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Renal arteriovenous malformation in a 14-year old girl with gross hematuria

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ABSTRACT

Renal arteriovenous malformation (AVM) is a rare cause of gross hematuria in children and adolescents that may present so acute and severe hematuria, leading to significant anemia in patients. We present a 14-year old girl with acute massive gross hematuria with no history of trauma or coagulation defect. Although AVM not common, it should be included in the differential diagnosis of gross hematuria in children and adolescents.

Keywords: Hematuria; Arteriovenous malformation, IgA nephropathy, Urinary stone, Loin pain-hematuria syndrome

Case Report

Implication for health policy/practice/research/medical education:
Trauma-unrelated hematuria rarely causes significant anemia and hemodynamic changes. Therefore, when it occurs, renal vascular malformation should be considered in the differential diagnosis.

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Introduction

Gross hematuria is defined by the presence of an increased number of red blood cells (RBCs) in the urine that is visible to the naked eye. Despite its uncommon occurrence in children, it is a disturbing finding for both the affected child and the family. General practitioners and pediatricians are first-line persons encountering hematuria in children (1).

The clinician generally is able to establish the underlying etiology by a complete history, physical examination, and urinalysis.

The most commonly identified etiologies for gross hematuria in children include IgA nephropathy, hypercalciuria, urethrorrhagia and hemorrhagic cystitis (2).

Vascular anomalies are rare causes of gross hematuria. Renal arteriovenous malformations (AVMs) are abnormal communications between the intra-renal arterial and venous systems. They cause hematuria and are associated with hypertension. Renal AVMs may be either congenital or acquired (often iatrogenic). Congenital arteriovenous malformations usually present with hematuria, while acquired fistulas usually present with hemodynamic changes such as hypertension, cardiomegaly and congestive heart failure (3). More commonly, the renal AVMs refers to the congenital type of malformation such as our case.

Case Presentation

A 14-year-old girl with a past medical history of 2 weeks, gross hematuria and clot retention was referred to our hospital. Other symptoms were nausea, vomiting, and fever. Hematuria deteriorated during the three days before referral. Patient was evaluated in the urology emergency ward. Then, due to severe anemia, she was referred to pediatric hematology/oncology division and consulted with pediatric nephrologist in our hospital.

She had no history of trauma, recent surgery, bleeding disorders, or consumption of any drug. There was no family history of bleeding disorders.

In physical examination, the patient showed severe pallor. Blood pressure at the time of presentation was 120/70 mm Hg and heart rate was 96 per minute. Her abdomen was soft without any distention or organomegaly.

Initial laboratory data were as follows; hemoglobin = 2.8 g/dL, creatinine = 0.9 mg/dL, BUN = 12 mg/dL, Na = 143 mEq/L, K = 3.6 mEq/L, total bilirubin = 2 mg/dl,

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direct bilirubin = 0.5 mg/dL, and urinalysis showed +3 blood and many RBCs.

Ultrasonography showed normal size kidneys with normal thickness of parenchyma and echo pattern. Additionally, mild hydronephrosis with an internal echo (hematuria) was seen in the right kidney. No stone or mass lesion was detected.

Bladder ultrasonography showed 10 ×18 mm lesion near the ureterovesical junction caused by the blood clot.

The patient received two units of packed RBCs. After improvement of weakness and pallor, color Doppler sonography and multi-slice abdominal computed tomography were conducted.

Color Doppler sonography showed a small vascular lesion in the right kidney (Figure 1).

In abdominal CT angiography, focal abnormal vascular enhancement (measured about 12 m × 15 mm) in the mid pole of the right kidney was seen, which is suggestive of AVMs (Figure 2). This diagnosis was confirmed by selective conventional renal vessels angiography (Figure 3). The patient was referred to an interventional radiologist for embolization of renal arteriovenous malformation.

Discussion

Hematuria is a common complaint of patients in pediatric nephrology or pediatric urology practice. The appearance of blood in the urine of adult patient, especially at ages over 50 years, is probably an alarming sign of urinary tract malignancy, which should be considered and ruled out at the first steps of the diagnostic approach. On the contrary, most of the causes (not all) of hematuria in children and adolescents are rather benign conditions such as urinary tract infection, urinary stone, and hypercalciuria and also post-infectious glomerulonephritis.

Rare causes of hematuria include hereditary hemorrhagic telangiectasia, radiation cystitis, schistosomiasis (not rare in endemic areas), AVMs, nutcracker syndrome, and the loin pain-hematuria syndrome.

