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RENAL ARTERIOVENOUS MALFORMATION MANAGED WITH EMBOLIZATION – CASE REPORT AND REVIEW OF LITERATURE

MALFORMAÇÃO ARTÉRIO-VENOSA RENAL E TRATAMENTO POR EMBOLIZAÇÃO – CASO CLÍNICO E REVISÃO DA LITERATURA

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Abstract

Vascular malformations of the kidney are pathologic processes that involve renal veins and arteries and include arteriovenous malformations (AVMs) and arteriovenous fistulas (AVFs). These lesions may present with a wide range of signs and symptoms that vary from hypertension, hematuria to renal masses. The presence of arterio-venous shunting characterizes AVMs and AVFs. We report the case of a congenital renal AVM in a woman who presented with hematuria and was successfully treated with endovascular embolization in an emergency setting. The lesion was selectively catheterized with a microcatheter and embolization was performed by injection of a mixture containing n- butyl 2-cyanoacrylate (NBCA) and lipiodol. Diagnostic imaging modalities and the technique of embolization are discussed.

Key-words

Renal arteriovenous malformations (AVMs); Renal arterious fistula (AVFs); Gross hematuria; Embolization; n-butyl 2- cyanoacrylate- NBCA.

Resumo

As malformações vasculares do rim são processos patológicos que envolvem as veias e as artérias renais incluindo-se neste grupo as malformações arterio-venosas e as fístulas arterio-venosas. Estas lesões podem apresentar-se clinicamente por um grande espectro de sinais e sintomas que variam desde a hipertensão, a hematúria ou por massas renais. A presença de um shunt arterio-venoso caracteriza quer as malfomações quer as fístulas. Reportamos um caso de uma mulher com uma malformação arterio-venosa congénita desconhecida, que se apresentou inicialmente no serviço de urgência com hematúria, tendo sido tratada com sucesso por embolização endovascular. A lesão foi cateterizada selectivamente com um microcateter tendo a embolização sido realizada pela injecção de uma mistura contendo N–butil-2-cianoacrilato e lipiodol. São discutidas as diferentes técnicas de diagnóstico radiológico assim como a técnica de embolização.

Palavras-chave

malformações renais artério-venosas; Fístula renal artério-venosa; Hematúria; N–butil-2-cianoacrilato.

Introduction

Vascular abnormalities within the kidney are rare and represent a heterogeneous group of diseases[1,2]. There has been controversy regarding their nature and classification. Thus, renal vascular abnormalities are categorised on the basis of their location, as central and peripheral for renal hilum. Arteriovenous malformations (AVMs) are always congenital and very rare in the general population, with an incidence of approximately 4 per 10.000 individuals[1]. On the other hand, arteriovenous fistulas (AVFs) are more common, represent about 70-80% of renal arteriovenous abnormalities, are almost always acquired, and usually result from penetrating trauma, percutaneous biopsy, surgery, malignancy or inflammation[3]. Congenital AVFs are part of the spectrum of congenital AVM. While AVF is defined as a single direct communication between a renal artery and a vein, AVMs are abnormal communications between the renal arterial and venous systems via a vascular nidus, a cluster of multiple, enlarged, tortuous arteriovenous communications[2].

Traditionally, congenital renal AVMs are classified into three forms, depending on the angioarchitecture: the cirsoid is the most common type, characterized by multiple varix-like vascular communications with multiple arteriovenous interconnections[3]; the angiomatous type consists of a single large artery feeding multiple interconnecting distal branches and draining veins[3]; and the aneurysmal type, which typically occurs in elderly patients when a pre-existing arterial aneurysm erodes into an adjacent vein[4]. The aneurysmal type of AVM may be difficult to differentiate from chronic acquired AVFs[2].

Gross hematuria and flank pain are the most common sign and symptom presented by patients with renal AVM[1,5].

Treatment for renal AVMs has evolved from nephrectomy to transcatheter embolization[1].

We report an interesting case of a congenital renal AVM in a woman who presented with hematuria and was successfully treated with endovascular embolization in an emergency setting.

