Pregnancy-associated congenital renal arteriovenous fistula as a cause of hematuria


ABSTRACT

Renal arteriovenous fistulas are a very rare pathology in clinical urology. They are classified as acquired and non-acquired. Acquired fistulas are categorized as iatrogenic, traumatic, spontaneous, and inflammatory. They generally present with hematuria and pain and in some cases with cardiovascular signs and symptoms. Definitive diagnosis is made through imaging studies and there are different types of treatment. Congenital renal arteriovenous fistula presenting during pregnancy is even more rare, with only seven cases reported in the biomedical literature.

Keywords: Renal arteriovenous fistula, pregnancy, hematuria, Mexico.

RESUMEN

Las fistulas arterio-venosas renales constituyen una patología muy poco frecuente en la urología clínica. Se clasifican en adquiridas y no adquiridas. Las fistulas adquiridas se clasifican en iatrogénicas, traumáticas, espontáneas e inflamatorias. Su forma de presentación generalmente con hematuria y dolor y en algunos casos signos y síntomas cardiovasculares, pero el diagnóstico definitivo es a través de estudios de imagen; así mismo, existen diversas formas de tratamiento. Es aún más rara la presentación de fistula arterio-venosa renal congénita en el embarazo, no más de siete casos se han publicado en la literatura biomédica.

Palabras clave: Fistulas arterio-venosas renales, embarazo, hematuria, México.
INTRODUCTION

In 1923 Varela was the first to describe a case of renal arteriovenous fistula (RAVF).\(^1,2\) It has an estimated prevalence of 0.04%, but its incidence is on the rise as a result of the advances in diagnostic imaging techniques.

Origin of arteriovenous fistula at the kidney level can be congenital (22-25%), idiopathic (3-5%), or acquired (70-75%).\(^3\) High flow vascular malformations (congenital) are localized in order of frequency in the upper pole (45%), middle third (30%), and lower pole (25%).\(^4\)

They can arise forming part of different hereditary diseases such as Sturge-Weber syndrome and Klippel-Trenaunay syndrome.\(^2\)

Characteristically their morphology is cirsoid with tortuous nests and multiple arteriovenous communications at the segmental or interlobar level. They are made up of dysplastic subepithelial vessels (absence of elastic lamina) localized at the pyelocaliceal level.\(^3\)

Acquired arteriovenous fistula (AVF) are more frequent and can be secondary to kidney trauma, tumors, or can be iatrogenic as a result of kidney biopsy, percutaneous kidney surgery, open surgery, or post-extracorporeal shock wave lithotripsy (ESWL). They are located in order of frequency in the upper pole (45%), middle third (30%), and lower pole (25%).\(^4\)

Only 7 cases were found in the literature review and they were reports on pregnant patients presenting with hematuria and renal arteriovenous fistula rupture.\(^5,7\) The hyperdynamic circulatory state during pregnancy may predispose vascular malfunction bleeding.\(^5\)

Clinical suspicion and a thorough and precise hematuria differential diagnosis are essential for diagnosis. Angiography shows a series of characteristic signs from which definitive diagnosis of renal arteriovenous malfunction can be made that include cirsoid nest, immediate filling of the renal and gonadal veins and vena cava, and slightly radio-opaque nephrographic phase.\(^4\)

Currently angiography and posterior embolization appear to be first treatment choice. However, therapeutic choice varies according to clinical symptoms, fistula output, and the technical means available in each center.

In asymptomatic cases and cases of low arteriovenous fistula output, observation is the initial approach.\(^4\)

Surgery is reserved for cases in which fistula output is elevated, affecting large vessels (extrarenal fistulas) and for cases in which percutaneous occlusion has failed.\(^8\)

CASE PRESENTATION

Patient is a 26-year-old female nurse with no important past medical history who sought medical attention at the emergency room of the Hospital Regional Lic. Adolfo López Mateos in her 11th week of pregnancy when she had sudden onset of total hematuria with amorphous, filiform clots and no other signs or symptoms. She was evaluated in the Urology Department where hematuria protocol was initiated with kidney and bladder ultrasound and laboratory tests due to her serious condition, ruling out urinary infection, tumor, and lithiasis.