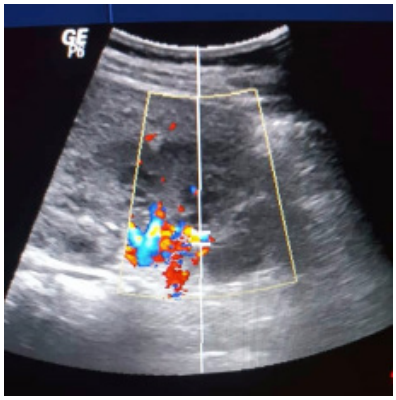

Figure 1. Color Doppler sonography showed aliasing in the middle region of right kidney in favor of AVM

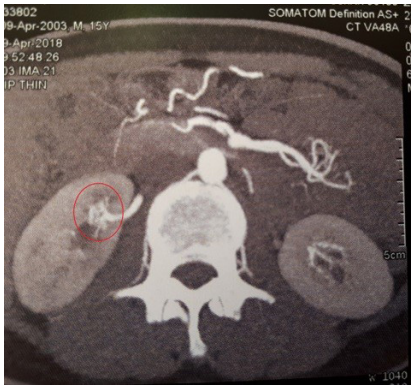

Figure 2. Multi slice spiral CT angiography of renal arteries showed 12*15 mm abnormal vascular lesion in middle portion of right kidney.

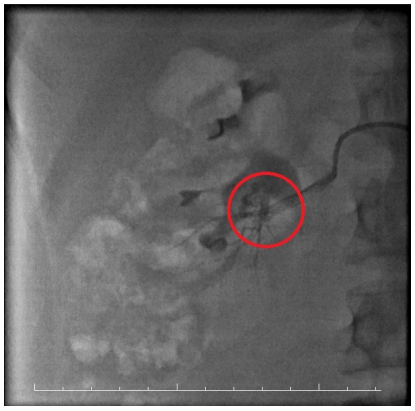

Figure 3. Selective renal artery angiography showed AVM of right kidney adjacent to pyelocalyceal system.

AVMs are abnormal communications between the intra-renal arterial and venous systems with an estimated prevalence of less than 0.04% (4,5).

Renal AVMs can be either congenital or acquired (often iatrogenic). AVM (in about 75% of cases) may be a consequence of renal biopsy, surgery, trauma, and pyelonephritis. These lesions appear in angiography as a single communication between a vein and an artery.

Congenital renal arteriovenous malformations (about 20% of cases) are specified in angiography as multiple communications between arteries and veins. They are usually located in the upper pole of the kidney; however, they also can be in the mid-point or the lower pole (6).

Individuals with renal arteriovenous malformation usually appear with microscopic hematuria due to the rupture of small venules into the kidney collecting system from increased intravascular pressure (7). Other patients with AVMs may present with signs of congestive cardiac failure such that up to half of them have symptoms of cardiomegaly and high blood pressure (8). There are few cases of AVM presenting as gross hematuria with clot

formation reported in the medical literature.

Color Doppler ultrasonography is an accessible and noninvasive imaging modality for screening patients with AVMs (9). One of the disadvantages of ultrasonography is poor sensitivity to distinguish AVMs from the normal kidney artery and vein (10). A high-flow lesion with possible pulsatility in the downstream drainingrenal vein is suggestive of AVMs.

In abdominal and pelvic computed tomography (CT) imaging, a well-marginated renal lesion that enhances similar to the blood pool and early enhancement of the draining renal vein is diagnostic for kidney AVMs.

The gold standard diagnostic modality for visualization and treatment of renal vascular abnormalities is conventional angiography. This procedure can selectively opacify small arterial branches to detect normal vessels from abnormal vessels (9).

Embolization is usuallythe most common treatment of renal AVM (11). Surgery can be performed when embolization fails or if AVM is associated with malignancy.

Conclusions

In our case, after renal color Doppler ultrasonography, we performed abdominal CT angiography and then conventional selective renal angiography to confirm renal AVM. The patient was followed up and referred for embolization of AVM.

Authors' contribution

PA as resident of pediatrics collected some history of the patient and helped to prepare the primary draft. HEM was responsible for approach and diagnosis of patient, completed the history of the patient and drafting the paper. HEM also edited the final draft.

Conflicts of interest

The authors declared no competing interests.

Ethical considerations

Ethical issues including plagiarism, double publication, and redundancy have been completely observed by the authors. The parents of the patient gave his consent to publish as a case report.

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