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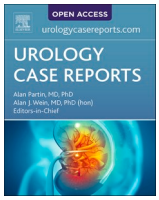

Trauma and reconstruction

Delayed symptomatic renal arteriovenous fistula in a 24 years old male following renal biopsy

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ABSTRACT

We report a case of a 24-year-old male with a history of kidney biopsy at young age due to chronic renal dysfunction and challenging hypertension, who presented with flank pain and hematuria. Initial imaging suggested renal pelvis enlargement, but MRI revealed a massive renal arteriovenous malformation (AVM). Angiographic embolization was abandoned due to extensive effluent flow, followed by successful surgical resection preserving healthy kidney tissue. This case underscores the importance of considering renal AVMs in the differential diagnosis of young patients with gross hematuria or refractory hypertension to prevent complications and improve patient outcomes.

1. Background

Renal arteriovenous malformations (AVMs) are infrequent abnormalities of the renal vasculature defined by the irregular connection of renal arteries and veins. The pathogenesis may either be congenital or acquired, with renal tumor or iatrogenic involvement among possible mechanisms.[1] With an estimated prevalence of 0.04%, the disease occurs extremely rarely.[2] AVMs pose a significant risk to health if not managed properly. Among potential symptoms secondary hypertension, gross hematuria, and cardiac insufficiency are particularly important. Accordingly, timely detection and intervention of renal AVMs can avert severe complications and enhance patient outcomes.

2. Case presentation

A 24-year-old male presented to a primary hospital emergency department with acute right-sided flank pain and gross hematuria. According to his medical history, he was diagnosed with Henoch-Schönlein purpura (IgA vasculitis) in early adolescence, resulting in chronic renal dysfunction. The diagnosis was obtained by biopsy of the right kidney at the age of 16. During follow-up, a cystic structure involving the right kidney had been noticed but left untreated since its presence was asymptomatic. Further preexisting conditions included therapy-refractory hypertension requiring quadruple medication and Crohn's disease.

Immediately ultrasonography unveiled what seemed to be an ectasia of the renal pelvis as well as a full bladder with intravesical coagula. After placement of a urinary bladder irrigation catheter, low-dose-computed tomography (CT) showed no signs of ureterolithiasis or tumor explaining a suspected postrenal obstruction. Magnetic resonance imaging (MRI) revealed an enormous renal AVM, mimicking a congested renal pelvis (Fig. 1). Subsequently, the patient was transferred to a tertiary medical center to receive further treatment.

Here, angiography with digital subtraction analysis (DSA) was performed. Upon examination, a normative superior renal artery accompanied by an inferior renal artery exhibiting a direct connection to the inferior vena cava (IVC) through a profoundly dilated vein was observed. (Fig. 2). The planned embolization of the AVM was not performed due to the significant risk for coil embolism arising from the substantial effluent flow into the VCI.

Given the symptomatic patient, surgical exposure of the kidney was performed. The AVM presented whirring, occupying almost two-thirds of the kidney. By inducing selective ischemia in the lower renal artery, the AVM collapsed, allowing for a clear visualization of the affected site. This revealed that the anomalous finger-sized vessels had entirely displaced the healthy renal parenchyma. The preservation of the healthy superior portion of the kidney was successfully achieved without encountering any additional complications throughout the intra- or postoperative course. (Fig. 3). The patient demonstrated a prompt recovery and eventually succeeded in discontinuing all antihypertensive

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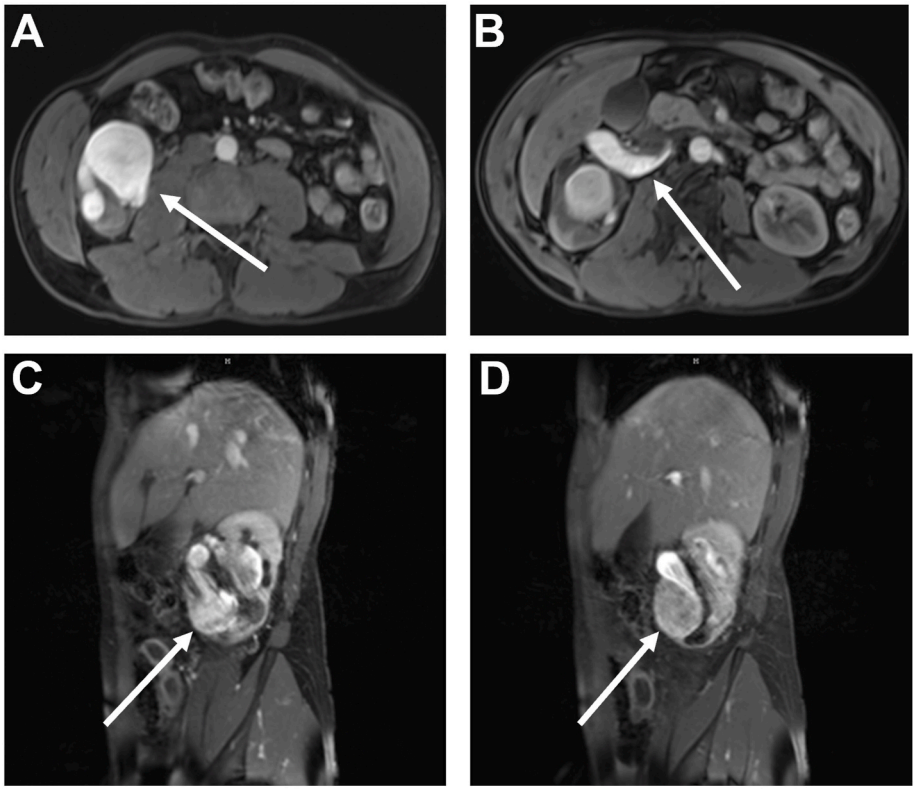

