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Congenital Renal Arteriovenous Malformation: A Rare but Treatable Cause of Hypertension

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Statistical Analysis C
Data Interpretation D
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Conflict of interest: None declared

Patient: Female, 29
Final Diagnosis: Renal arteriovenous malformation
Symptoms: Hypertension
Medication: —
Clinical Procedure: Angiography
Specialty: Cardiology

Objective: Rare disease

Background: Congenital renal vascular anomalies have been classified into 3 categories: cirroid, angiomatous, and aneurysmal. These classifications are based on the size, location, and number of vessels involved. Aneurysmal malformations, such as the one reported here, have a single (and dilated) feeding and draining vessel. The prevalence of renal AVMs is estimated at less than 0.04%, making them rare causes of secondary hypertension.

Case Report: A 29-year-old white woman was seen in the hypertension clinic as a referral from high-risk obstetric clinic for management of hypertension (HTN). A secondary hypertension workup with Doppler waveforms of the renal arteries revealed prominent diastolic flow in the left compared to the right. For confirmation, an MRA was done, which showed a large left renal upper-pole arteriovenous malformation (AVM) with associated vascular shunting and early opacification of the left renal vein. This congenital AVM was identified as the cause of her hypertension. Angiography and coil embolization were performed. The patient's BP normalized within a few days and she was taken off her antihypertensive medications.

Conclusions: This case illustrates that a careful review of duplex waveforms beyond just peak velocity and ratios is important to identify uncommon pathologies. This is important, as renal AVMs respond well to embolization, with resolution of hypertension in 59% of patients treated.

MeSH Keywords: Arteriovenous Malformations • Catheterization, Peripheral • Hypertension, Renovascular • Renal Artery

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Background

Congenital renal vascular anomalies are rare but potential causes of significant morbidity. They have been classified into 3 categories: cirroid, angiomatous, and aneurysmal [1]. These classifications are based on the size, location, and number of vessels involved. The prevalence of renal AVMs is estimated at less than 0.04%, making them rare causes of renovascular cause of hypertension. Despite the rarity of these lesions, they are treatable, with high rates of resolution of secondary hypertension. It is imperative that physicians ordering renal ultrasounds for secondary hypertension workups be aware of findings that may indicate the presence of these vascular lesions. This case illustrates the diagnosis and unique treatment of this rare etiology of renovascular hypertension.

Case Report

A 29-year-old white woman contemplating pregnancy was seen in the hypertension clinic as a referral from a high-risk obstetric clinic for management of hypertension (HTN). She had been diagnosed with HTN at age 19 and had been on antihypertensive medication (methyldopa 500 mg BID) with uncontrolled HTN. She had an extensive negative workup for secondary HTN, including pheochromocytoma, hyperaldosteronism, Cushing's, and thyroid disorders. A renal artery duplex study done elsewhere was reported as moderately increased left renal artery peak systolic velocity but normal renal-to-aortic ratio (ARR) not suggestive of renal artery stenosis. Our review of the images (Figure 1A) showed that the peak proximal left renal artery velocity was 225 cm/s with ARR of 1.3, which is within the normal limits (renal-to-aortic velocity ratios greater than 3.5 to 1.0 are indicative of renal artery stenosis) [2]. However, the diastolic flow was especially prominent in the proximal left renal artery as compared to the right side (Figure 1B), and a color Doppler US image of the left kidney showed a large-diameter

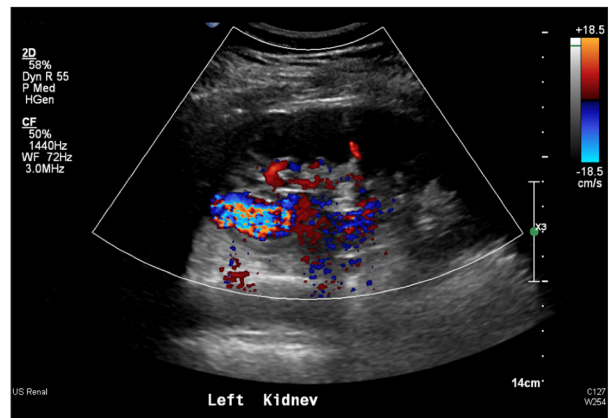


Figure 2. Spectral Doppler of the left-sided vascular malformation.

vascular structure at the hilum (Figure 2) that was not shown on spectral Doppler. An MRA was done, which showed a large left renal upper-pole arteriovenous malformation (AVM) with associated vascular shunting and early opacification of the left renal vein. The elevated diastolic velocity seen on Doppler imaging was therefore understood as being caused by high flow in the area of the vascular malformation, as the high-pressure arterial system shunted into the low-pressure venous system through the large feeding vessel. Since the patient had no history of trauma or surgery, this was considered likely to be a congenital AVM. Selective arteriograms identified a single-origin vessel from the proximal renal artery (Figure 3), and this was closed successfully with coil embolization (Figure 4, Video 1). Fortunately, the arterial branch involved was only perfusing a small part of the renal parenchyma; therefore, there was no significant renal injury after embolization. The patient's BP normalized within a few days and she was taken off of her antihypertensive medications. At 6-month follow-up, she remained normotensive while off all medications and was discharged from the hypertension and high-risk obstetric clinics.

