Int J Clin Exp Med 2019;12(3):2883-2887
www.ijcem.com /ISSN:1940-5901/IJCEM0085468

*Case Report*

# Spontaneous rupture of kidneys triggered by microscopic polyangiitis

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Received April 17, 2018; Accepted February 12, 2019; Epub March 15, 2019; Published March 30, 2019

**Abstract:** Rationale: Microscopic polyangiitis (MPA) is defined by the 2012 revised Chapel Hill Consensus Conference Nomenclature of Vasculitides as necrotizing vasculitis, with few or no immune deposits, predominantly affecting small vessels (i.e. capillaries, venules, or arterioles) and granulomatous inflammation is absent. MPA is clinically characterized by small-vessel vasculitis primarily affecting the kidneys and lungs but other organs may be involved as well. Spontaneous rupture of kidneys is a rare but extremely dangerous event in clinical practice. Here is reported a successfully treated case of spontaneous renal rupture triggered by MPA. Patient concerns: A 57-year-old female complaining of fever for 2 weeks and edema for 1 week presented with newly developed severe lumbago, delirium, acute renal failure, and hemorrhagic shock. Radiological imaging revealed large bilateral peri-renal hematoma and compression of renal parenchyma. Diagnoses: Acute renal failure and hemorrhagic shock caused by spontaneous rupture of kidneys which was triggered in turn due to MPA. Interventions: Measures of absolute bed rest, blood transfusion, hemostasis, and rehydration were immediately taken as first aid measure to stabilize vital signs. Methylprednisolone pulse therapy was the core of MPA treatment. Oral administration was given after discharge. During the follow-up period, the dosage of glucocorticoid reduced step by step and lasted for 15 months. Outcome: Renal function was recovered and binaural listening was improved markedly. Hypoalbuminemia, proteinuria, and hematuria was cured, CT examination demonstrated remarkable absorption in peri-renal hematoma and less compression of renal parenchyma. Conclusion: MPA could trigger spontaneous rupture of kidneys without warning and should be included in the differential diagnosis of spontaneous renal rupture; conservative treatment based on glucocorticoid is safe and effective for the renal rupture caused by MPA.

**Keywords:** Spontaneous rupture of kidneys, microscopic polyangiitis

## Introduction

Spontaneous rupture of kidneys is a rare but extremely dangerous event in clinical practice. There is often no precursor before onset. The common causes of spontaneous rupture of kidneys include renal tumor, anticoagulation therapy, vascular disorders such as aneurysms associated with autoimmune diseases mainly polyarteritis nodosa, renal cyst, renal calculi, severe pyelonephritis and renal allograft rupture because of acute rejection of the implant. Although surgical intervention and interventional embolization therapy can be used to treat the massive bleeding from the ruptured kidney, spontaneous renal rupture still has a high mortality rate. Here, a case of spontaneous renal rupture caused by MPA is reported, which was successfully cured through conservative treatment based on glucocorticoid.

## Case report

A 57-year-old woman presented to our department complaining of fever for 2 weeks and edema for 1 week. A past history of Otitis media and nerve deafness for half a year could be elicited. The demographic and laboratory features at baseline are presented in **Table 1**. Physical examination showed increased temperature (38.5°C), normal blood pressure (Bp, 138/86 mmHg) and heart rate (HR, 68 bpm). Laboratory tests showed abnormal findings in blood routine (white blood cell $20.2\times10^9$/L, neutrophil

**Table 1.** Baseline data of the patient

| | | | |
|---|---|---|---|
| Age (years) | 57 | eGFR | 21.21 ml/min*1.73 $m^2$ |
| Gender | Female | Albumin | 26 g/L |
| Temperature | 38.5°C | 24 h urinary protein | 1.17 g/24 h |
| Blood pressure | 138/86 mmHg | Urine erythrocyte | 1090/μl |
| Heart rate | 68 bpm | ANCA-P | Positive |
| White blood cell | $20.2\times10^9$/L | ANCA-C | Negative |
| Neutrophil ratio | 89% | Myeloperoxidase | Positive |
| Hemoglobin | 129 g/L | Proteinase 3 | Negative |
| Platelet | $228\times10^9$/L | Other immunology indexes | Negative |
| D-Dimer | 4200 μg/L | Serum tumor markers | Negative |
| Fibrinogen | 522.7 mg/dL | Sputum bacterial culture | Negative |
| Serum creatinine | 220 μmol/L | Blood bacterial culture | Negative |