Case Report

A 38-year-old woman was admitted to our hospital with urinary retention, gross hematuria and right flank pain. The patient reported a history of lumbar trauma one month ago. She denied history of hypertension, known urolithiasis or recent medical intervention. She also denied any bleeding disorder and was not taking any medication. Her physical examination was normal, no abdominal bruit on auscultation was found. The patient's blood pressure was 110/80 mmHg, and her heart rate was 103 bpm. Laboratory parameters were within normal range, except for a low hematocrit (19%) and hemoglobin (6,4 g/dl). Urine examination showed massive amounts of erythrocytes.

A bladder ultrasonography (US) revealed a movable 8 cm hyperecogenic mass in keeping with a clot (Fig. 1). No parenchymal or collecting system abnormalities detected.

Bladder wash and catheterisation were performed.

A computed tomography (CT) was also performed in order to better understand the origin of the hematuria (Fig. 2). After an unenhanced acquisition, 100 mL of endovenous contrast were administered at a flow rate of 4 ml/second. Triphasic CT was then performed in the corticomedullary, nephrogenic phases and excretory phase. The unenhanced CT revealed spontaneous hyperdense images on the right collecting system in relation to fresh blood. The enhanced CT scan showed a significant delay in nephrographic and pielographic phase in the right kidney. Additionally, a parenchymal 2 cm mixed lesion was identified in the inferior third of the same kidney, showing fast corticomedullary enhancement. A tumoral diagnostic hypothesis was raised.

On colour Doppler US, a small intraparenchymatous lesion with turbulent high flow was observed in the inferior third of the right kidney, seeming compatible with a vascular malformation. The patient underwent ureteroscopy, which was unremarkable for cancer.

Considering gross hematuria with negative endoscopy findings, and the lesion found on CT and renal Doppler US, differential diagnosis included a renal cell carcinoma (RCC) and a renal AVM. Selective right renal artery angiography was performed using a right transfemoral approach and a 5-French sheath with a hydrophilic guidewire coupled with a 5-French cobra shaped catheter (Fig. 3). Digital subtraction imaging demonstrated the feeding artery to the AVM. The lesion was selectively catheterized with a microcatheter and embolization was performed by slow injection of a mixture containing n- butyl 2-cyanoacrylate - NBCA (Histoacryl, Braun ®) and lipiodol (Lipiodol UltraFluide, Guerbet®) in a concentration 1:2. Once the nidus of the AVM was filled, injection was stopped and the microcatheter was withdrawn. No complications occurred during the procedure. The control angiogram revealed complete exclusion of the AVM.

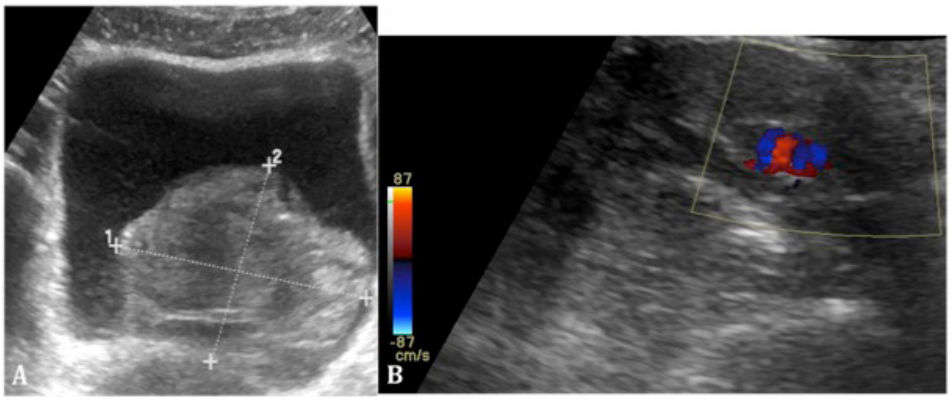

Figure 1 – (A) Uninary bladder sonography showing a large heterogeneous clot. (B) Right renal Doppler showing aliasing in the inferior pole that highlights the AVM.

