealed extensive lobulated subcutaneous T2-hyperintense lesions without deep tissue infiltration. Lymphoscintigraphy confirmed congenital lymphatic dysplasia. Active ulcers were successfully treated with collagen dressings and a long-term regimen of combined decongestive and heat therapy was advised. **Literature Review:** A literature review was conducted to differentiate classic PCL presentations with rare ipsilateral variants, evaluating the diagnostic utility of multi-extremity MRI and the long-term efficacy of combined conservative wound and lymphedema management. **Conclusion:** This case illustrates a severe, atypical PCL phenotype, emphasizing the necessity of multi-modal imaging for precise anatomical mapping and demonstrating the clinical importance of collagen dressings and heat therapy for chronic, limb-threatening lymphatic complications.

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INTRODUCTION

Primary congenital lymphedema (PCL) is a rare congenital disorder characterized by the developmental dysplasia or aplasia of the lymphatic system, leading to chronic, debilitating interstitial fluid accumulation[1,2]. The classic phenotype, associated with Milroy disease, typically presents at birth or during early infancy. As documented in foundational literature, such as the classic infant presentation described by Kitsiou-Tzeli et al., the standard clinical trajectory involves symmetric, bilateral non-pitting edema predominantly localized to the lower extremities[3]. While bilateral lower extremity involvement is the established hallmark of PCL, strictly unilateral presentations are remarkably rare[4]. Furthermore, ipsilateral manifestations, where lymphatic dysplasia aggressively affects both the upper and lower limbs on a single side of the body, represent an exceptionally uncommon clinical entity. Diagnosing and mapping these atypical variants requires high clinical suspicion and advanced multi-modal imaging, primarily lymphoscintigraphy and magnetic resonance imaging (MRI), to definitively differentiate primary congenital lymphatic anomalies from secondary, acquired, or syndromic causes of unilateral swelling[5,6]. In this article, we present a comprehensive review of the literature alongside the 23-year natural history of a patient exhibiting a highly atypical, ipsilateral variant of PCL. This case is distinguished by severe anatomical asymmetry, specifically involving the left upper limb digits while paradoxically sparing the left lower limb digits. By detailing her clinical progression from an in utero manifestation to the development of chronic lymphatic ulcers, we aim to highlight the critical role of precise anatomical mapping via MRI[7,8]. Furthermore, this review demonstrates the importance and long-term efficacy of combining advanced wound care, specifically collagen dressings, with conservative heat therapy in managing severe, progressive lymphatic complications[9,10].

Case Illustration: A 23-year-old female presented to the surgical outpatient department with a history of chronic atypical unilateral swelling and recurrent non-healing ulcers on her left hand (Figure 1a). According to the patient and maternal recall, this swelling was clinically apparent at birth and served as the primary

indication for full-term lower segment cesarean section. The patient reported normal childhood growth and development with no family history of similar lymphatic or vascular anomalies (Figure 1b).

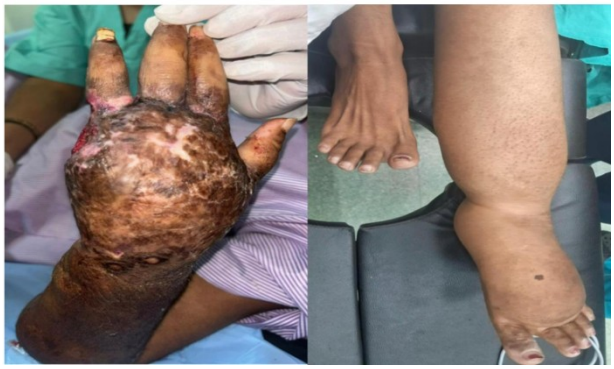

Figure 1a. The current 23-year-old adult phenotype featuring severe asymmetrical left-sided edema with ulceration

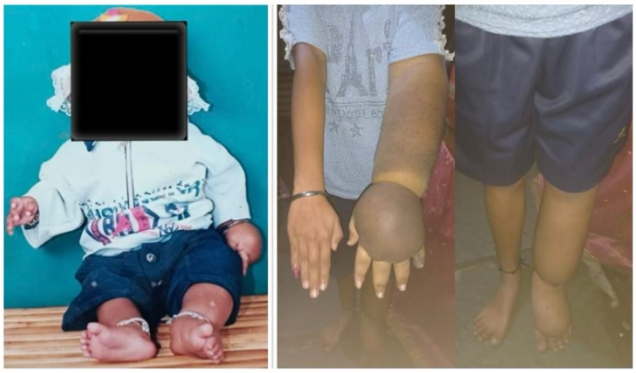

Figure 1b. Chronological clinical photographs illustrating the progression of ipsilateral swelling from infancy to adolescence. The images show evident swelling at 1 year of age with further progression noted at 13 years

