Vesical Hemangioma in a Patient with Klippel-Trenaunay-Weber Syndrome

A 3-year-old child with Klippel-Trepanauy-Weber syndrome (KTWS), was admitted to our hospital with recurrent episodes of hematuria. Subsequent clinical history revealed 2 episodes of hematuria some weeks before, not initially mentioned by the parents. Urine analyses were negative for erythrocytes and proteinuria. The child manifested a normocytic anemia (hemoglobin, 9.9 g/dL; mean corpuscular volume, 74 μm^3; iron, 13 $\mu g/d$). Coagulation tests were normal; her D-dimers were 2060. An ultrasound examination of the urinary tract demonstrated a giant mass in the vesical dome (**Figure 1**). Magnetic resonance imaging revealed a complex vascular malformation at the left leg. Additional to this finding, a large vesical vascular malformation of 4.7 × 2.4 cm was visualized (**Figures 2** and **3**; available at www.jpeds.com). After multidisciplinary counseling, conservative treatment of close follow-up was chosen as therapy; during 6 years of follow-up, she has had transabdominal ultrasound and clinical examinations.

KTWS is a rare disorder characterized by capillary, venous and lymphatic, nonmalignant malformations that affects 1 in 30 000 live births. The syndrome consists typically of cutaneous capillary malformations, varicose veins, and bone and soft tissue hypertrophy of the affected extremity, leading to limb enlargement.[1] In 95% of cases, these 3 signs are limited to a lower limb.[2] Seventy-five percent of patients present with symptoms before the age of 10 years. Urinary tract involvement is reported in up to 23% of patients with KTWS.[3] The syndrome was described for the first time in 1900 by Klippel and Trenaunay as "nevus varix osteohypertrophique." In 1907, Weber described a similar syndrome, but with the presence of large arteriovenous fistulae.[4,5] Some authors consider the 2 syndromes as separate conditions. Tian et al identified VG5Q, an angiogenic factor, as an agent in the pathogenesis of KTWS.[6,7] Gene mutations that upregulate the VG5Q expression lead to increased angiogenesis. Contrary to previously held beliefs, urogenital manifestations of KTWS are not rare.[8] In 1971, Lindenauer reported a high frequency of hematuria of unknown cause in patients with KTWS.[9] Hemorrhagic shock in a 2-year-old child with a bladder hemangioma emphasized that early diagnosis is important to identify patients at risk.[10]

In cases of moderate and infrequent episodes of hematuria, a conservative approach (observation or bladder irrigation) is recommended.[1,6] When conservative management is insufficient, a more invasive approach is considered. Partial cystectomy is the current standard therapy for benign bladder tumors when located extratrigonally. Because of the significant morbidity (reduced storage function of the bladder, impaired voiding function) that is associated with this surgical intervention, less invasive treatment options have been proposed with their specific limitations and risks of complications. Good results have been reported with laser photocoagulation, which is a feasible treatment for smaller lesions.[4,6,10,11] Radiotherapy is not invasive, but has a high associated morbidity. Transurethral resection carries the risk of excessive bleeding and selective embolization of the internal iliac arteries can result in rapid collateralization.[1,4] ■

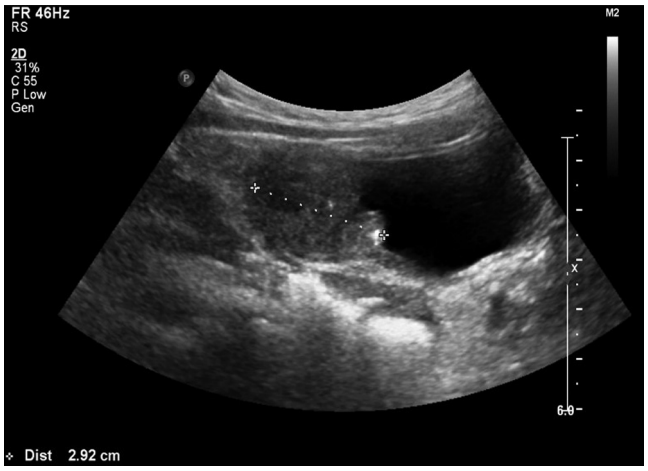

Figure 1. Transabdominal ultrasound image shows a large hyperechoic mass arising from the dome of the bladder.

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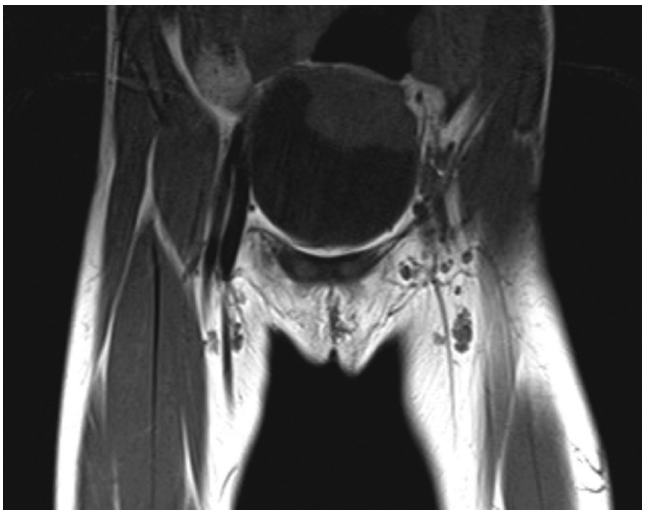

Figure 2. A T1-weighted coronal magnetic resonance image shows a lobullary, isointense mass in the vesical dome.

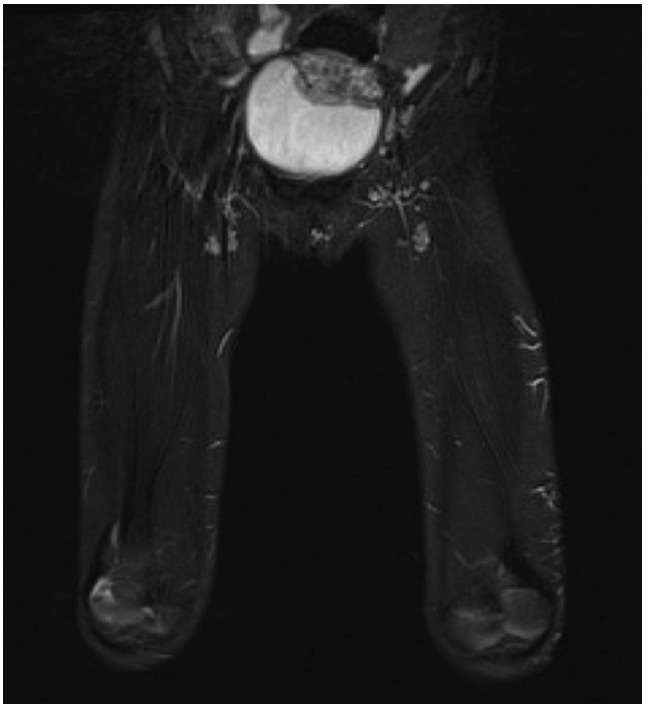

Figure 3. A T2-weighted inversion and T1-weighted recovery magnetic resonance image revealing a large mass in the vesical dome.