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Granulomatosis with polyangiitis presenting as cholangitis and acute kidney injury

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Case Study: Granulomatosis with polyangiitis (GPA) is a small-vessel vasculitis, but it does not usually involve medium-size vessel. A 74-year-old male visited the emergency department for dyspepsia and fever unresponsive to digestive medicine. The laboratory data were as follows: white blood cell, 16210/μL; serum creatinine, 4.30 mg/dL; amylase, 1115 IU/L; lipase, 1161 IU/L; and C-reactive protein, 176.97 mg/L. Computed tomography showed edematous changes around common bile duct (CBD). Magnetic resonance cholangiopancreatography (MRCP) showed CBD dilatation and renal cysts (Fig. 1a). His symptoms improved after endoscopic nasobiliary drainage. However, serum creatinine level increased to 8.50 mg/dL after two weeks. We performed renal biopsy during hemodialysis. The pathologic results showed granuloma with fibrous crescent (Fig. 1b). Serum cytoplasmic antineutrophilic cytoplasmic antibody was positive. GPA was diagnosed based on above results. We performed high dose-steroids and cyclophosphamide treatment concurrently with hemodialysis. Two months later, magnetic resonance imaging (MRI) was performed. Interestingly, as compared to previous MRCP, contrast-enhanced lesions with increased number and size were observed in the kidneys, and a contrast-enhanced lesion was observed in the pancreatic head. The lesions on the kidney were arterial aneurysms (Fig. 2). The immunosuppressive agents were continued, but pneumonia developed, and progressed to sepsis and death. GPA is a systemic vasculitis characterized as necrotizing granulomatous inflammation. GPA presents as a phenotype of rapidly progressive glomerulonephritis in the glomeruli. However, GPA of a long duration can lead to medium-sized arterial aneurysms. Abdominal symptoms in this case might result from the pancreatic aneurysm. The pancreatic aneurysm may result from GPA because the renal cysts of the MRCP was identified to be arterial aneurysms caused by GPA. Arterial aneurysms are rare in GPA. However, chronic inflammation of GPA can lead to aneurysm of the middle artery. When patients present with abdominal symptoms and acute kidney injury, clinicians need to consider systemic vasculitis.

Figure 1. (A) Computed tomography demonstrates showed only edematous change and central hypo-dense lesion around common bile duct and pancreatic head portion T2-weighted image of magnetic resonance cholangiopancreatography showed distended gallbladder and dilated common bile duct and several tiny renal cysts. (B) The pathologic finding of the kidney demonstrates fibrous crescent and diffuse mesang

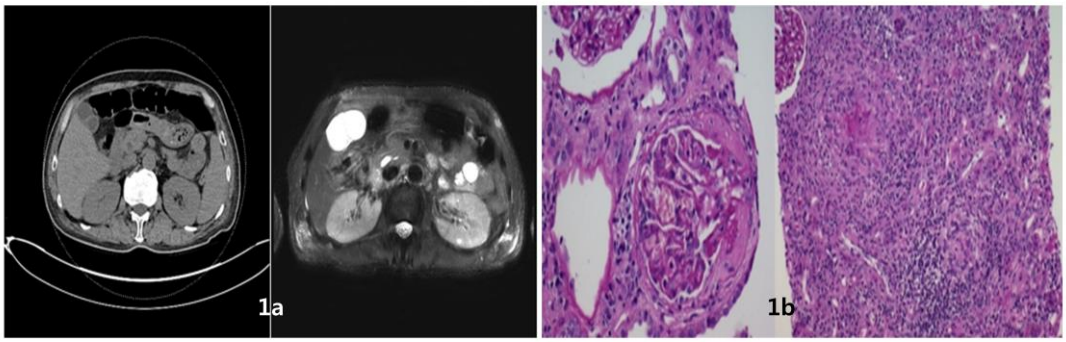

Figure 2. T1-weighted image of magnetic resonance imaging showed contrast-enhanced nodular lesion with various sizes on the both kidney. Renal artery angiography shows multiple microaneurysms on the interlobular or arcuate arteries without rupture.