

VESICAL HEMANGIOMA IN PATIENT WITH KLIPPEL-TRENAUNAY-WEBER SYNDROME

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ABSTRACT

Klippel-Trenaunay-Weber syndrome is characterized by cutaneous hemangiomas, varicosity and bony hypertrophy of extremities. Urinary tract hemangiomas may occur in 3 to 6% of these patients. This work intends to report a case of a patient with a huge vesical hemangioma, who presented this syndrome.

A 5 year-old boy with Klippel syndrome sought our services due to 3 episodes of gross hematuria in the past 30 days. Excretory urography and computerized tomography were performed, indicating the presence of a swelling in vesical dome. An exploratory cystotomy was conducted and the dark colored mass in vesical dome was excised by partial cystectomy. The histopathologic finding confirmed that it was a vesical hemangioma. Though rare, urinary tract hemangiomas must always be considered in patients with Klippel-Trenaunay syndrome.

Key words: bladder; hemangioma; Klippel-Trenaunay-Weber syndrome; therapeutics
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INTRODUCTION

The Klippel-Trenaunay-Weber syndrome is a syndrome due to vascular malformations, characterized by cutaneous hemangiomas, varicosities and bony hypertrophy of extremities (1). Lesions are present at birth and about 75% of patients manifest symptoms before 10 years old (2). In 3 to 6% of patients, there are urinary tract hemangiomas (3). This work intends to report the case of a patient with this syndrome who presented hematuria caused by a vesical hemangioma.

CASE REPORT

Male, 5 year-old patient, who presented Klippel-Trenaunay-Weber syndrome, sought the urology service due to 2 episodes of gross hematuria in

the past 30 days. Apart from characteristic syndromic changes (cutaneous hemangiomas in lower limbs, hypertrophy of right lower limb and varices in lower limbs), the patient did not present any other relevant finding on physical examination. He was submitted to an ultrasonography of the urinary tract, which revealed a huge mass in vesical dome, with characteristic signs of hemangioma. An excretory urography was made, showing a large filling defect. The assessment was complemented by abdominal and pelvic computerized tomography, which showed a 4.0 x 3.5 cm mass in vesical dome (Figure-1).

Following the analysis of tests, it was decided to perform a partial cystectomy. During exploratory cystotomy, a dark colored mass was seen in vesical dome (Figure-2). The partial cystectomy was made with a safety margin. The histopathologic finding confirmed that it was a vesical hemangioma.



Figure 1 – Pelvic computerized tomography evidencing a large mass in the vesical dome area (a, b).

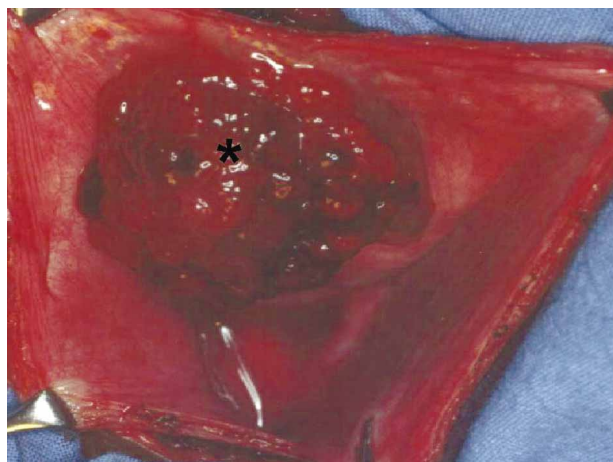


Figure 2 – Photograph of swelling situated in vesical dome (*), during performance of partial cystectomy.

DISCUSSION

In Klippel-Trenaunay-Weber syndrome, symptoms in genitourinary system occur in the illness' more severe forms, mainly in patients with visible vascular lesions in trunk and pelvis (2). One of the most frequent symptoms in these patients is hematuria, due to hemangiomas.

Vesical hemangiomas have an incidence of less than 1% of the organ's primary tumors, being, however, the most common connective tissue benign tumor in bladder (3). Painless gross hematuria is a characteristic of this type of tumor, as it was observed in the present case (2).

In Klippel-Trenaunay-Weber syndrome, vesical hemangiomas are frequent and preferably located on the bladder's anterior wall and vesical dome (2). Several treatments are proposed for this lesion: fulguration or endoscopic excision (2), treatment with YAG-Laser (3) and partial cystectomy. Endoscopic excision can lead to profuse bleeding,

and is contra-indicated (3), laser treatment is expensive and unavailable in many centers, which makes partial cystectomy the treatment best suited for this type of tumor.

Patients with Klippel-Trenaunay-Weber syndrome who present hematuria must be investigated, as they can have urinary tract hemangiomas.

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