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Vascular anomalies of the male genitalia☆,☆☆

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Abstract

Purpose: Vascular tumors and malformations of the male genitalia can affect urinary, sexual, reproductive, and emotional function.

Methods: Male patients with a genital lesion evaluated or treated at our center from 1995 to 2010 were reviewed to analyze presentation, diagnosis, treatment modalities, and outcome.

Results: Of the 3889 male patients, 117 had a vascular anomaly of the genitalia: 12 tumors and 105 malformations. The referring diagnosis was accurate in 72.7% of patients with a tumor, whereas 46.3% of malformations were misdiagnosed. Tumors included infantile hemangioma (n = 10) and kaposiform lymphatic anomaly (n = 2). Common vascular malformations were lymphatic (n = 46), venous (n = 33), and capillary-lymphatic-venous (n = 16). Presenting signs for tumors included ulceration (33.0%) and ambiguous genitalia (25.0%). Malformations manifested with swelling (40.0%), fluid leakage (16.2%), and pain (16.2%). Treatment was necessary for 69.9% (79/113) of patients. The remaining lesions (34/113) were observed. Tumor management included observation, pharmacotherapy, and excision. Malformations were largely treated with sclerotherapy and/or surgical procedures.

Conclusions: Vascular anomalies of the male genitalia are uncommon and frequently misdiagnosed. Accurate diagnosis can be made and appropriate treatment can be instituted based on presentation, natural history, and radiographic imaging. Observation and pharmacotherapy are the mainstays of tumor management. Malformations require sclerotherapy and/or resection. Interdisciplinary care optimizes outcomes for males with these often-disfiguring vascular lesions.

Vascular anomalies are disorders of blood vessels that usually present in infancy and childhood. They are divided into 2 groups, tumors and malformations, based on biologic and clinical behavior (Table 1) [1]. Tumors have a proliferating endothelium, whereas malformations exhibit normal endothelial cell turnover. Vascular anomalies, particularly those involving the male genitalia, cause considerable distress. These lesions can adversely impact a patient's self-

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image, inflict pain, and alter function. Accurate diagnosis is essential to ensure proper treatment and to optimize outcomes for these potentially life-altering anomalies.

1. Materials and methods

1.1. Study population

After approval by the Committee on Clinical Investigation of Children's Hospital Boston, the Vascular Anomalies Center (VAC) database was culled for all male patients with a vascular anomaly of the genitalia who presented to our center from 1995 to 2010. Patients were included if the lesion involved the penis, scrotum, urethra, and/or prostate. Records were retrospectively reviewed to study presentation, anatomical location, symptoms, referring diagnosis, VAC diagnosis, imaging, treatment modalities, and outcome.

1.2. Statistical analysis

Statistical analysis of results was performed using the Mann-Whitney U tests for comparison between tumors and malformations. Differences were considered significant when $P < .05$. Calculations were performed in GraphPad Prism version 16.0 (GraphPad Software, Inc, La Jolla, CA).

2. Results

2.1. Tumors and malformations

Of the 3889 male patients registered with a vascular anomaly, only 117 (3.0%) had a lesion of the male genitalia. Tumors comprised 10.3% (n = 12) and malformations 89.7% (n = 105) of the study population (Table 2). Tumors presented in early infancy (0.1 ± 0.1 years) compared with malformations (2.8 ± 5.0 years) ($P < .001$). The referring diagnosis was accurate in 72.7% of tumors and only 53.7% of malformations. Tumors were either infantile hemangioma (IH) (n = 10) or kaposiform lymphatic anomaly (KLA) (n = 2). Slow-flow malformations (n = 99) were more numerous than fast-flow lesions (n = 6). The most frequent slow-flow anomalies were lymphatic (LMs; n = 46), venous (VMs; n = 33), and capillary-lymphatic-venous (CLVMs; n = 16) malformations. The fast-flow anomalies were arteriovenous (n = 2), capillary malformation (CM)–arteriovenous (n = 2), and capillary-lymphatic-arteriovenous (n = 2).

The external male genitalia, penis or scrotum, was involved in all patients. Tumors were limited to the external genitalia. Malformations extended into the urethra (n = 10), bladder (n = 6), and prostate (n = 1). Involvement or extension beyond the genitalia occurred in 50.0% of tumors and 75.2% of malformations. Presenting signs for tumors included ulceration (33.0%), ambiguous genitalia (25.0%), and bleeding (16.7%). Additional findings in the subgroup of patients with perineal IH included cloacal exstrophy, microphallus, cryptorchidism, and infantile hepatic hemangiomas.

Vascular malformations presented with swelling (40.0%), pain (16.2%), fluid leakage (16.2%), bleeding (14.3%), and infection (10.5%). More specific genitourinary symptoms were hematuria (n = 15), chylous reflux (n = 8), altered urinary mechanics (n = 6), dysuria (n = 5), cryptorchidism (n = 4), bladder outlet obstruction (n = 3), depression (n = 3), erectile dysfunction (n = 2), and ambiguous genitalia (n = 1).

2.2. Diagnosis

The diagnosis of a vascular anomaly was made in 25.6% (n = 30) of patients on the basis of clinical history and physical examination. Radiographic studies were used for diagnostic clarification in 57.3% (n = 67) of cases. Histopathology was reviewed in 17.1% (n = 20) of patients.

Table 1 International Society for the Study of Vascular Anomalies classification

Tumors	Slow-flow malformation	Fast-flow malformation
IH	Capillary	Arteriovenous fistula
Congenital hemangioma	Lymphatic	Arteriovenous
Rapidly involuting congenital hemangioma	Microcystic	
Noninvoluting congenital hemangioma	Macrocystic	
	Lymphedema	
KHE	Venous	
KLA		
Pyogenic granuloma	Combined	Combined
	CLVM	CM-AVM
	Capillary-venous	Capillary-lymphatic-arteriovenous malformation
	Lymphatic-venous	