CASE REPORT

A Rare Case of Idiopathic Vascular Renal Tumor Renal Hemangioma: A Case Report

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Renal hemangioma is a rare benign vascular tumor which can present with painful or painless gross hematuria. Its preoperative diagnosis is extremely difficult due to non-specific imaging findings and can also mimic other disease entity.
This is a case of a 31-year-old female who presented with recurrent gross hematuria with no antecedent cause. KUB ultrasound and CT stonogram showed insignificant findings of the cause of hematuria. Renal Angiogram revealed multiple vascular channels with arterial and venous connections at the upper pole of the right kidney with the impression of gross hematuria secondary to AV malformation. Simple Nephrectomy was done to address the hematuria wherein histopathologic findings were indicative of renal hemangioma. Patient did not report any episode of gross hematuria since the operation.

Key words: recurrent gross hematuria, renal hemangioma, vascular anomaly

Introduction

Hemangioma is a rare vascular tumor found in the urinary tract. The kidney is most frequently affected, followed by the urinary bladder, ureter, and urethra.[1] Diagnosis is often difficult and preoperative diagnosis is rare.[2] Internationally, approximately 200 cases were reported since this condition was first described by Virchow in 1867.[2] Literature review for local data only revealed a case of renal hemangioma in a pediatric patient reported by Dr. Caballes, et al. in 2018. There is no published data for adult cases locally. The authors report a case of gross hematuria due to idiopathic renal hemangioma. Their objective was to discuss its clinical presentation, emphasize the difficulty in preoperative diagnosis and the different treatment modalities.

The Case

A 31-year-old female was hospitalized due to recurrent gross hematuria. In 2012, she had her first episode of gross total hematuria with no other accompanying symptoms such as flank pain and fever. She denied any history of trauma nor any blood dyscrasia. She was initially managed as a case of urinary tract infection and was given unrecalled antibiotics. Afterwhich, she claimed to have spontaneous resolution of the hematuria. She was asymptomatic until 2019 wherein another spontaneous gross hematuria happened. Several imaging modalites were requested. KUB ultrasound was done which revealed unremarkable kidneys, urinary bladder was well-distended with echogenic intraluminal structure at the posterior aspect measuring 10.1cm x 9.6cm x 2.6cm with

the impression of hematoma to rule-out urinary bladder mass. CT stonogram was requested which showed non-obstructing nephrolithiasis on the right kidney measuring 0.2cm with Hounsfield unit of +417. Cystoscopy was done to evacuate intravesical blood clots. No gross mass lesion was seen. Spontaneous resolution of hematuria was reported for the second time. After 4 months, another episode of spontaneous gross hematuria occurred. This time, CT scan of the whole abdomen with contrast was done which revealed branching hyperdense lesion at the upper pole and interpolar region of the right kidney (Figure 1). She was tachycardic with a heart rate of 103. On laboratory work-up, her hemoglobin level was below normal at 74mg/dL. Hence, 3 units of packed red blood cell were transfused. Also at this time, renal angiogram was requested which showed two renal arteries and single renal vein in the right kidney with multiple intertwining vascular structures with venous and arterial connection in its upper pole suggestive of AV malformation. (Figure 2.) There was a single renal artery and single renal vein in the left kidney. Urinary bladder was noted to be filled with blood clots. Cystoscopy and evacuation of blood clots were rendered for the second time. The patient consulted at this institution and authors' initial impression was recurrent gross hematuria secondary to AV malformation of the right kidney. She underwent Simple Nephrectomy of the right kidney to address the hematuria. Histopathology was suggestive of renal hemangioma. The operation went well with no complication. Postoperatively, there was no reported complaint of any recurrence of gross hematuria within a 2-month follow-up as of this writing.

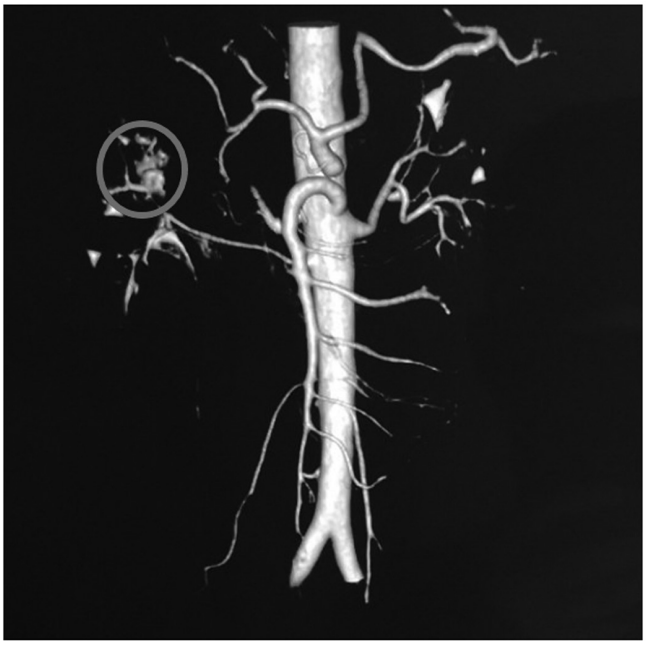

Figure 2. Multiple intertwining vascular structures with venous and arterial connection in the upper pole of right kidney measuring 1.5cm.

