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Male genital lymphedema: clinical features and management in 25 pediatric patients☆,☆☆

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ABSTRACT

Purpose: Genital lymphedema in the pediatric population is poorly understood. The purpose of this study was to determine the epidemiology, morbidity, and treatment outcomes for males with genital lymphedema.
Materials and methods: Male patients with genital lymphedema evaluated at our vascular anomalies center between 1995 and 2011 were reviewed. Etiology, age-of-onset, location, morbidity, and treatment were analyzed.
Results: Of the 3889 patients with vascular anomalies, 25 (0.6%) had genital lymphedema: 92% (23/25) with primary and 24.0% (6/25) with familial/syndromic lymphedema. For primary disease, the mean age-of-onset was 4.5 ± 6.3 years with 60.9% (14/23) presenting in infancy, 13.0% (3/23) in childhood, and 26.1% (6/23) in adolescence. Combined penoscrotal lymphedema was identified in 72.0% (18/25) of patients; 19 children (76.0%) had concomitant lower extremity involvement. The most common complication was cellulitis (24.0%). Surgical contouring was performed in 44.0% (11/25) of patients. Patients with operative intervention and follow-up ($n = 6$) had sustained improvement after a median of 4.2 years (range: 0.3–11.0).
Conclusions: Lymphedema of the male genitalia is typically idiopathic. Most patients develop swelling in infancy but can present in adolescence and occasionally childhood. The penis and scrotum are usually both involved and concurrent lower-extremity swelling is common. Surgical debulking can improve symptoms and appearance.

Lymphedema is chronic swelling of tissue owing to poor lymphatic function from either anomalous lymphatic development of (*primary*) or injury to (*secondary*) lymphatic nodes or vessels. Affected tissue initially swells from accumulation of subcutaneous lymph; resultant inflammation stimulates adipose deposition and fibrosis causing further enlargement over time [1]. Primary lymphedema is uncommon; it affects 1.2 per 100,000 persons less than 20 years of age [2]. Secondary lymphedema affects the majority (90%) of patients, principally the lower extremities of adults, owing to infection or treatment for malignancy [3]. In developing countries, filarial infestation is the most common cause of genital lymphedema [3]. The prevalence of primary genital lymphedema is unknown. Complications of lymphedema include infection, functional disability, and chronic cutaneous changes. Disfigurement from lymphedema, particularly of the genitalia, can cause considerable psychosocial distress.

Genital lymphedema in the pediatric population is not only infrequent but also poorly understood. Previous reports on genital lymphedema have been limited by small numbers of patients [4–12] and/or are comprised largely of adults [13–19]. Conflicting opinions on the utility of compression therapy [10,14,16,20] and timing of operative interventions (early versus post-pubertal) exist [4,7,10,14,16]. The purpose of this study was to determine the epidemiology, morbidity, and treatment outcome for males with genital lymphedema in an effort to improve patient counseling and offer management options for practitioners.

1. Materials and methods

After approval by the Committee on Clinical Investigation at Boston Children's Hospital, the Vascular Anomalies Center database was queried for all male patients with “lymphedema” of the penis and/or scrotum who presented to our center between January 1995 and January 2011. Records were retrospectively reviewed for referring diagnosis, age at presentation and intervention, anatomic location, morbidity, treatment modalities, and outcome. Diagnosis was made by clinical history, physical examination, and/or imaging studies. Age-of-onset was categorized based on physiological age:

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infancy (<12 months), childhood (1–9), and adolescence (10–21 years) [21–23]. Data are presented as mean ± standard deviation.

Excisional debulking is utilized in our center for lymphedema of the penis and scrotum. The procedures were performed by two surgeons in the following manner. The genitalia are prepared and draped widely and a urinary catheter of the largest size that can be easily inserted is placed. For penile lymphedema, a circumferential incision is made in the penile skin several millimeters proximal to the corona of the glans penis, using the scar from previous circumcision, if relevant. The skin of the shaft of the penis is then dissected just below the dermis circumferentially to the base. The entirety of the shaft skin is unfurled proximally using blunt, sharp, and/or cautery dissection. The underlying lymphedematous adipose tissue is then dissected off Buck's fascia circumferentially. Care is taken to avoid injury to the skin, neurovascular bundles, or urethra. Once the lymphedematous tissue is removed, the redundant skin is advanced back down the penile shaft. Excess circumference of skin is resected to leave just enough to comfortably wrap the penile shaft. When possible, the least abnormal skin is saved for penile coverage, discarding as much of the skin containing vesicles, dimples, or fibrosis. The skin is approximated to create a longitudinal scar at the ventral midline. The excess length of skin is excised to facilitate reapproximation at the distal pre-coronal incisional location. A small round closed-suction drain is placed under the skin flap, positioned dorsally away from the urethra and brought out in a suprapubic position. Interrupted intradermal absorbable sutures followed by a running subcuticular suture are used for closure. A penile block with 0.25% bupivacaine is performed prior to dressing the wound. The penis is gently wrapped with circumferential gauze, leaving the urinary catheter in place for 24 hours.