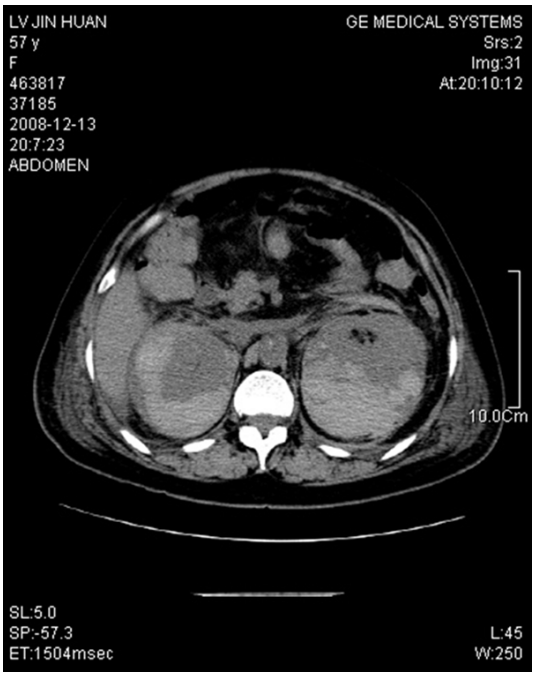


**Figure 1.** Abdomen CT on the day of shock. Urgent abdomen CT examination on the day of shock showed large bilateral peri-renal hematoma and compression of renal parenchyma (2008.12.13).

ratio 89%), renal function (serum creatinine, Scr, 220 μmol/L), liver function (albumin, ALB, 26 g/L, no obvious abnormal changes in other indicators) and coagulation function (D-Dimer 4200 μg/L and fibrinogen, FIB, 522.7 mg/dL). The markedly elevated 24 hour urinary protein (1.17 g/24 h) and urine erythrocyte (1090/μl) were found in urine inspection. To ascertain the causes of hematuria and proteinuria, immunological tests were carried out that included: antineutrophil cytoplasmic antibody (ANCA)-P(+), ANCA-C(-), myeloperoxidase (MPO)(+), proteinase 3 (PR3)(-), others like ANA, dsDNA, SSA, SSB and serum tumor markers, were negative. Sputum and blood bacterial culture were also negative. The patient was diagnosed as acute renal failure (ARF), ANCA-associated vasculitis, microscopic polyangiitis (MPA), and nerve deafness based on the above abnormal findings. Methylprednisolone (20 mg, iv), anti-infection and diuretic therapy were immediately given.

One week after admission, the patient developed sudden chills, fatigue, shortness of breath, severe lumbago, and delirium. Monitoring results showed: Bp 90/60 mmHg, HR 90bpm, hemoglobin (Hb) 29 g/L, platelet (PLT) $407\times10^9$/L and Scr increased to 480 μmol/L rapidly. Urgent abdomen CT showed large bilateral peri-renal hematoma and compression of renal parenchyma (**Figure 1**). This condition was considered as spontaneous renal rupture and hemorrhage caused by MPA. Measures of absolute bed rest, blood transfusion, hemostasis and rehydration were immediately taken. Methylprednisolone pulse therapy (200 mg/day) was used to treat the primary disease. After 3 consecutive days of 200 mg/day, the dose of methylprednisolone reduced to 80 mg/day, which also lasted 3 days. Then the dose of methylprednisolone decreased to 40 mg/day until discharge. After intermittent blood transfusion (1600 ml in total) and rehydration, consciousness and blood pressure gradually returned to normal. Hb increased to 80 g/L one week later. Scr progressively increased and then began to fall from the peak of 578 μmol/L 3 days after the renal rupture. After treatment, the patient was discharged when Bp 150/90 mmHg, Scr 155 μmol/L, ALB 32 g/L, and renal CT examination for the second time showed