Initially, the condition presented as painless swelling without functional limitations or overlying skin changes. The patient noted that prolonged limb elevation and bed rest provided transient alleviation of the lower limb swelling. At 18 years of age, the left upper limb swelling progressed, complicated by the development of non-healing ulcers. The disease course severely worsened at 22 years of age when this ulceration progressed to wet gangrene. She underwent debridement of the left little finger and the dorsum of the hand; tissue swab cultures at that time were positive for Klebsiella pneumoniae. Despite targeted antibiotic therapy, the progression of wet gangrene necessitated the amputation of the fifth digit of the left hand. Upon current presentation, general physical and systemic examinations were unremarkable, with a normal body mass index (BMI) of 22 kg/m3. On local examination, highly asymmetrical, non-pitting edema was prominently noted in both the left upper and lower limbs. Notably, the anatomical distribution demonstrated specific digit discrepancy: the edema involved the digits of the left upper limb but spared the digits (toes) of the left lower limb. Skin thickening was present, though there was no evidence of hyperkeratosis, papillomatosis, or venous dilation. Stemmer's sign was positive in both the left upper and left lower extremities. Limb measurements recorded a left forearm circumference of 30 cm and a left calf circumference of 40 cm. Active, recurrent, nonhealing ulcers were observed on the dorsum of the left hand. A recent swab culture from the active ulcers on the left hand was negative for microbial growth. An Anteroposterior, Oblique and Lateral X-ray of the left hand was performed and magnetic resonance imaging (MRI) of the left forearm and left hand was conducted to map the extent of soft tissue involvement, at 21 years of age (Figure 2a) (Figure 2b).

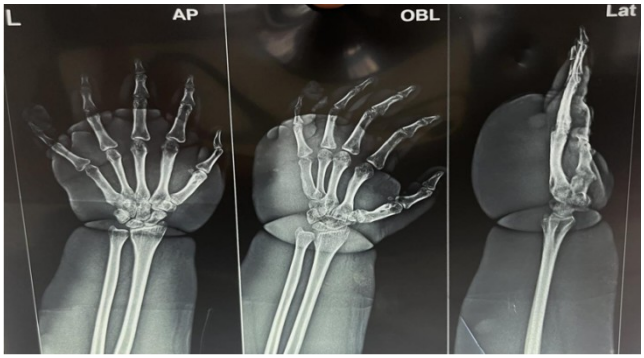

Figure 2a. Anteroposterior, Oblique and Lateral X-ray of the left hand at 21 years of age

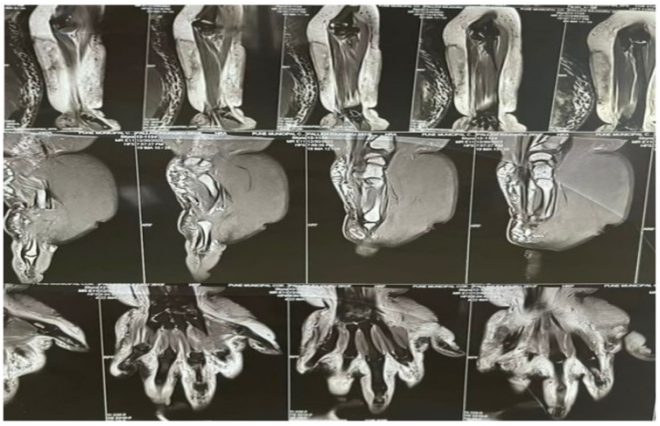

Figure 2b. T2-weighted 3 Tesla MRI slice through the forearm and hand at 21 years of age, demonstrating hyperintense, lobulated subcutaneous soft tissue lesions without infiltration into musculature

The lesions were confined strictly to the subcutaneous plane between the skin and underlying musculature, with no evidence of deep tissue invasion. Lymphoscintigraphy at 23 years of age revealed persistent and significant lymphatic obstruction in both the left upper and left lower limbs, confirming the diagnosis of a primary congenital lymphatic anomaly[6] (Figure 3). The patient was managed conservatively for her chronic ulcerations using collagen dressings applied twice over the duration of one week (Figure 4). The patient exhibited a highly favorable clinical response[9] . Upon discharge, she was advised to maintain a strict regimen of complex decongestive therapy[5] combined with localized heat therapy for long-term lymphedema volume reduction[10].