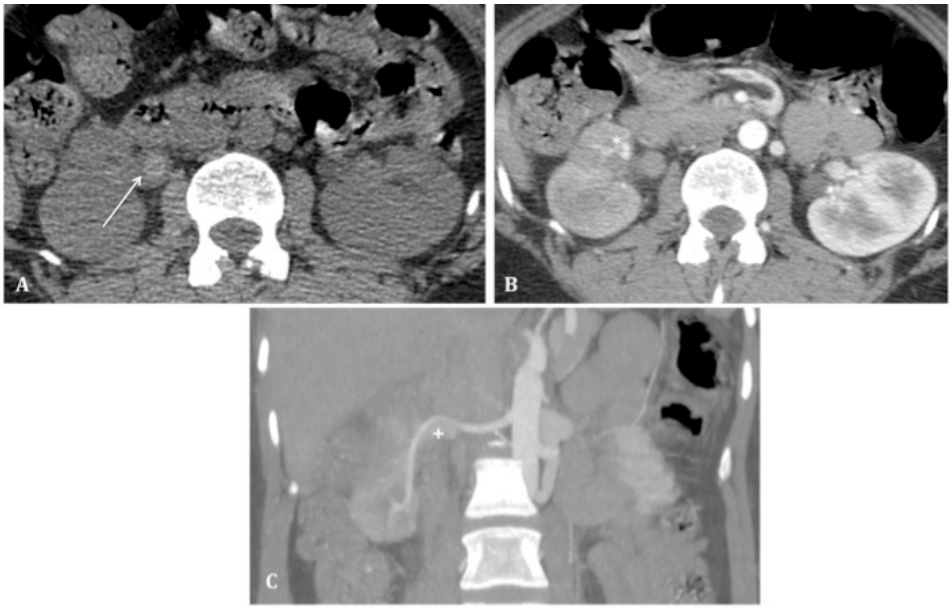

Figure 2 – Abdomen CT on the day of admission. (A) Unenhanced CT axial view demonstrating blood in the right collecting system (arrow). Enhanced CT (B) axial view and (C) MIP coronal view, during the corticomedulary phase showing assimetry perfusion between the kidneys and enhancing tangle of vessels involving the right inferior pole (*) with early renal vein opacificacion (+).

