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A Case Of Granulomatosis With Polyangiitis Accompanied By Cerebral Aneurysm: Possibility Of IgG4-related Disease Overlap Syndrome.

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Case Study : Granulomatosis with polyangiitis (GPA) is an ANCA-associated vasculitis (AAV) that affects in small sized blood vessels. GPA is characterized by involvement of respiratory tracts, kidney, and any organ. IgG4-related disease (IgG4-RD) is a systemic fibroinflammatory disease by infiltration of IgG4 plasma cells. Although no always common, both diseases can present common features. We report a case of GPA with which possibility of an IgG4-RD overlap syndrome cannot be ruled out. A 47-year-old female visited due to weakness and both lower legs edema accompanied by petechiae for one month, followed by bilateral flank pain and hemoptysis. Her vital signs were stable. Initial laboratory tests showed followings: serum creatinine, 6.0mg/dL; $HCO3^-$, 15.6mmol/L; anti-PR3 ANCA, 158U/mL; IgG subclass IV, 106.5mg/dL. CT findings showed abdominal aortitis and retroperitoneal fibrosis, and bilateral hydronephrosis due to both ureteral strictures were also shown. After percutaneous nephrostomy, clinical symptoms and signs were marked improved but, generalized edema and proteinuria were worsened, so we performed kidney biopsy. On the kidney biopsy, fibro-cellular crescents were observed in 16 of 21 glomeruli, and the GBM showed normal thickness. Several days after starting induction treatment with high-dose glucocorticoid and cyclophosphamide for GPA, she abruptly died from subarachnoid hemorrhage secondary to anterior communicating artery rupture. Structural changes in the blood vessels in GPA may occur as vasculitis worsens, but it is rare to report that GPA affecting small arteries is associated with aneurysm development or rupture. She showed clinical features such as aortitis, retroperitoneal fibrosis, and cerebral aneurysm, which are difficult to explain in patients with GPA affecting small arteries. GPA has a poor prognosis due to irreversible organ failure. If IgG4-RD is also accompanied by cerebral aneurysm, the prognosis may be poor. Thus, if AAV patient presents with features suggesting the possibility of another disease, a more aggressive investigation should be performed.

Figure 1. .jpg

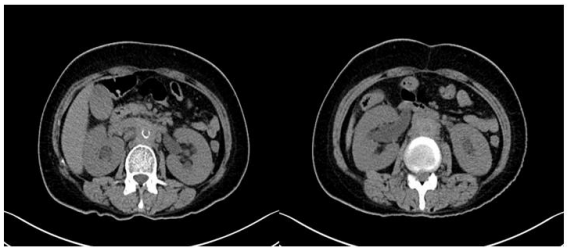

Figure 1. .jpg