

Life-threatening bleeding from an aneurysm in the arcuate artery of the kidney as a rare complication of granulomatosis with polyangiitis

Piotr Skonieczny¹, Zbigniew Heleniak¹, Tomasz Liberek¹, Barbara Bułło-Piontecka¹, Bartosz Baścik², Alicja Dębska-Ślizień¹

¹ Department of Nephrology, Transplantology and Internal Medicine, Medical University of Gdańsk, Gdańsk, Poland

² Department of Radiology, Medical University of Gdańsk, Gdańsk, Poland

Granulomatosis with polyangiitis (GPA) is a systemic necrotizing inflammation of small and medium-sized vessels. It most commonly affects the upper respiratory tract and kidneys,¹ but it can also develop in other organs.² Bleeding from aneurysms formed during the disease is a rare complication. In a Japanese review, 21 cases of arterial aneurysms in GPA were identified. In 4 cases, renal arteries or their branches were affected.³ Usually, aneurysms in the course of vasculitis were incidental findings during radiologic examination or was identified after aneurysm rupture.⁴

A 50-year-old, previously healthy man was hospitalized at a district hospital for 1 month due to fever, night sweats, and progressive pedal edema. Blood tests revealed high C-reactive protein (CRP) levels, while computed tomography (CT) suggested pneumonia. The patient's condition did not improve with broad-spectrum antibiotic therapy. Development of kidney failure (increase in creatinine levels from 0.7 mg/dl to 3.7 mg/dl) and progression of pulmonary infiltrates with cavity formation raised the suspicion of GPA, and the patient was transferred to our unit.

On admission, the creatinine level was 4.1 mg/dl; CRP, 211 mg/l; and albumin, 15 g/l; while cytoplasmic antineutrophil cytoplasmic antibody (cANCA) titer was 1:2560 and anti-proteinase 3 antibody, 188 RU/ml. The patient presented with significant shortness of breath, and massive pulmonary infiltrates typical of GPA were noted on a CT scan (FIGURE 1A).

Severe, life-threatening GPA with rapidly progressing glomerulonephritis was diagnosed. On a concomitant use of piperacillin/

tazobactam, co-trimoxazole and fluconazole, the patient started intravenous methylprednisolone, followed by pulse cyclophosphamide and plasma exchange therapy. Hemodialysis was started due to oliguria, overhydration, and lung edema. Vegetations on the aortic valve suggestive of endocarditis were found on echocardiography, and therapy with vancomycin was added.

On the 15th day of hospitalization, the patient experienced severe pain in the right flank, with a significant reduction in hemoglobin levels. Computed tomography angiography revealed a massive hematoma adjacent to the right kidney, with active bleeding from the ruptured aneurysm of the arcuate artery (FIGURE 1B). Numerous

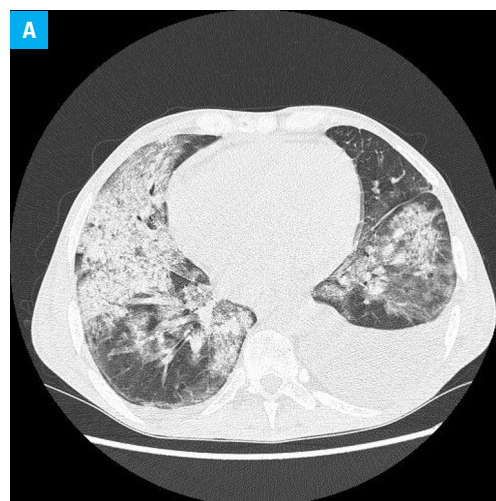


FIGURE 1 A – admission chest computed tomography scan showing massive pulmonary infiltrates

Correspondence to:
Piotr Skonieczny, MD,
Department of Nephrology,
Transplantology and Internal
Medicine, Medical University of
Gdańsk, ul. Dębinki 7, 80–952 Gdańsk,
Poland, phone: +48 58 349 25 05,
email: piotr.skonieczny@gumed.edu.pl
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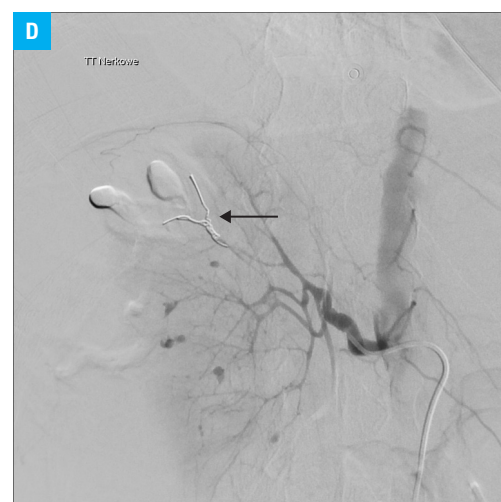
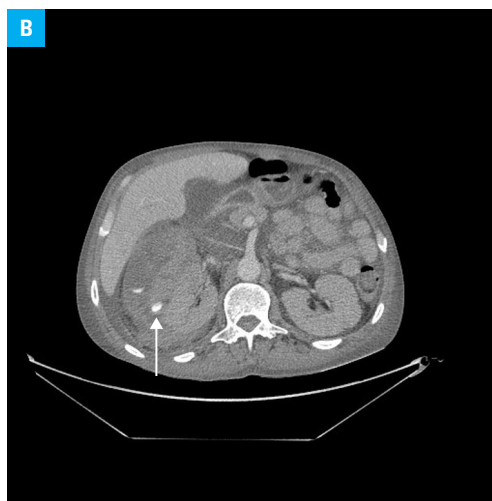


FIGURE 1 **B** – abdominal computed tomography: hematoma adjacent to the right kidney with active bleeding (arrow) on day 15 of hospitalization; angiography of the bleeding vessel: **C** – ruptured aneurysm of the arcuate artery (black arrow) and numerous nonbleeding aneurysms (white arrows); **D** – a scan performed after insertion of embolizing coils (arrow)

nonbleeding aneurysms in both kidneys were also visualized. Urgent embolization of the bleeding vessel was performed, which stopped the hemorrhage (FIGURE 1C and 1D). During hospitalization, control CT was performed twice, showing no further bleeding.

After the third pulse of cyclophosphamide, the patient's condition improved and regression of pulmonary infiltrates was observed, but the patient remained dialysis dependent. Transesophageal echocardiography showed no signs of endocarditis. The patient was discharged home on a reduced dose of steroids. Laboratory tests at 1 month showed cANCA titers of 1:1280 and anti-proteinase 3 antibody titers of 147 RU/ml. Clinically, the patient presented again with endocarditis and pneumonia. His condition deteriorated and he died due to septic shock.

In conclusion, major hemorrhage from an aneurysm of a medium or large-sized artery can be a rare manifestation of ANCA-associated vasculitis. The signs suggestive of internal hemorrhage should raise the suspicion of this complication and prompt urgent diagnostic CT followed by intravascular intervention or surgery. In the present case, the hemorrhage was successfully managed with endovascular intervention, and the immunosuppressive therapy with steroids and cyclophosphamide induced remission

of GPA.⁵ Unfortunately, the patient died of infectious complications, which is the most common cause of mortality in that population.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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