A Mild and Rare Form of Klippel-Trenaunay Syndrome Presenting With Urethral Bleeding Due To Penile Hemangioma

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Klippel-Trenaunay syndrome (KTS) is characterized by a triad of cutaneous port-wine capillary malformations, hemihypertrophy, and varicose veins. Intermittent gross painless hematuria is usually the first clinical sign. An 8-year-old boy with multiple hemangiomas, including glans penis, and associated with KTS presented with urethral bleeding. Radiologic and endoscopic evaluation revealed neither intra-abdominal nor intravesical hemangioma. Urethral bleeding was thought to be related to glanular hemangioma extending to the anterior penile urethra. Although we were able to manage the case conservatively, many patients require endoscopic or surgical interventions. Radiologic and endoscopic evaluations and careful follow-up is essential for diagnosis and prompt treatment.

CASE HISTORY

An 8-year-old boy was admitted to our emergency department several times because of intermittent episodes of urethral bleeding. Hematuria resolved spontaneously after insertion of a urethral catheter. Bleeding through external urethral meatus appeared one day after removal of the catheter and resolved by compression of the glans penis. The child had been diagnosed with KTS at 5 years of age and had no medical or surgical treatments.

Physical examination revealed a macular, telangiectatic vascular hemangioma of “port-wine” variety, present in a diffuse distribution over the entire right lower extremity, right side of trunk, and especially the cheek and upper lip (Figure 1). The lesion was found to be expanding over the right gluteal region and posterolateral of the right thigh and involving the glans penis, causing a curvature (Figure 2). The patient had been circumcised at 2 years old. Brownish-black angiokeratomatous nodules were distributed over the underlying hemangioma and both lower extremities. Congenital pes planus deformity was present on the right affected side. A right hemihypertrophy was definitely present (Figure 3).

Imaging modalities such as computed tomography and magnetic resonance imaging showed no intra-abdominal or intrapelvic abnormality. Endoscopy was performed to exclude a urethral or bladder hemangioma. Cystoscopy revealed no hemangioma or active bleeding in the bladder or posterior urethra. There was inner extension of glanular hemangioma to the anterior penile urethra. Because it was very small in size, we decided to follow up conservatively. At the last follow-up, the patient was asymptomatic and the size of hemangioma remained stable.

COMMENT

Since KTS was described in 1990, many patients with bleeding in the genitourinary or lower gastrointestinal tract have been reported.1-8 Three components of KTS are hemangioma/port-wine stain, varicose veins, and
hemihypertrophy of bone and soft tissues. In addition, orthopedic anomalies, such as polydactyly, syndactyly, and scoliosis are noted in the literature. Our case presented all main components of KTS and was associated with pes planus deformity.

Hemangioma and varicose veins, which are the major components of KTS, are usually responsible for manifestations. According to Servelle et al., the dysplastic posterolateral vein that drains the lower extremity overloads the pelvic veins. Because of this circulatory overload, the collateral veins of this dilated internal iliac system (vesical, genital, and rectal veins) are not able to drain normally. Dilated varicose veins appear on the bladder, internal genital organs, and the rectum. These large varicose veins may rupture into the bladder, vagina, or rectum. Painless intermittent hematuria and rectal bleeding are usually the first clinical signs. The present case was admitted to the emergency department because of urethral bleeding. Lower gastrointestinal tract bleeding was not observed.

Bladder hemangiomas are generally reddish-blue in color and sessile, pedunculated, lobulated, or flattened lesions that are commonly located on the anterior wall and dome of the bladder. Partial cystectomy and other endoscopic methods are treatment alternatives. Fortunately, we did not detect any hemangioma or abnormality in the bladder during cystoscopy.

Urethral hemangiomas are the other cause of hematuria or urethral bleeding in patients with KTS. They may be related to other congenital disorders such as systemic angiomatosis and Sturge-Weber syndrome. The treatment modalities are selected according to the size, number, and site of involvement of the hemangioma and the presence and severity of symptoms. Total excision or transurethral resection, fulguration, arterial embolization, and photo-ablation with laser and sclerotherapy are treat-
All of these aggressive interventions have the potential for significant complications, such as urethral stricture, impotence, and massive hemorrhage. Because our case was stable and associated with relatively mild symptoms, we treated him conservatively.

In conclusion, patients with KTS present multiple systems manifestation. A multidisciplinary approach is essential for prompt treatment and follow-up of this group of patients.

References