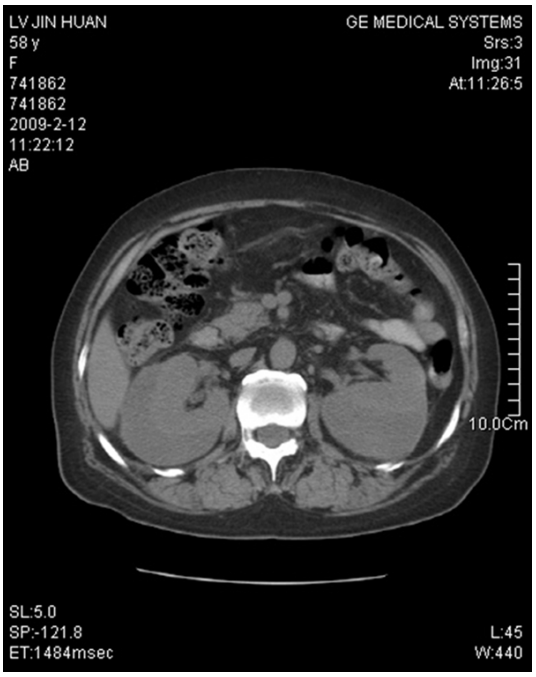


**Figure 2.** Abdomen CT 3 months after renal rupture. Abdomen CT re-examination 3 months after renal rupture showed remarkable absorption in peri-renal hematoma and obvious alleviation in renal parenchymal compression (2009.03.05).

peri-renal hematoma that was partially absorbed. After discharge, methylprednisolone (24 mg, po) continued to be used. 2 months after discharge, CT examination for the third time demonstrated remarkable absorption in peri-renal hematoma and less compression of renal parenchyma (**Figure 2**). Methylprednisolone usage for 3 months normalized injured renal function (Scr: 94 μmol/L), anemia (Hb: 126 g/L) and hypoalbuminemia (ALB: 37 g/L). Simultaneously, proteinuria reduced from 1.17 gram/day to 0.5 gram/day and even binaural listening improved markedly. The dosage of glucocorticoid began to be reduced step by step. The usage of methylprednisolone has been lasted for 15 months. During the follow-up period, the therapeutic effects are shown in **Table 2**. This patient was followed for 29 months. At the end of the last follow-up, blood and urine auxiliary examination data showed as follow: Hb: 128 g/L, ALB: 46 g/l, Scr: 67 μmol/L, 24 h urine protein: 288 mg, no hematuria, ANCA-P(-), ANCA-C(-), MPO(-), PR3(-).

## Discussion

The patient can be diagnosed as ANCA-associated vasculitis, microscopic polyangiitis (MPA) based on the manifestation of glomerulonephritis such as proteinuria, hematuria, increased Scr and the positive MPO-ANCA(P) according to the new nomenclature and classification of vasculitis in CHCC 2012 [1] after excluding secondary vasculitis such as rheumatoid or lupus vasculitis, etc. One week after admission, spontaneous renal rupture happened without warning, which should be responsible for the sudden decrease in hemoglobin and blood pressure, progressive increase in serum creatinine and the clinical symptoms of severe lumbago, delirium, and shock. Radiographic analyses and blood tests verified MPA was the chief contributor to the spontaneous rupture of kidneys after excluding sepsis and coagulopathy.