Discussion

Gross hematuria is one of the most common manifesting symptoms in urologic diseases. The usual causes of gross hematuria are urinary tract

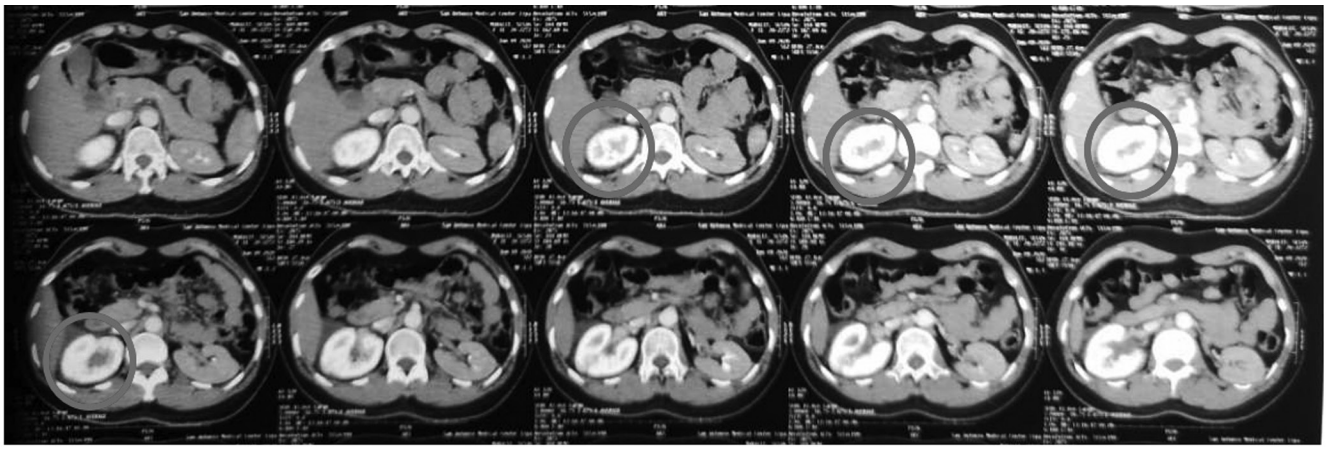

Figure 1. Branching hyperdense lesion at the upper pole and interpolar region of the right kidney.

infection, urinary tract calculi, genitourinary trauma and genitourinary malignancies. Rare causes are vascular anomalies such as hemangioma and arteriovenous malformation. In literature, these diseases account for 0.04% of cases.[3] Out of this 0.04%, 75% are traumatic in origin and 25% are idiopathic.[3] Idiopathic renal vascular anomalies are extremely rare, and which was found in the present case. Renal hemangioma usually affects young adult 30-40 years of age. It has no gender predilection. Hemangiomas are said to be benign vascular tumors that probably arise from embryonic remains of unipotent angioblastic cells.[4] Although renal hemangioma can be found in any part of the kidney, the most frequent location is the tip of the papilla. The submucosal region, the papilla, and the medulla account for 90% of anatomic locations.[4] In the present case, it was found at the upper pole of the right kidney. It has wide array of symptoms from asymptomatic microscopic hematuria to painless or painful total gross hematuria which can have hemodynamic consequences.[2] It is usually solitary and unilateral in 80% of cases and considered to be an isolated disease, but it can be associated with different syndromes such as Tuberous Sclerosis, Sturge-Weber syndrome, and Klippel-Trenaunay-Weber syndrome.[1] The size of a renal hemangioma ranges from 1 to 2 cm in diameter to as large as the kidney itself.[5] The one seen in the present case measured 1.5cm. The radiological features of this tumor are not well described in the literature and the findings are non-specific. Initial diagnostic imagings for cases of gross hematuria are ultrasonography and CT scan. Renal hemangiomas can present with normal findings to a nonspecific hyper or hypodensity on these modalities. Renal angiography is said to be the gold standard and is usually requested in cases of high suspicion of vascular anomalies with inconclusive cause of hematuria in initial imaging.[6] The findings on angiography ranges from a normal appearance to a hypervascular mass producing varying degrees of caliceal deformity and can be associated with rapid arteriovenous shunting.[6] Since, there are no specific clinical or radiological findings for the tumors, renal hemangiomas are often missed or can be mistaken with other disease entity such as other vascular anomalies or even renal cell carcinoma. Histologically, the tumors have a dense network of thick-walled vessels lined with typical endothelial cells with hemorrhages.[6] Management includes observation, nephrectomy, partial nephrectomy and embolization. In a healthy patient with mild to moderate hematuria who is clinically well, observation is not contraindicated.[1] There are reported cases of spontaneous resolution but this is reserved for lesions less than 1cm with no hemodynamic instability. Embolization has a wide range of 53-90% success rate for small lesions. Generally, surgery is considered when the tumor is large or it already causes symptoms such as life-threatening hemorrhage, or when it is difficult to distinguish the lesion from a disease such as carcinoma which requires nephrectomy.[1] Simple nephrectomy was done in the present case because the patient already has hemodynamic instability. The preference of doing partial nephrectomy versus nephrectomy depends on the size of the lesion and the preference of the surgeon. Renal hemangioma is seldom diagnosed preoperatively, and it is therefore difficult to recommend specific treatment.[4] To the authors' knowledge, this is the first report of renal hemangioma in a young adult in the local setting.

Conclusion

In conclusion, idiopathic renal hemangioma is extremely rare which can present with different clinical manifestations. What makes it more interesting is that the preoperative diagnosis of this disease remains difficult because it produces no particular clinical symptoms or images on common radiological studies such as sonography and CT scan. Renal angiography is the gold standard in diagnosing these cases. Management includes observation, nephrectomy, partial nephrectomy and embolization. Lastly, always keep in mind that when a patient presents with recurrent gross hematuria with no definite findings in imaging such as tumor or stones, hemangioma should be on the list of differential diagnoses.

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