Fig. 1. Computed tomography in frontal (A) and axial (B) view. The arteriovenous malformation on the right kidney is marked with arrows and may be mistaken for hydronephrosis. Magnetic resonance imaging in frontal (C, D) and sagittal (E, F) view reveals that the inferior pole of the right kidney is almost completely occupied by an angiographically contrasted mass marked with arrows and draining into the inferior vena cava (D).

medications. Furthermore, the hematuria and proteinuria resolved.

3. Discussion

Numerous case reports on AVM have been published. However, to date, no clinical trials have been published, most likely due to the rarity of the disease.

The pathogenesis distinguishes between congenital, acquired, and idiopathic AVMs, with acquired AVMs accounting for by far the largest proportion.[3] The reasons delineated in the existing literature for acquired AVM include renal biopsy, surgery, traumatic injury, or routine exercise.[1,3,4] Based on the patient's history of receiving an ipsilateral renal biopsy 8 years ago, we hypothesize this to be the causal factor in the development of the AVM. Development of renal arteriovenous fistula (AVF) following renal biopsy has been described as a complication.[4] However, unlike an AVF evident within a few days or months after biopsy, clinically silent courses with the development of an AVM symptomatic years later have not been reported yet.[4] We assume that the vascular malformation grew progressively over time through arterial insufflation but was only correctly diagnosed upon invasion of the renal pelvis. The gradual development of secondary hypertension and a cystic structure of the right kidney in between kidney biopsy and the onset of acute symptoms reinforce this hypothesis.

While spontaneous remission is possible, the disease may become symptomatic by increasing extent and resulting hematuria or cardiovascular disorder arising from significant volume shifts.[2] Accordingly, reported patients' symptoms leading to early AVM diagnosis include predominantly gross hematuria and flank pain, and less frequently hypertension or heart failure.[5] Given that some of these symptoms frequently manifest in urology emergency departments, several authors have noted a tendency for delayed AVM diagnoses, as more common preliminary working diagnoses may take precedence initially like in our case.[3,5] Hence, the consideration of AVM as a pertinent differential diagnosis, along with the application of an appropriate imaging modality, is key to achieving optimal and timely treatment, thereby mitigating potential long-term complications.

In the initial assessment, AVM frequently mimics ectasia of the renal pelvis as observed in the present case. In this context, authors particularly point out the necessity to perform color-coded Doppler ultrasonography even in point-of-care setting.[5] The definite diagnosis follows from contrast-enhanced CT or MRI and/or angiography, as in the present case.

Subsequent treatment planning should involve a multidisciplinary team of urology and interventional radiology.[5] DSA-guided transarterial embolization is the procedure of choice in most cases, however, it has limitations as described previously and shown in our case. Surgical resection remains a viable option in individual cases, allowing for a complete resection of the AVM while preserving the healthy renal sections.[1] In the presented case, surgical exposure of the kidney and subsequent resection of the AVM resulted in a successful outcome, with the patient experiencing a swift recovery and eventual discontinuation of quadruple hypertension medications. In almost any reported case in the literature, successful treatment of AVM results in the resolution of both acute and secondary symptoms, highlighting that treatment provides a causal therapy approach.[1,4]

4. Conclusion

This case highlights the importance of considering renal AVMs in the differential diagnosis of patients presenting with gross hematuria, ectasia, and/or hypertension refractory to treatment at a young age. The medical history of a previous renal biopsy, even if performed years ago, should be included in the assessment of patients with unusual ectatic alterations of the renal pel vicaliceal system, flank pain, and gross