She was managed with bladder washes and cystoscopy but hematuria did not remit. Hemoglobin and hematocrit value then dropped significantly and cystoscopy was carried out that showed a bladder with no apparent pathology. Bright red blood was observed in the left ureteral meatus meriting left ureteroscopy that showed slight continuous pulsating bleeding arising from left kidney renal cavities.

Conservative management with hemotransfusions was decided upon and there was spontaneous hematuria remission after ureteroscopy. The following day new hematuria symptoms presented with the same characteristics and together with the hospital perinatology department the decision was made to carry out renal angioresonance study and superselective renal angiograph, confirming left cirsoid arteriovenous fistula (Images 1, 2, 3 and 4).

Disease progression had periods of spontaneous hematuria remission and recurrence, the latter of which did not alter patient or product vital signs or hematic or hydrometabolic values.

Because of gestational age conservative management and strict surveillance were continued enabling the patient to reach full-term pregnancy. She underwent cesarean section at 39th gestational week resulting in live single birth. Neonate was in excellent condition with an Apgar score of 9/9 and no congenital malformation. After surgery, patient progressed satisfactorily with no symptom recurrence. Radiologic surveillance has been maintained and there has been no evidence of congenital renal arteriovenous fistula.

DISCUSSION

Congenital renal arteriovenous fistula (RAVF) is a rare entity. However, since its description in 1923, great advances have been made in relation to its diagnosis and treatment, importantly allowing for the conservation of kidney function through less invasive techniques.\(^6\)

This pathology is even rarer during pregnancy with only 7 cases reported in the literature.\(^5,7\) Estimated prevalence is under 0.04%.\(^6\)
The origin of RAVF at the kidney level can be congenital (22-25%), idiopathic (3-5%), or acquired (70-75%). High flow vascular malformations (congenital) are localized in order of frequency in the upper pole (45%), middle third (30%), and lower pole (25%).

Congenital arteriovenous malformations appear to arise from genetic alterations that distort the cellular ability to direct and regulate the periendothelial environment during angiogenesis. These alterations are usually present at birth and usually manifest themselves in infancy/adolescence when they acquire sufficient size to be clinically expressed. Likewise, they can be associated with complex syndromes such as Sturge-Weber, Klippel-Trenaunay, Parkes-Weber, Cobb, Wyburn-Mason, Gorham-Stout and others. They more commonly present at 50-60 years of age although their appearance in infancy or even during gestation is not uncommon, given that the hyperdynamic circulatory state during pregnancy predisposes bleeding.

Magnetic resonance imaging with low signal in T1 and T2 has shown multiple hypertrophied arteries with enlarged vascular spaces connected by lineal or focal shunts, with no evidence of identifiable mass. Nevertheless, both explorations present high sensitivity but low specificity and when there is diagnostic doubt
angio-CAT scan or angioresonance study should be carried out.4,8

Presently angiography and posterior embolization appear to be first treatment of choice. However, therapeutic choice varies according to clinical symptoms, fistula output, and the technical means available in each center.

In asymptomatic cases and cases of low arteriovenous fistula output, observation is the initial approach and in cases of complete remission periodic radiologic surveillance should be continued.4

Surgery is reserved for cases in which fistula output is elevated, affecting large vessels (extrarenal fistulas) and for cases in which percutaneous occlusion has failed.8

## CONCLUSIONS

Congenital renal arteriovenous fistula (RAVF) is a rare entity with a prevalence of 0.4%. This pathology is even rarer during pregnancy, with only 7 cases reported in the literature.

Clinical suspicion and a thorough and precise differential diagnosis are essential for diagnosis. Treatment should be directed at conserving the greatest amount of renal parenchyma. The best alternative is selective embolization when circumstances permit. Observation is an alternative when RAVF is of low output and when there are no hemodynamic repercussions. Strict radiologic surveillance should then be continued.

## BIBLIOGRAPHY