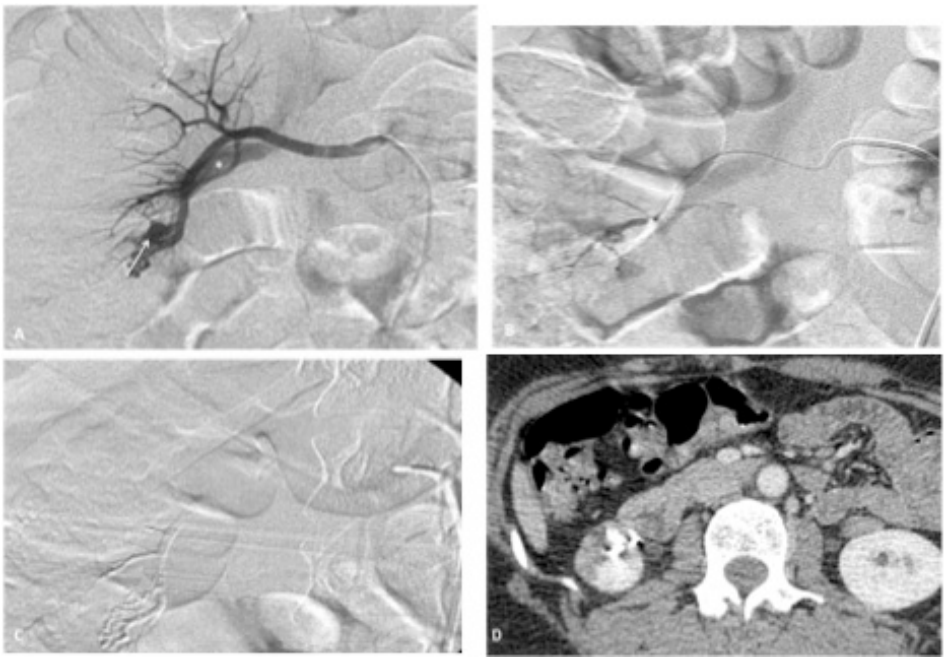

Figure 3 – (A) Right renal angiography, digital subtraction imaging showing glomerulus-like arteriovenous malformation with distinct nidus (arrow) and early draining inferior medial vein (*). (B) Superselective angiography with enlarged depiction of the renal AVM microcatheter tip in wedge position. (C) Control angiography after complete embolization with NBCA, nidus is filled with glue. (D) Enhanced CT 1 month after embolization demonstrated no recurrence of AVM, and a small high density areas indicated residual lipiodol, no hematuria was evident after the first year of follow-up.

The patient was discharged with no signs of hematuria. 12 months after, the patient remained free of symptoms. An angio-CT was performed showing the same delay in nephro and excretory phases on the right kidney as it was seen on the first CT, although no enhancement lesion was observed. In its place, radiopaque post-embolization material was identified.

Discussion

Renal AVMs are rare lesions, with only a little more than 200 reported in the literature[4]. The congenital AVMs are usually composed of multiple tortuous arteriovenous communications in contrast to fistulas that usually have the form of a solitary arteriovenous channel.

AVMs are usually located on the kidney upper pole (45%), but they also can be detected in the mid-point or in the kidney lower pole in an equal ratio, as it was observed in our patient[6]. The left kidney is more frequently involved, and women are affected twice as often as men[2]. The peak incidence is between 30 to 40 years, in keeping with the case reported[6].

Our patient had suffered a blunt lumbar trauma, which is a known risk factor for AVM formation[3]. However, the presence of numerous feeding vessels and multiple arteriovenous interconnections, depicted on CT and confirmed on angiography, was much more suggestive of a cirsoid type renal AVM. These congenital vascular anomalies are presented in 72% of cases, as gross hematuria due to rupture of small venules into the calyces from abnormally increased intravascular pressure[1,4].

Beyond cystoscopy that is needed to rule out any urinary bladder pathology, the radiological workup of a patient with gross hematuria and suspicion of an AVM should include the US as the preferred initial diagnostic method for evaluation of the kidneys[2]. Grayscale US findings of renal AVM include hypoechoic cystic or tubular-like structures of varying sizes. Colour Doppler US may demonstrate the vascular nature of the lesion showing turbulent high flow[2]. CT enhanced imaging may demonstrate a vascular mass particularly in the corticomedullary phase[2]. Delayed CT images may better show the exact size and the relation of the AVM to the pielocaliceal system[2].

The differentiation between AVMs and RCC on CT may be challenging but essential in selecting appropriate management.[5] Varying degree of vascular shunting is observed in both RCC and renal AVMs. Thus after symptomatic treatment, a close follow-up is needed in order to rule out neoplasic lesion, if any doubt persist a biopsy should be carried out[5]. Although CT might be useful for diagnosis, patients with severe characteristic symptoms should proceed directly to angiography and undergo immediate treatment if needed[2,4]. Catheter angiography remains the gold standard for depicting detailed vascular anatomy of renal vascular malformations. Arteriography can define: the main arterial supply to the vascular malformation; the presence of a nidus; the size of arteriovenous shunting, and the venous drainage[2].

Our aim was to immediately treat the AVM by performing endovascular embolization to stop the bleeding, preserve renal parenchymal function, and eradicate the symptoms and hemodynamic effects associated with the abnormality that was diagnosed in this patient, who presented with acute anemia and tachycardia.

Indications for treating an AVM are: a progressive increase in the size of the fistula; recurrent or persistent hematuria; and hemodynamic effects associated with the abnormality, especially hemodynamic descompensation, hypertension, and high-output cardiac failure[4].