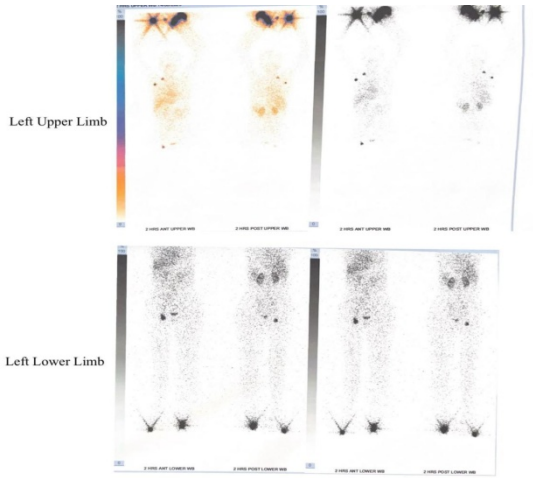

Figure 3. Lymphoscintigraphy performed at 23 years of age. Left upper limb (top) and left lower limb (bottom) panels reveal persistent significant lymphatic obstruction on the left side, confirming a primary congenital lymphatic anomaly

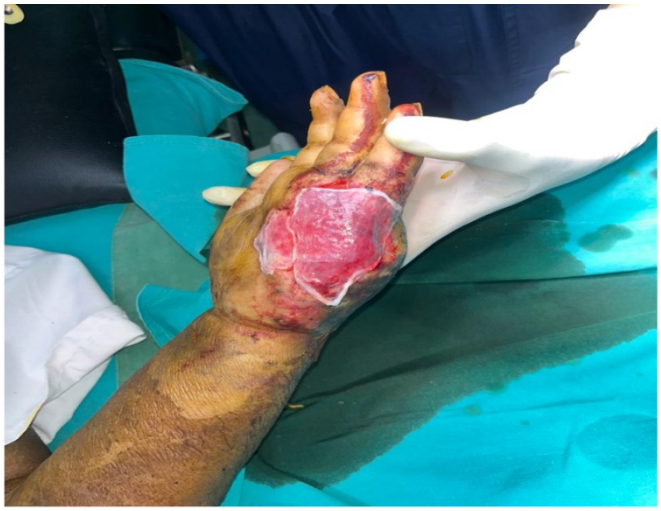

Figure 4. Clinical photograph illustrating the application of collagen dressing on the chronic, recurrent lymphatic ulcerations of the dorsum of the patient's left hand

DISCUSSION

Primary congenital lymphedema falls under the broader spectrum of primary lymphatic dysplasias, which are increasingly categorized by their distinct phenotypic presentations and anatomical distributions[2, 4]. While classical textbook descriptions of PCL emphasize bilateral, symmetrical lower limb edema presenting at birth[1, 3], our patient presented with a strictly unilateral, ipsilateral involvement affecting both the left upper and lower limbs. This asymmetric distribution, coupled with differential digit involvement, represents a rare deviation from standard clinical expectations. The diagnosis and classification of atypical PCL relies heavily on objective, multi-modal imaging. Lymphoscintigraphy remains the established gold standard for assessing gross lymphatic function and confirming congenital obstruction[6] . However, as demonstrated in this case, non-contrast MRI or MR lymphangiography is indispensable for precise anatomical mapping. MRI effectively delineates the extent of subcutaneous lobulated lesions and definitively confirms the absence of deep fascial or muscular infiltration, which is critical for surgical planning and ruling out other complex vascular anomalies[7, 8].

Long-term management of PCL focuses on volume reduction and the prevention of secondary complications, such as severe Klebsiella pneumoniae infection and subsequent gangrene observed in this patient. The International Society of Lymphology recommends complex decongestive therapy as the cornerstone of conservative management[5] . In the presence of chronic, non-healing lymphatic ulcers, advanced wound care is required. Collagen dressings have been shown to significantly accelerate wound healing compared to conventional dressings by promoting healthy granulation tissue in chronic beds[9]. Additionally, integrating simple and cost-effective modalities, such as heat therapy, has demonstrated excellent efficacy in reducing lymphedema volume and improving local tissue compliance in chronic cases[10].

CONCLUSION

Atypical unilateral presentations of primary congenital lymphedema present significant lifelong clinical challenges. This case highlights the critical necessity of utilizing multi-extremity MRI in conjunction with lymphoscintigraphy to accurately map anomalous lymphatic phenotypes. Furthermore, it demonstrates that a robust, combined conservative approach utilizing collagen dressings for acute ulceration and heat therapy for chronic volume reduction can successfully manage limb-threatening complications and improve long-term patient outcomes.

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