Journal of Urology and Renal Diseases
Nunes TF, et al. J Urol Ren Dis 07: 1251.
www.doi.org/10.29011/2575-7903.001251
www.gavinpublishers.com

Case Report

Spontaneous Rupture of Renal Pseudoaneurysm in Microscopic Polyangiitis: A Case Report and Literature Review

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Citation: Nunes TF, Marchiori E, Motta-Leal-Filho JM (2022) Spontaneous Rupture of Renal Pseudoaneurysm in Microscopic Polyangiitis: A Case Report and Literature Review. J Urol Ren Dis 07: 1251. DOI: 10.29011/2575-7903.001251

Received Date: 10 March, 2022**; Accepted Date:** 25 March, 2022**; Published Date:** 28 March 2022

Abstract

Background: Renal artery pseudoaneurysm is a rare vascular lesion usually caused by trauma or percutaneous urological procedures. Spontaneous rupture of pseudoaneurysms without predisposing events, especially in hemodialysis patients, has rarely been reported. **Case Presentation**: A 52-year-old woman had a diagnosis of glomerulonephritis with polyangiitis (ANCA negative). After 15 days of hospitalization, she developed severe abdominal pain and hemorrhagic shock. Contrast-enhanced computed tomography revealed a left perirenal hematoma with pseudoaneurysms. Renal angiography showed multiple pseudoaneurysms in the left renal artery branches, and embolization was performed. Post-angiography images showed no pseudoaneurysms. Her abdominal pain improved, and she is scheduled to be discharged to home care. **Conclusions**: When patients on maintenance dialysis complain of severe abdominal pain, spontaneous rupture of a renal pseudoaneurysm should be considered as a differential diagnosis, even if the patient has no history of trauma or previous urological procedures.

Keywords: Case report; Hemodialysis; Percutaneous embolization; Renal artery pseudoaneurysm; Spontaneous rupture

Abbreviations: MPA: Microscopic Polyangiitis; ANCA: Antineutrophil Cytoplasmic Antibody; PAN: Polyarteritis Nodosa; AAV: ANCA-Associated Vasculitis; GPA: Granulomatosis with Polyangiitis; PR3: Proteinase 3; GBM: Glomerular Basement Membrane; MPO: Myeloperoxidase; CVVHDF: Continuous Venovenous Hemodiafiltration; MZR: Mizoribine

Introduction

Microscopic Polyangiitis (MPA) is a rare, systemic, necrotizing vasculitis that often occurs in the presence of Antineutrophil Cytoplasmic Antibody (ANCA), affecting small vessels such as arterioles, venules, and capillaries [1]. In contrast, formation of aneurysms and rupture of medium-sized vessels are typically associated with Polyarteritis Nodosa (PAN). Rupture of arterial aneurysms or hemorrhage of medium vessels is a rare occurrence in ANCA-Associated Vasculitis (AAV) that has been previously described in only a few patients with granulomatosis with polyangiitis (GPA) [2-5] and MPA [6]. We describe the clinical presentation, evaluation, and diagnosis of a patient with MPA who developed spontaneous rupture of a renal artery pseudoaneurysm and was treated with transarterial embolization.

Case Report

Patient Information

A 52-year-old woman presented to the emergency department

with a 2-day history of upper abdominal pain, which worsened when coughing and speaking. She reported coughing for 10 days but tested negative for COVID-19. As for comorbidities, the patient had type 2 diabetes mellitus controlled with oral medications.

Clinical Findings

Physical examination showed blood pressure of 110/70 mm Hg, pulse rate of 115 beats per minute, and respiratory rate of 20 breaths per minute. She was afebrile. Patient's height was 160 cm and weight was 70 kg. Examination revealed regular heartbeat without murmur, lungs clear to auscultation, and a distended abdomen with diffuse tenderness to palpation and pain on percussion. There were no inflamed joints, nasopharyngeal abnormalities, or dermatological manifestations. On admission, she did not require intubation or mechanical ventilation, was hemodynamically stable and had a normal neurological examination.