In the past, partial/total nephrectomy or surgical ligation of feeding arteries were commonly performed in the presence of symptomatic renal AVMs[1]. Arterial embolization has gained ground compared with surgery, and nowadays is considered the standard therapy to preserve renal function and reduce morbidity. Surgery on the other hand, is still only recommended for large AVMs due to the risk of systemic embolization of the injected material[3].

The benefits of percutaneous embolization treatment are: avoidance of nephrectomy; reduction of peri-operative risk and post-operative morbidity; reduced surgical time and hospital stay; and decreased incidence of renal ischemia[4].

In order to successfully embolize renal AVMs, it is important to achieve complete occlusion of the nidus, where the artery and vein communicate. As previously stated, AVMs usually receive blood supply from multiple arteries and only permanent occlusion of all feeders or occlusion of the nidus can provide successful embolization. Recurrence of AVMs has been reported after technically successful embolization, by recanalization of feeding vessels or recruitment of new feeders after incomplete vascular nidus occlusion[2]. The recurrence frequency is related with the embolization agents used.

Several embolic agents have been proposed and applied: autologous clot; gelatin sponge[7]; stainless steel coils and platinum microcoils[7]; alcohol[7,8]; and NBCA.[7] Many authors have reported primary success of embolization with autologous clot and gelatin sponge, but almost 50% of reported AVMs showed late partial recanalization[9]. Alcohol embolization is performed using an emulsion of absolute ethanol and iodized oil (lipiodol), with a maximum ethanol dose of 0,4 ml/Kg body weight, the emulsion must be slowly injected via the afferent arteries in order to prevent reflux[10]. Recanalization of vessels, with alcohol embolization, has been reported in patients with facial AVMs[10]. NBCA or glue is diluted with lipiodol with the ratio of 1:3; the ratio is sometimes modified, depending on the distance between the nidus and the microcatheter tip, and the velocity of venous return[10]. Lipiodol is used to opacify the embolic agent and to slow the polymerization time of the glue. Prior to the injection of NBCA mixture, a manual test injection with contrast media is performed, to evaluate the velocity of flow through the AVM and the venous return, and also the reflux into non targeted areas. Just before the administration of NBCA mixture, the lumen of the microcatheter is filled with glucose solution in order to prevent the contact with blood, which can induce conjugation of the agent. NBCA mixture is injected in the same way as the test injection and the microcatheter should be quickly removed after injection. Controlled injection of an adequate amount and dilution of liquid embolic agent, such as NBCA or alcohol, is very important in order to prevent ischemic complication or pulmonary embolization[2].

In a recent review by Murata et al[10]. concerning endovascular embolization strategy for renal AVMs, 12 patients were examined during at least 48 months after embolization in the setting of gross hematuria. This study assessed technical and clinical success, and also complications. Different materials have been used for embolization including gelatin sponge particles, coils, ethanol and NBCA. The results suggest that embolization using coils alone is not preferable, while the procedure using liquid agents such as NBCA or ethanol are efficient enough to obtain sustained relief of hematuria. Another study with 7 cases of congenial AVMs that used micro coils as an embolization strategy, suggested as a general rule that coil occlusion is safer mainly in large AVMs with high blood flow[8]. One of the advantages of the liquid embolization is the fact that these agents do not interfere with possible future retreatments[10].

In the reported case, we did not choose to use coils, alcohol or gelatin sponge based on the fact that the patient's lesion was not very large. Accordingly to the literature in renal AVMs, NBCA mixed with lipiodol, if placed superselectively, is safe, produces excellent results and does not interfere with possible future treatments.

The embolization value of renal AVMs has not been fully established, as well as which embolization agent should be used, due to the lack of clinical evidence supported by statistically significant results. Nonetheless, embolization by selective catheterization can be considered safe and effective in the setting of gross hematuria due to renal AVM.

Conflict of interst disclosure statement

Author 1, Author 2, Author 3 and Author 4 declare that they have no conflicts of interest.

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