Urology Case Reports 39 (2021) 101818

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Congenital renal arteriovenous fistula presenting with gross hematuria and its management

Mohammad Hatef Khorrami [a], Niloofar Javadi [b], Hossein Ebrahimi [c], Farbod Khorrami [d], Zahra Zandi [e,*]

[a] Department of Urology, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran
[b] Student Research Committee, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran
[c] Resident of General Surgery, Department of General Surgery, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran
[d] Department of Human Biology, University of Toronto, Toronto, ON, Canada
[e] Resident of Urology, Department of Urology, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran

ARTICLE INFO

Keywords:
Renal arteriovenous fistula
Hematuria
Percutaneous embolization

ABSTRACT

Renal arteriovenous fistula (AVF), an abnormal connection between artery and vein, results from development failure or following vascular injury. AVFs may cause various complications, i.e., secondary hypertension and hematuria. To manage AVF, it is recommended to bypass it from blood circulation surgically or by percutaneous embolization. The present study describes a woman with AVF, who primarily was managed percutaneously and then surgically.

1. Introduction

Renal arteriovenous fistula (AVF) is an abnormal extra-capillary link between artery and vein. AVF is a consequence of vascular development process failure, vascular injury due to tumor, trauma or iatrogenic.[1]

AVFs are usually symptomized by microhematuria or gross hematuria. Management of symptomatic AVFs is removing the lesion from blood circulation, either surgically or through percutaneous embolization. Untreated AVF may cause complications, such as hypertension, urinary obstruction or congestive heart failure.[1] In this article, we reported a case of AVF and its management.

2. Case presentation

A 37-year-old woman was referred to our urology center with permanent gross hematuria, first detected ten days ago. She also complained of dull pain in her left flank with no radiation. Hematuria occurred without pain, dysuria or frequency, and it was reported as a worm-shaped clot.

Her medical documents showed hypertension, which was resistant to three full-dose antihypertensive medications.

In physical examination, there was no noticeable finding except high blood pressure. Systolic and diastolic blood pressures were 165 and 95 mmHg, respectively.

As the first step in the diagnosis approach, renal and bladder ultrasonography was done. Due to an ultrasonography report which detected a suspicious malignant lesion in the inferior pole, the patient underwent an abdominopelvic computed tomography scan (CT scan) with/out intravenous contrast. CT scan reported left renal vein engorgement and enhanced multi-lobular mass without evidence of soft tissue fed from the left renal artery and drained to the left renal vein (video). Considering venous drainage was observed in the arterial phase of the CT scan, this mass was highly suspicious of AVF. No evidence of active bleeding and hematoma was detected.

Supplementary video related to this article can be found at https://doi.org/10.1016/j.eucr.2021.101818

Since the AVF diagnosis was confirmed with the CT scan, the patient was a candidate for angioembolization. During this procedure (video still), the implanted coil passed AVF because of its large size. As the angioembolization was failed, the patient underwent a left partial nephrectomy After successfully removing the left kidney inferior pole lesion, the bleeding continued despite the kidney's packing and suturing the incision. Due to persistent bleeding and clinical suspicion of the lesion remnant, the surgery team performed a total left nephrectomy.

* Corresponding author. Department of Urology, Alzahra Hospital, Shohadye Sofe St, Isfahan, Iran.
E-mail addresses: Khorrami386@gmail.com (M.H. Khorrami), Javadi.niloofar75@gmail.com (N. Javadi), Hosseinebrahimimed@gmail.com (H. Ebrahimi), Z.zandi70@gmail.com (Z. Zandi).

https://doi.org/10.1016/j.eucr.2021.101818
Received 6 August 2021; Received in revised form 18 August 2021; Accepted 20 August 2021
Available online 24 August 2021

Immediately after total nephrectomy, the previously elevated systolic blood pressure, despite consuming antihypertensive agents, dropped to the normal range amazingly.

3. Discussion

The failure of the vascular development process may reflect in arteriovenous fistula (AVF) or malformation (AVM). The anatomy of the feeder artery makes the difference between these two conditions; AVM is formed by complex arterial branches followed by tortuous vascular nidus. In contrast, AVF is built of a single artery connected directly into a dilated draining vein. The etiology of AVF is categorized into three groups: congenital, acquired and idiopathic. The congenital form is mostly seen in women, threefold of men, in their 30's and 40's—the acquired AVF results from penetrating trauma, tumor or iatrogenic procedures, e.g., renal biopsy. In case of an unknown significant cause, it is called idiopathic.[1]

As our case characteristics are similar to congenital characteristics, woman and 37 years old, and no previous history of mentioned causes of the acquired form was reported, the diagnosis of our case was congenital AVF.

