Inflammation and infection

Spontaneous perinephric hematoma in a patient with Granulomatosis with polyangiitis

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Introduction

Granulomatosis with polyangiitis (Wegener's) is a necrotizing granulomatous vasculitis with predominant involvement of lung, kidney and upper airways. It was reported by Klinger in 1931 at first and then described in detail by Wegener.1

In histopathology it involves venules, capillaries and arterioles but also may affects arteries and veins. It is strongly associated with antineutrophilic cytoplasmic antibodies (ANCA), particularly PR3-ANCA.1,2

A 60 year old man, known case of Granulomatosis with polyangiitis(GPA) with previous history of recurrent sinusitis and history of right side hearing loss and positive titer of c-ANCA that was treated with prednisolone 5 mg daily and methotrexate 5 mg weekly, presented with fever, pleuritic chest pain and productive cough, admitted with impression of pneumonia. Initial creatinine was 1.3 mg/dl. Imaging studies revealed left upper lobe pulmonary cavitary lesion. Therefore treatment with broad spectrum antibiotics, were started. Five days following initial antibiotic therapy he improved with creatinine 1.4 mg/dl. On the eighth admission day, his creatinine increased gradually to 1.7 mg/dl that was accompanied with pyuria. It was proposed that it can be due to drug induced acute interstitial nephritis (AIN). Then his antibiotic changed to other agents with less nephrotoxic effect. On the eleventh day, he got to rapid rise of creatinine to 3.4 mg/dl that was accompanied with deteriorating general condition, malaise and fever. The urinalysis revealed 3+ proteinuria, hematuria and pyuria. c-ANCA was positive with high blood level. Other vascular markers such as p-ANCA, Anti-nuclear antibody (ANA), double-strand DNA (ds-DNA), C-3, C-4 and Anti glomerular basement antibody (GBM), were negative. Methyl prednisolone 500 mg daily for 3 consecutive days was administered. Additional treatments were administration of cyclophosphamide and plasmapheresis.

After first dose of prednisolone pulse therapy, his condition improved and became afebrile. But he got to sudden onset severe right upper quadrant abdominal pain and right side flank pain, about 6 hours after second dose. CT-Angiography revealed subcapsular perinephric hematoma of right kidney about 164 × 78 mm in size (Fig. 1) and a vascular aneurysm about 8mm in upper pole of right kidney (Fig. 2). Finally right side nephrectomy was done due to hemodynamic instability and significant hemoglobin drop.

One day following nephrectomy, third dose of methyl prednisolone and 500 mg Rituximab, were administered. Plasmapheresis with albumin replacement and hemodialysis were administered every other day. During ICU course creatinine concentration had no improving changes and increased to 4.8 mg/dl. On the seventh day after nephrectomy, he got to hemoptysis and alveolar hemorrhage, and cardiopulmonary arrest occurred. CPR was not successful and he expired.

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The first case of spontaneous perinephric hematoma was diagnosed in 1856 that called “Wuderlich syndrome”. A meta-analysis reviewed the hematoma location: 35% right sided, 41% left and 3% bilateral. The most common cause of perinephric hematoma was neoplasms and the second cause was vascular diseases, within this cause, PAN was most common cause.

The clinical manifestation can be with Lenk’s triad; flank pain, hypotension and lumbar or abdominal mass. Common presentations also include hematuria and hypertension.

The most useful diagnostic imaging study is CT scan, as it helps to diagnose the underlying cause, too. Magnetic resonance imaging (MRI) and ultrasound also can be useful. Then angiography is useful if a vascular cause is suspected. Repeated imaging after hematoma resolution is important to rule out underlying malignancy.

We described a rare case of granulomatosis with polyangiitis (GPA) complicated by spontaneous perinephric hematoma which occurred soon after initiation of immunosuppressant therapy. The usual course of GPA is granulomatous inflammation of small and medium size vessels.

Our patient presented chronic sinusitis and involvement of middle ear with conductive deafness, initially. Also had high titer c-ANCA directed against proteinase 3 (anti-PR3) that confirms Granulomatosis with polyangiitis. The biopsy findings after nephrectomy confirm the pattern of crescentic glomerulonephritis that was accompanied with high titer of c-ANCA (Fig. 3). These data confirm the diagnosis of GPA as the main etiology of RPGN in this case. But presence of vascular aneurysm is a rare manifestation of GPA.

The first case of GPA with spontaneous perinephric hematoma was reported by Backer et al., in 1978 that was a 24 year old man with multiple bilateral renal arterial aneurysms. Hartman reported a case with lethal bleeding from a ruptured renal arterial aneurysm in GPA in 1987. In 1995, Aoki et al., reported a 50 year old man with massive intraperitoneal hemorrhage from a ruptured aneurysm that died. Bakker et al., Moutsopoulos et al., and Pumpe et al., reported cases with GPA and spontaneous perinephric hematoma.

Conclusion

The great dilemma of these case reports is that, is it necessary to perform precise vascular assessment for every patient that got to GPA or not. According to possible chance of life threatening hemorrhagic events following vascular insults, it may be logical to perform screening vascular study in every patient with diagnosis of GPA to prevent such lethal vascular insults. So we suggest further studies to confirm this concept that, is it really rational to perform screening vascular imaging study in every patient with diagnosis of GPA or not?
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