For scrotal lymphedema, a lenticular incision is made vertically encompassing the median raphe or transversely at the most dependent portion of the scrotum. The testes and spermatic cord structures are identified and protected. A full thickness wedge of scrotal tissue, encompassing skin, adipose tissue and dartos is excised. If hydroceles are present, the tunica vaginalis is similarly excised. The excess scrotal septum is resected and the testes are secured with non-absorbable sutures to the reduced septum. Scrotoplasty is performed to create a new midline raphe or transverse closure parallel to the rugae. In cases of extreme scrotal enlargement, additional redundant tissue is resected from the lateral aspect of each hemi-scrotum and closed in a V-Y advancement fashion. Closure is accomplished with multiple layers of absorbable suture in the tunica vaginalis, dartos, subcutaneous tissue, and skin. Post-operative scrotal elevation is employed for 1 week and patients are encouraged to wear a compressive undergarment thereafter.

2. Results

Of the 3889 male patients with a vascular anomaly, 25 (0.6%) had lymphedema of the genitalia. Primary lymphedema occurred in 92.0% (23/25) of patients and six children (24.0%) had syndromic or familial lymphedema [Noonan syndrome ($n = 2$), lymphedema–distichiasis ($n = 2$), and Milroy disease ($n = 2$)] (Table 1). Two patients (8.0%) had secondary lymphedema from operative intervention for undescended testes ($n = 1$) or non-infectious granulomatous disease ($n = 1$). Average age of onset was 4.5 ± 6.3 years (range: 0 to 15). Fourteen (60.9%) patients presented in infancy. Childhood onset occurred in 13.0% (3/23) of patients; the remaining 26.1% (6/23) presented during adolescence. The referring diagnosis was accurate in 64.0% (16/25) of patients; the most common erroneous diagnosis was capillary-lymphatico-venous malformation (Klippel-Trenaunay syndrome). Diagnosis was made by history and physical examination in almost all patients. No adjunctive imaging studies were performed in 8.0% (2/25) of children. MRI [72.0% (18/25)] and/or lymphoscintigraphy [36.0% (9/25)] were reviewed when available; of these studies, most were obtained at prior institutions or primarily for evaluation of non-genital anomalies. Pathologic specimens were reviewed in addition to imaging and clinical evaluation in nine (36.0%) children.

Penoscrotal lymphedema was identified in 72.0% ($n = 18$) of patients. Isolated scrotal and penile disease was found in 24.0% ($n = 6$) and 4.0% ($n = 1$) of patients, respectively. Concomitant involvement of the lower extremities occurred in 76.0% ($n = 19$) of children. Genitourinary symptoms were observed in 60.0% ($n = 15$) of patients and included hematuria [12% ($n = 3$)], altered urinary mechanics [8.0% ($n = 2$)], and phimosis [8.0% ($n = 2$)] (Table 2). Additional morbidities included cellulitis [24.0% ($n = 6$)], lymphorrhea [24.0% ($n = 6$)], and skin changes [12% ($n = 3$)].

Treatment was administered to 72.0% (18/25) of patients; seven (28.0%) children were observed (Table 3). Compression therapy was prescribed to 12 patients (48.0%). Of the patients receiving compression therapy, four (33.3%) later received surgical debulking. Contouring to improve symptoms and/or the appearance of the genitalia was performed in 44.0% ($n = 11$) of patients with a mean of 1.5 ± 1.0 procedures per patient. One patient underwent concomitant debulking of a lymphedematous lower extremity. Initial operative intervention occurred at average 13.9 ± 8.0 years of age. Post-operative complications ($n = 2$) included the appearance of granulomatous tissue along a penile shaft suture line that improved with time and a superficial wound infection treated with antibiotics. Post-operative follow-up was available for 54.5% (6/11) of patients with surgical procedures at a median of 4.2 years (range: 0.3–11.0). All patients with operative treatment and follow-up had sustained improvement.

3. Discussion

Genital lymphedema is an uncommon but troubling cause of deformity of the male genitalia. The epidemiology and morbidity are unclear in part owing to the rarity of the condition. Furthermore, diagnostic criteria, timing of therapies, and surgical techniques have been debated. To our knowledge, this is the largest series of male pediatric patients with genital lymphedema.

The etiology of genital lymphedema in children in the United States is predominately idiopathic. While most patients develop symptoms in infancy or early childhood, the diagnosis is often delayed for several years. Lymphedema is most commonly confused with other vascular anomalies, particularly if the lower extremities are also affected. An accurate diagnosis is imperative in providing appropriate

Table 1
Characteristics of genital lymphedema in 25 male pediatric patients.

Etiology	Primary	92.0%	(23/25)
	Familial/syndromic	24.0%	(6/25)
	Secondary	8.0%	(2/25)
Age of onset	Infancy	60.9%	(14/23)
	Childhood	13.0%	(3/23)
	Adolescence	26.1%	(6/23)
Location	Genital		
	Combined penile and scrotal	72.0%	(18/25)
	Isolated scrotal	24.0%	(6/25)
	Isolated penile	4.0%	(1/25)
	Associated lower extremity involvement	76.0%	(19/25)

Table 2
Complications of genital lymphedema in 25 male pediatric patients.

Cellulitis	24.0%	(6/25)
Lymphorrhea	24.0%	(6/25)
Skin changes	12.0%	(3/25)
Hematuria	12.0%	(3/25)
Pain	8.0%	(2/25)
Bleeding	8.0%	(2/25)
Altered urinary mechanics	8.0%	(2/25)
Chylous reflux	8.0%	(2/25)
Phimosis	8.0%	(2/25)
Dysuria	4.0%	(1/25)