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Case Report

Hematuria in a young severe hemophilia patient – A case report on a rare radiological finding

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ABSTRACT

Renal ectopia or ectopic kidney is one of the common renal anomalies in which there is faulty migration of the fetal pelvis during the embryonic period. Hemophilia is an inherited coagulation disorder characterized by recurrent and spontaneous bleeding due to a deficiency of clotting factor VIII or IX. Clotting factor concentrates are used as standard therapy for the management of bleeding episodes. Hematuria is the presence of blood in urine that is either visible to the naked eye (macroscopic) or nonvisible (microscopic). Common etiologies of hematuria include congenital anomalies, calculus, inflammation, and infectious and malignancy of the renal and urogenital tract. Here, we present a 14-year-old male with severe hemophilia A who presented with severe pain abdomen and hematuria. On evaluation, he was found to have a left-sided ectopic kidney. In addition to the overdue discovery of the left-sided ectopic kidney, there is the coexistence of a bleeding disorder which may pose a challenge in the patient's management.

Keywords: Severe hemophilia, Hematuria, Renal ectopia

INTRODUCTION

Renal ectopia refers to the malposition kidney due to faulty migration of the fetal pelvis during embryonic development. The most common location is pelvic followed by abdominal and lumbar position based on its position in the retroperitoneum. The incidence of the pelvic kidney is about one in 725 births. A simple ectopic kidney is usually silent. However, if malrotated there is a hazard of calculus formation with consequent hydronephrosis, leading to colicky pain and hematuria.[1] When associated with a coagulation disorder, pre-existing ectopic kidney might pose a challenge in management.

Hemophilia A is an X-linked hereditary bleeding disorder characterized by deficiencies or absence of functionally active Factor VIII.[2,3] There are very few case reports showing the coexistence of renal ectopia and hemophilia. Hematuria is a common clinical finding in a setting of such coexistence. Hence, specific history along with appropriate imaging plays a crucial role in the right diagnosis and proper treatment of complications, along with specific guidance for follow-up.[4]

CASE REPORT

A 14-year-old male presented with progressive pain abdomen for the past 7 days associated with reddish discoloration of urine for the past 2 days. The patient was diagnosed with severe

hemophilia A at 1 year of age and was on regular prophylaxis at the dose of 30 IU/kg recombinant Factor VIII (rFVIII) 3 times a week up to 10 years of age following which he developed high titer inhibitor and was thereafter put on emicizumab prophylaxis, along with injection rFVIIa for breakthrough bleeding. Inhibitor status was negative screened 4 weeks back. On naked eye examination, macroscopic hematuria was present without any evidence of target joint or organ bleeding. No previous history of hematuria was present and annualized bleeding rate was 2.

A complete hemogram showed hemoglobin 13.3 g/dL, total leukocyte count 4500/cu mm, differential counts within normal range, and platelet 1.6 lakh/cu mm. Biochemical workup revealed normal creatinine (0.7 mg/dL), normal serum electrolytes, and l liver function test. Urine routine examination revealed plenty of red blood cells and the urine culture was sterile. Ultrasonography of the whole abdomen showed a malrotated left kidney in the mid-pelvis. Contrast-enhanced computed tomography whole abdomen [Figure 1] was done which showed similar findings, along with swollen left kidney with peripheral inflammatory stranding. Delayed enhancement and retention of contrast were also seen in the left kidney suggestive of acute nephritis. The patient was infused with 100% factor correction with rFVIII for 5 days followed by 50% factor correction for 7 days, along with adequate hydration along with adequate hydration and supportive management after which his symptoms resolved.[3]

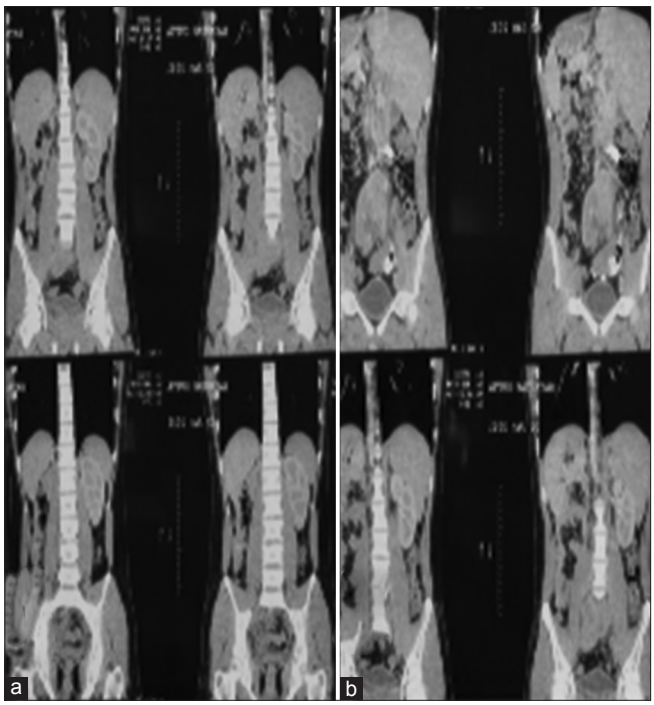

Figure 1: 14-year-old male with severe hemophilia A who presented with hematuria. (a) Contrast-enhanced computed tomography scan of the abdomen showed an ectopic kidney located in the left pelvic region. (b) There is also the presence of delayed enhancement and retention along with peripheral inflammatory changes in the left malrotated kidney.

DISCUSSION

Between the 6th and 9th weeks of intrauterine life, the kidneys ascend to the lumbar region just underneath the adrenal glands, along the dorsal aorta.[5] Ectopic kidney may rarely present as pain in the abdomen or macroscopic hematuria. In the present case, the left ectopic kidney was present in the mid-pelvis region.[6]

Young hemophiliacs presenting as isolated hematuria are rare. However, if present, as in the present case, ruling out local pathology is of utmost importance for proper management. Renal infection and calculus formation can worsen hematuria in a patient with an ectopic kidney in a background of coagulation disorder like hemophilia A.[7] It is also pertinent to properly investigate for any other associated congenital anomalies. The use of non-steroidal anti-inflammatory drugs should be restricted in such a situation as it causes nephropathy hence adequate intake of oral fluids with dietary counseling should be advised to avoid calculus formation.

CONCLUSION

Unilateral ectopic kidney in patients with hemophiliacs is rare. Clear knowledge of the association of renal anomaly with the bleeding tendency in hemophiliacs specially in young people is of utmost importance for proper management.

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Ethical approval

The research/study complied with the Helsinki Declaration of 1964.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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