The most common causes of spontaneous rupture of kidneys include renal tumor, anticoagulation therapy, vascular disorders such as aneurysms associated with autoimmune diseases mainly polyarteritis nodosa, renal cyst, renal calculi, severe pyelonephritis and renal allograft rupture because of acute rejection of the implant [2-6]. MPA predominantly affect small vessels, in 90% of patients the kidneys are involved. Renal involvement is manifested by microscopic hematuria with cellular casts in combination with proteinuria which is, generally, not massive. In addition, deterioration of renal function frequently occurs clinically apparent as rapidly progressive glomerulonephritis [7]. Additionally, inflammation of MPA can weaken the vessel wall and lead to aneurysm formation. Disruption of the internal and external elastic laminae is noted. However, arterial aneurysm rupture or medium-vessel hemorrhage is a rare occurrence in ANCA-associated vasculitis. To the best of our knowledge, renal artery aneurysms have previously been reported in only three cases of MPA in the English literature [8-10], of these, only one included rupture of renal artery aneurysm and active bleeding. Although arterial angiography was regrettably not performed to confirm the presence of aneurysms because of urgent condition and unstable signs. It is still rare to report that spontaneous rupture of kidneys can be caused by MPA and these data further demonstrate that medium arteries and veins may also be affected by MPA. In addition, the severity of this case is unprecedented, hemorrhagic shock caused by spontaneous rupture of kidneys which was triggered in turn due to MPA has never been reported before.

**Table 2.** Changes of vital signs and laboratory tests before and after treatment

| *Indicators* | *On the day of spontaneous renal rupture* | *After methylprednisolone usage for 3 months* | *After methylprednisolone usage for 15 months* |
|---|---|---|---|
| *Blood pressure* | 90/60 mmHg | Normal | Normal |
| *Heart rate* | 90 bpm | Normal | Normal |
| *Hemoglobin* | 29 g/L | 126 g/L | 128 g/L |
| *Platelet* | $407\times10^9$/L | $312\times10^9$/L | $260\times10^9$/L |
| *Serum creatinine* | 480 μmol/L | 94 μmol/L | 67 μmol/L |
| *eGFR (ml/min*1.73 m$^2$)* | 8.62 | 56.59 | 83.64 |
| *PRO* | 1.17 g/24 h | 0.5 g/24 h | 0.288 g/24 h |
| *Albumin* | 26 g/L | 37 g/L | 46 g/L |
| *ANCA-P* | Positive | Positive | Negative |
| *ANCA-C* | Negative | Negative | Negative |
| *Myeloperoxidase* | Positive | Positive | Negative |
| *Proteinase 3* | Negative | Negative | Negative |

Surgical intervention or interventional embolization therapy is the predominant treatment of spontaneous rupture of kidneys [2, 4]. Even so, spontaneous renal rupture still has a high mortality rate. In this case, in addition to emergency management, measures of conservative treatment based on glucocorticoid but not nephrectomy or interventional embolization were taken. Methylprednisolone usage for 3 months normalized injured renal function, anemia, hypoalbuminemia, abnormal urine protein as well as peri-renal hematoma and binaural listening. These data demonstrate that conservative treatment based on glucocorticoid is safe and effective for this patient with spontaneous rupture of kidneys caused by MPA. The 29-month follow-up data show that the long-term result is also effective. In the study of Ishiwatari A et al. mentioned above, emergency arterial angiography was performed, microaneurysm of superior segmental renal artery and the interlobular artery was found, and embolization of the right segmental renal artery was performed using micro-coils [10]. As far as we know, this is the first report of rapid deterioration of renal function and hemorrhagic shock caused by spontaneous renal rupture can be successfully suppressed by conservative treatment based on glucocorticoid. The fact that treatment of methylprednisolone without immunosuppressive agents give this patient complete remission clearly indicates that compared with other cases of vasculitis which predominantly affects capillaries, ANCA-associated vasculitis with renal rupture and hemorrhage might be involved in medium artery and small artery/arteriole. This condition may also be much more sensitive to glucocorticoid treatment. Conclusion: MPA could trigger spontaneous rupture of kidneys without warning and should be included in the differential diagnosis of spontaneous renal rupture. Conservative treatment based on glucocorticoid is safe and effective for the renal rupture caused by MPA.

## Acknowledgements

The work should be attributed to the Department of Nephrology, Tianjin Hospital.

## Disclosure of conflict of interest

None.

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