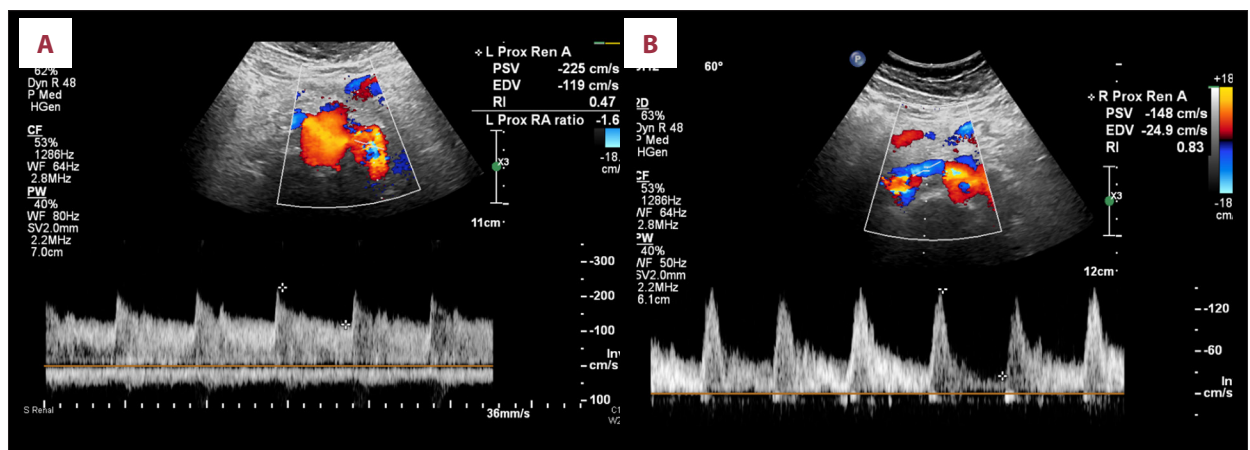


Figure 1. (A, B) Right and left renal Doppler ultrasounds.



Figure 3. Selective angiogram of the vascular malformation.

Discussion

The classification of renal arteriovenous malformations is first categorized into congenital and acquired. Acquired vascular malformations are more common than congenital ones [3]. These acquired malformations are usually caused by renal biopsy, trauma, infection, or vasculitis. In the congenital category, the renal arteriovenous malformations can be categorized into 3 classes based on morphology: angiomatous, cirroid, and aneurysmal [1]. Cirroid aneurysms have more than 4 feeding vessels and are greater than 1 cm in size, whereas angiomatous malformations have a single large feeding vessel supplying multiple draining veins and are less than 1 cm in size [1]. Aneurysmal malformations, such as this one, have a single feeding and draining vessel.

The diagnosis of these lesions can be multimodal. Initial testing can be performed with ultrasound. Renal ultrasound is a common modality for the screening of renovascular hypertension. In the evaluation for renovascular hypertension, the indices used are often renal artery velocities and renal-to-aortic ratio. An elevated renal artery velocity greater than 2 m/s is indicative of renal artery stenosis [4]. Additionally, a renal artery-to-aortic velocity ratio greater than 3.5 to 1 is abnormal and suggest stenosis [4]. In our case, the renal artery velocity was elevated but the ratio was normal. The abnormality in this case was in the elevated diastolic velocities of the affected artery, as shown on Doppler. This was due to increased flow caused by the large-diameter arteriovenous connection and the resultant shunting.



Figure 4. Post-coiling angiogram still photo.



Video 1. Selective angiogram performed after complete coiling of renal AVM.

Diagnosis of these vascular anomalies is often confirmed with CT angiography, MR angiography, or angiography [5]. Angiography with a direct selective angiogram of the malformation is the most helpful in complete characterization [6,7], and is the method of choice for interventional treatment [6].

Treatment of these lesions can be performed with various modalities. The most common methods of treatment involve embolizing the feeding arterial vessel or vessels of the AVM [7]. This can be accomplished with coils, alcohols, glue, or gelatin sponges. In the setting of high-flow vessels, coils are efficient occluding agents and, in some cases, can be followed with n-butyl 2-cyanoacrylate (NBCA), a liquid embolic agent that can easily fill the spaces left by coils [6]. Low-flow areas may

be more amenable to liquid agents or small, injectable embolic particles; however, the risk of distal embolization must be kept in mind when using these agents [6,8]. Embolization leads to resolution of hypertension in 59% of patients treated [8]. Additionally, these intravascular techniques minimize the invasiveness required to correct these malformations and have replaced the previously preferred treatment, which was nephrectomy [9].

No evidence in the literature currently compares medical treatment to therapeutic interventions. There are, however, reports of patients being medically managed and obtaining control of their secondary hypertension without any invasive intervention [10]. However, patients are more likely to achieve control of their hypertension with interventional therapy. Medical management is ultimately unable to resolve

the underlying cause. This treatment comparison certainly warrants further investigation.

Conclusions

This case illustrates that a careful review of duplex waveforms beyond just peak velocity and A-to-R ratios is important to identify uncommon pathologies. In reviewing the patient's previously performed studies, an incorrect diagnosis of early-onset essential hypertension was rejected for a correct and treatable diagnosis of secondary hypertension. Renal arteriovenous malformations should be considered in all secondary hypertension workups for which no alternative explanation has been found in the initial testing. Treatments are usually minimally invasive and highly effective.

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