Incidence of AVFs is rare; however, it's possibly underestimated because of a significant portion of asymptomatic cases. Various symptoms may be observed based on the size and the localization of the AVF. Congestive heart failure is a result of a large fistula that contains a considerable amount of blood. Hypertension is may associated with renal ischemia followed by activation of the renin-angiotensin pathway[2] and altered flow dynamics of the renal artery.[1] Gross hematuria is the most common symptom among symptomatic AVFs. Even bleeding of small AVF causes hematuria unless AVF is localized somewhere other than the pelvicalyceal system.[1]

Our case was presented with gross hematuria and resistant hypertension. In our case, hypertension occurred earlier than hematuria. Also, the patient suffered from dull pain in her flank due to urinary obstruction.

Several modalities are used to diagnose AVF, including ultrasonography, computed tomography scan, magnetic resonance imaging and digital subtraction angiography (DSA). DSA is the gold standard assessment; however, ultrasonography is the initial step. AVF is a hypoechoic lesion in B-mode; while, Doppler ultrasound demonstrates the AVF as a flow-filled lesion with the turbulent flow.[3] The protocol must include arterial and delayed venous phases when AVF or other vascular lesions were investigated with the CT scan. DSA is both diagnostic and therapeutic.

In our study, the US as the initial step showed a lesion with a vascular portion. We performed a triphasic CT scan to discriminate between vascular lesion and malignancy, which revealed the AVF at the inferior pole of the left kidney.

The goal of management is to save kidney function and removing AVF from blood circulation to eradicate symptoms simultaneously. AVFs found incidentally without any symptoms are managed conservatively. Since less invasive therapies are the first choice in management, percutaneous angioembolization with local anesthesia is the first-line unless the patient is unstable or inappropriate anatomically.[4] Based on the material used, embolization is divided into four types: gel foam, alcohol, coil, and liquid embolic. Complications or the need for a large number of coils cause angioembolization to be changed to surgical interventions, including partial and total nephrectomy.[1]

First, our case underwent percutaneous embolization with a coil. Because of the large size of AVF, the coil was passed the AVF and entered to the right atrium. Because of the complication, partial nephrectomy was chosen as the second step. After partial nephrectomy, despite performing adequate measures, e.g., packing, the bleeding continued. Out of necessity, a total nephrectomy was done. Immediately as surgery finished, hypertension was cured, and her blood pressure dropped to normal range. The pathological assessment revealed that the lesion was arteriovenous fistula.

4. Conclusion

AVFs are rare non-malignant vascular malformation; however, they may cause important complications. It should be listed as a differential diagnosis in different symptoms, i.e., hematuria and drug-resistant hypertension. Management can be divided into two procedures, percutaneous embolization and surgery.

Consent

A written informed consent was obtained from the patient.

Author statement

Mohammad Hatef Khorrami: Conceptualization, Writing - Review & Editing. Niloofar Javadi: Investigation, Writing - Original Draft. Hossein Ebrahimi: Visualization, Writing - Original Draft. Farbod Khorrami: Investigation, Writing - Original Draft. Zahra Zandi: Supervision, Writing - Review & Editing.

Declaration of competing interest

None.

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

References

1. Hatzidakis A, Rossi M, Mamoulakis C, et al. Management of renal arteriovenous malformations: a pictorial review. *Insights Imag*. 2014;5(4):523–530.
2. Sarramon JP, Cerene A, Gorodetski N, Bernadet P, Durand D. Spontaneous renal arteriovenous fistula and arterial hypertension — conservative treatment and healing. *Eur Urol*. 1978;4:214–216.
3. Cisternino SJ, Malave SR, Neiman HL. Congenital renal arteriovenous malformation: ultrasonic appearance. *J Urol*. 1981;126(2):238–239.
4. Gandhi SP, Patel K, Pandya V, Raval M. Renal arteriovenous malformation presenting with massive hematuria. *Radiol Case Rep*. 2015;10(1):1068.