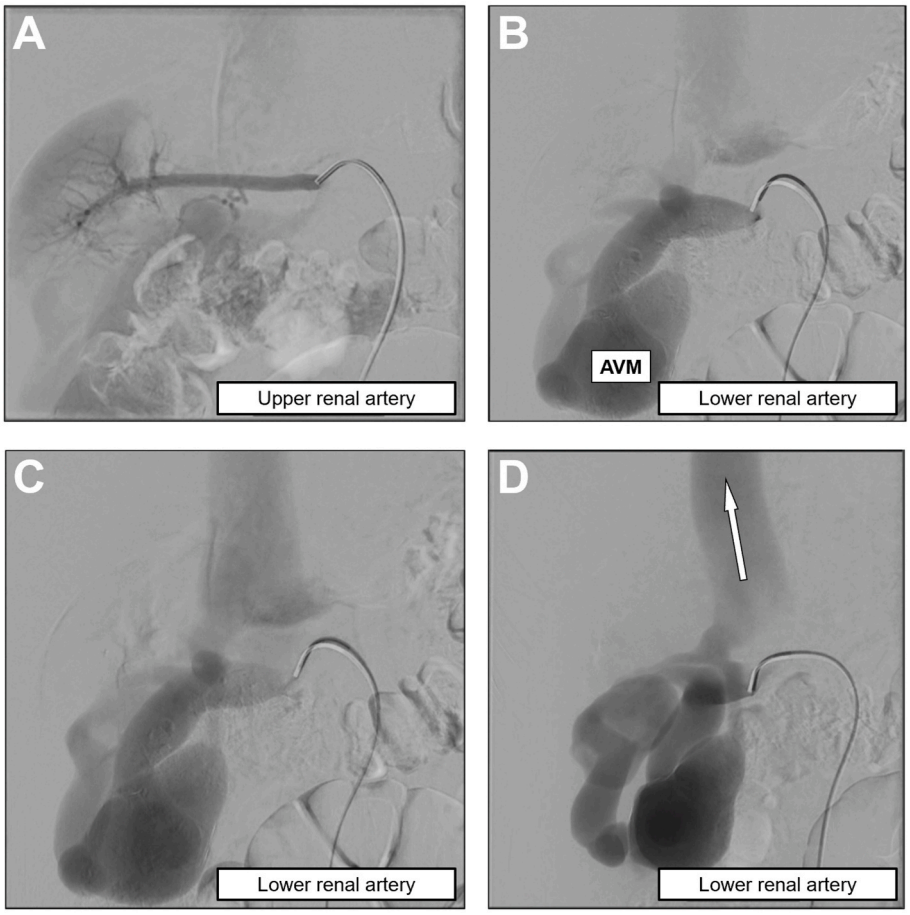

Fig. 2. Angiography with digital subtraction analysis. The upper renal artery shows normal configuration and perfusion of the upper pole (A). After application of the contrast medium into the lower renal artery (B), the convoluted arteriovenous malformation (AVM, C) appears with rapid and massive outflow into the inferior vena cava (D).

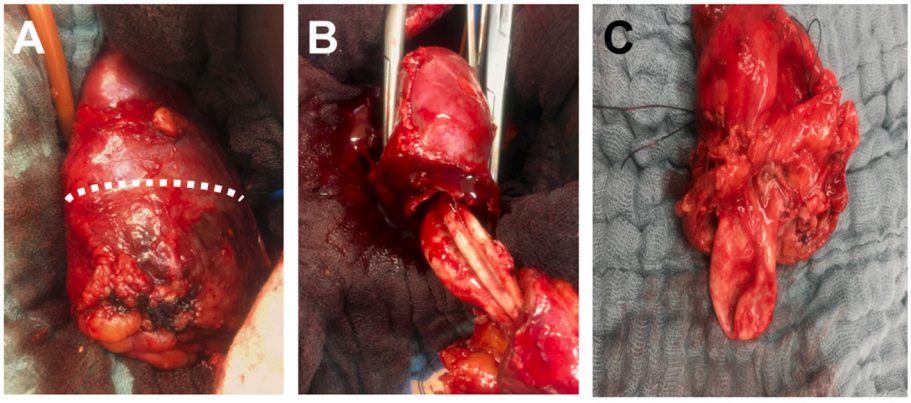

Fig. 3. Surgical removal. Following tourniquet of the lower renal artery, the arteriovenous malformation (AVM) convolute collapses demarking the vital upper portion (A). The dashed line indicates the cutting line of the partial nephrectomy. The AVM is removed (B) and shows finger-sized vascular structures without remaining renal parenchyma (C).

hematuria, along with color-coded renal Doppler ultrasonography. Early recognition and treatment prevent long-term complications, and modern imaging techniques such as MRI and DSA are indispensable to detect and characterize these lesions. DSA-guided embolization is often the preferred method of treatment, but the surgical approach represents a viable option where endovascular treatment fails or reaches its limits.

Consent for publication

Written informed consent for publication was obtained from the patient.

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CRediT authorship contribution statement

Thomas Büttner: Writing – original draft, Visualization, Data curation. **Jörg Ellinger:** Writing – review & editing, Data curation. **Guido Fechner:** Writing – review & editing, Data curation. **Stefan Hauser:** Writing – review & editing, Data curation. **Alexander Cox:** Writing – review & editing, Data curation. **Johannes Stein:** Writing – review & editing, Data curation. **Philipp Krausewitz:** Writing – review & editing, Investigation, Data curation, Conceptualization.

Declaration of competing interest

All authors declare no conflicts of interest.

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