Diagnostic Assessment

Complete laboratory tests and blood gas analysis were obtained, and Computed Tomography (CT) of the lungs and upper and lower abdomen was performed. Lung CT scans showed small bronchopneumonic consolidation in the posterior segments of both upper lobes, associated with bilateral pleural effusion, more intense on the left side, and slight cavitation in the left apex. Abdominal CT scans showed no significant abnormalities. Laboratory test results were as follows: white blood cell count 16 × 103 /μL (80% neutrophils, 10% lymphocytes, 5% monocytes, and 2% eosinophils); hemoglobin 7.2 g/dL; hematocrit 21.1%; platelet count 235 × 103 /μL; C-reactive protein 46 mg/dL; urea 236 mg/dL; serum creatinine 7.6 mg/dL (2 months before admission, the level was 0.7 mg/dL); and albumin 2.3 g/dL. She had normal liver enzyme and electrolyte levels. The erythrocyte sedimentation rate was 122 mm/hour. Her urine test showed protein 4+ and blood 3+, but no red cell casts or dysmorphic red blood cells. Serological testing was negative for proteinase-3 (PR3)-ANCA, c-ANCA, antinuclear antibody, anti-Glomerular Basement Membrane (GBM) antibody, and anti-myeloperoxidase (MPO)-ANCA. Biopsy of the right kidney together with clinical and laboratory data confirmed the diagnosis of glomerulonephritis with polyangiitis (ANCA negative). After 15 days of hospitalization, with the patient developing severe abdominal pain and hemorrhagic shock, CT angiography showed massive retroperitoneal hematoma formation on the left side with multiple renal micropseudoaneurysms with evidence of active bleeding (Figure 1).

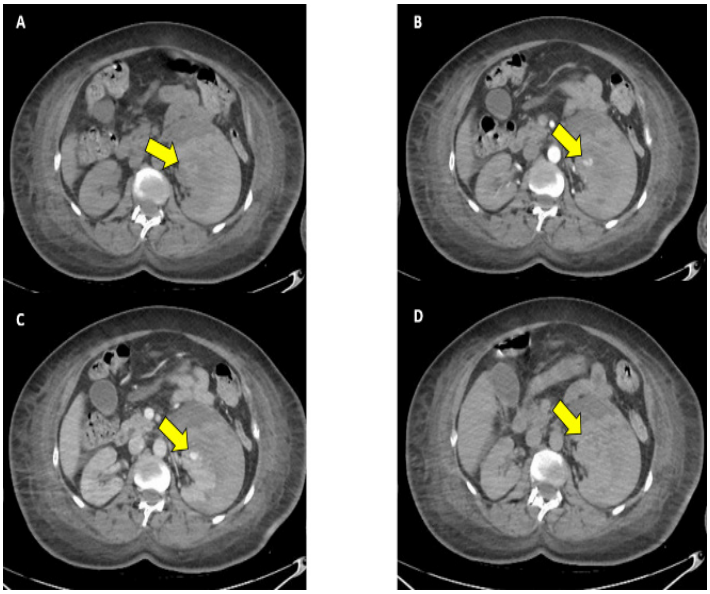

Figure 1: Contrast-enhanced abdominal computed tomography in the axial plane showing a large left perirenal hematoma **(A)** with evidence of active bleeding better visualized in the arterial phase **(B)** and portal phase **(C).** Excretory-phase image, after 10 minutes of acquisition **(D),** showing absence of contrast medium clearance, thus confirming nephropathy. Yellow arrows indicate the focus of active bleeding from the ruptured pseudoaneurysm.

Therapeutic Interventions

Spontaneous rupture of the aneurysms with concomitant retroperitoneal hematoma was suspected after CT angiography, and urgent angiography was performed. Angiography showed multiple left intrarenal artery aneurysms, and transarterial embolization was performed because these arteries were the source of hemorrhage (Figure 2). One day after embolization, the patient improved with extubation and no further use of vasoactive drugs. Continuous venovenous hemodiafiltration (CVVHDF) was initiated due to acute renal failure, and she received pulse therapy with methylprednisolone (1 g/day for 3 days), followed by maintenance therapy with prednisolone (50 mg/day) and pulses of 700 mg of cyclophosphamide, after adequate hydration, twice every 2 weeks. The daily dose of prednisone was gradually reduced.

