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Authors: Piotr Skonieczny, Zbigniew Heleniak, Tomasz Liberek, Barbara Bulło-Piontecka, Bartosz Baścik, Alicja Dębska-Ślizień

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

Authors:
Piotr Skonieczny\textsuperscript{1}, Zbigniew Heleniak\textsuperscript{1}, Tomasz Liberek\textsuperscript{1}, Barbara Bułło-Piontecka\textsuperscript{1}, Bartosz Baścik\textsuperscript{2}, Alicja Dębska-Ślizień\textsuperscript{1}

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

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

Bleeding from artery aneurysm as a complication of GPA

Corresponding author:
Piotr Skonieczny, M.D.
Department of Nephrology, Transplantology and Internal Medicine, Medical University of Gdańsk, Poland; 80-952 Gdańsk, Dębinki 7, tel. +48 58 349 25 05
email: piotr.skonieczny@gumed.edu.pl

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Granulomatosis with polyangiitis (GPA) is a systemic necrotizing inflammation of small and medium-size vessels. It most commonly affects the upper respiratory tract and kidneys [1], but can also affect other organs [2]. Bleeding from aneurysms formed during the disease is a rare complication - a review of the literature made by the Japanese identified 21 cases with arterial aneurysms in GPA. In four cases renal arteries or their branches were occupied [3]. Usually, aneurysmal vasculitis was discovered incidentally during radiological examination or when they ruptured [4].

50y old men, previously healthy, was hospitalized in the District Hospital for one month with fever, night sweats and progressive pedal edema. The blood test revealed high C-reactive protein (CRP), the computer tomography (CT) scan suggested pneumonia. The patient’s condition did not improve with broad spectrum antibiotic therapy. Development of kidney failure (creatinine increased from 0.7mg/dl to 3.7mg/dl) and progression of pulmonary infiltrates with cavity formation raised the suspicion of GPA and the patient was transferred to our unit.

On admission creatinine was 4.1mg/dl, CRP 211mg/l, albumin 15g/l, cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) 1:2560, anti-proteinase 3 (PR3) titer 188RU/ml. The patient suffered from significant shortness of breath and massive pulmonary infiltrates in the CT scan, typical for GPA, were noticed (Figure 1A).

The diagnosis of severe, life-threatening GPA with rapidly progressing glomerulonephritis was made and in the cover of piperacillin/tazobactam, co-trimoxazole and fluconazole, the patient started intravenous methylprednisolone, then a pulse of cyclophosphamide and plasma exchange therapy. Hemodialysis, because of oliguria, overhydration and lung edema, was started. Vegetations on the aortic valve suggestive of endocarditis were found in echocardiography and vancomycin was added.
On the 15th day of hospitalization the patient had a very strong pain in the right flank with significant hemoglobin lowering. In angio-CT massive hematoma adjacent to the right kidney with active bleeding from the raptured aneurysm of the arcuate artery were identified (Figure 1B). Numerous non-bleeding 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 made twice which did not show further bleeding.

After the third pulse of cyclophosphamide patient’s condition improved, regression of the pulmonary infiltrates was observed, although he remained dialysis dependent. In the transesophageal echocardiography no signs of endocarditis were seen. The patient was dismissed home on a reduced dose of steroids. After one month in laboratory tests cANCA was 1:1280, anti-PR3 titer 147RU/ml. Clinically he presented again with endocarditis and pneumonia, his condition deteriorated and died in a septic shock.

In conclusion, major hemorrhage from an aneurysm of a medium or large-size artery can be a rare manifestation of ANCA-associated vasculitis. Signs suggestive of internal hemorrhage should raise suspicion of this complication, and necessitate urgent diagnostic CT and intravascular radiology intervention or surgery. In the presented case endovascular intervention was successful in management of the hemorrhage, and the immunosuppressive treatment induced remission of GPA by steroids and cyclophosphamide [5]. Unfortunately, patient died of infectious complications, which is the most common cause of mortality in these patients.
Figure 1A. Computer tomography scan of the chest performed on admission shows massive pulmonary infiltrates.
Figure 1B. Hematoma adjacent to the right kidney with active bleeding (white arrow).
Figure 1C. Visualization of the ruptured aneurysm of the arcuate artery (black arrow).

Numerous non-bleeding aneurysms (white arrows).
Figure 1D. Scan after insertion of embolizing coils (black arrow).

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