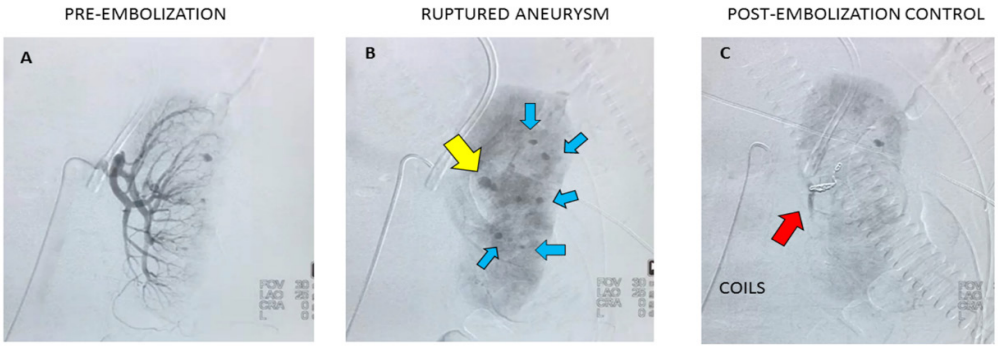

Figure 2: Left renal arteriogram showing multiple small pseudoaneurysms (blue arrows in 2A and 2B). Note the one located in the medial portion, which showed active bleeding (yellow arrow). Embolization of the artery supplying the ruptured pseudoaneurysm was performed using microcoils (red arrow in 2C). The procedure was technically successful and uneventful.

Follow-Up and Outcomes

The patient remains stable with no evidence of hypervolemia after cessation of antimicrobial therapy. There have been no reports of fever. Blood glucose levels are stable, and percutaneous gastrostomy was performed. The patient is alert and interacts with the examiner but has hearing difficulties. She is using tracheostomy with compressed air (5 L/min) with stable vital signs. She is scheduled to be discharged to home care.

Discussion

In this case report, we presented a rare case of renal hemorrhage in a patient with ANCA-associated MPA who was successfully treated with transarterial embolization of active bleeding. The patient received therapy with steroids and mizoribine (MZR), and then maintenance hemodialysis for end-stage renal disease. Diagnosis was confirmed by previous biopsy of the right kidney along with laboratory tests. Renal vascular disease is often described as a complication of classic PAN. The occurrence of multiple artery aneurysms is a classic feature of PAN and detected in 50%-60% of cases, with renal involvement in 80%-90% of cases [6,7]. Spontaneous renal hemorrhage is a rare but fatal clinical condition. In the present case, the patient had undergone a biopsy of the right kidney 15 days earlier, with diagnostic confirmation of Wegener granulomatosis, and then developed hemorrhagic shock, with her imaging tests showing a large perirenal hematoma resulting from a ruptured renal microaneurysm. She was successfully treated with microcoil embolization.

Treatment of AAV complicated by a ruptured renal artery should include supportive therapy, such as fluid resuscitation and blood transfusion, and transcatheter arterial embolization if active bleeding is suspected. Cases of renal artery rupture have been reported [6,7]. Occasionally, additional immunosuppressive therapy may be considered as a treatment option if vasculitis is the cause. In the reported cases of AAV-associated renal artery ruptures listed in Table 1, all patients survived.

References	Sex/age (years)	Autoantibody	Presentation	Therapy	Outcome
[6]	M/55	MPO	Abdominal pain, shock	Blood transfusion, steroids	Survived, HD
[7]	F/82	MPO	Nausea, shock	Coil embolization, steroids, mizoribine	Survived, HD
Our case	F/52	ANCA -	shock	Coil embolization	Survived, HD
MPO: antineutrophil cytoplasmic antibody with myeloperoxidase; HD: Hemodialysis					

Table 1: Reports of perirenal hemorrhage and spontaneous rupture of renal pseudoaneurysms in patients with microscopic polyangiitis ANCA-associated vasculitis.

Conclusion

This case report describes an unusual complication of MPA. Although small-vessel vasculitis predominates in MPA, inflammation of medium-sized arteries may also occur. A ruptured renal artery should be considered in patients with MPA, even in the absence of previous trauma and renal biopsy, when unexplained anemia or signs of shock are observed. Renal artery embolization provides an effective treatment option for these patients in the emergency setting.

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