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Immunoglobulin G4 연관 타액선염과 동반한 후복막 섬유증과 급성신손상 1예

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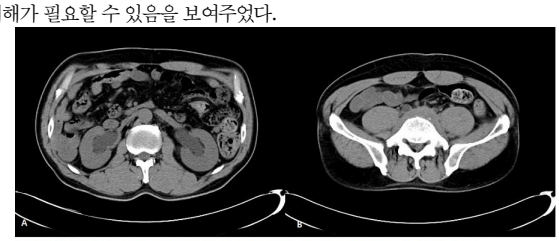
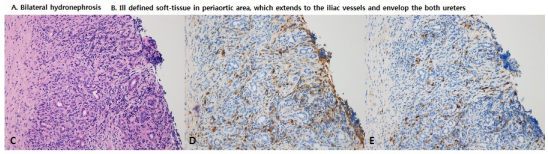
서론: IgG4 연관 질환은 IgG4 양성 형질세포가 풍부한 림프구의 침윤과 다양한 정도의 섬유화가 특징적이며 주로 췌장, 담관, 침샘, 눈물샘, 후복강, 갑상선, 폐, 간, 신장 등을 침범, 한가지 이상의 장기에서 발현되는 경우도 종종있다. 본 저자는 IgG4 와 연관된 타액선염 환자에서 후복막섬유증과 급성신손상이 동반 되어진 1례를 경험하여 이를 보고하고자 한다. **증례:** 58세 남자가 6개월전부터 지속되는 허리 및 좌측 다리로 방사되는 통증으로 시행한 MR spine상 L2/L3 spinal stenosis로 진단, 신경외과에서 시행한 수술 전 검사에서 혈중 요소질소 43.1mg/dL, 크레아티닌 4.02mg/dL 로 측정되어 신장내과로 의뢰되었다. 과거력상 2년전 좌측 턱밑샘에 1cm 크기의 종괴로 CT 및 조직생검으로 IgG4연관 타액선염 진단받은 후 절제술하였다. 진찰에서 경부, 흉부, 복부 검진시 이상소견 없었고 grade 2 이상의 양쪽 하지 부종이 있었다. 혈액검사 결과는 혈색소 9.2g/dL, 혈중 요소질소 41.8mg/dL, 크레아티닌 4.06mg/dL(6개월전 0.77mg/dL), IgG4는 357.0mg/dL(참고치, 3.9-86.4mg/dL)로 측정되었다. 전산화단층 촬영상 양측 수신증과 대동맥에서 장골혈관까지의 주변 연조직들이 양측 요관을 감싸 요관이 좁아진 소견으로 후복막 섬유증이 확인되었다. 2년전 IgG4 연관 타액선염을 진단받았던 환자로 후복막 섬유증 및 당시 혈액 IgG4 level 357로 측정되어 IgG 연관된 후복막 섬유증으로 인한 급성 신손상으로 진단하고 프레드니솔론(prednisolone) 60mg을 투여하였고, 크레아티닌 감소 소견은 있었으나 신장 초음파 경과에서 수신증 호전없어 D-J catheter를 삽입 후 퇴원하였다. 치료 4주 경과 후 혈액검사에서 혈중 요소질소 34.0mg/dL, 크레아티닌 1.95mg/dL였으며 양측 다리 부종 및 허리 통증은 호전되었다. **결론:** 본 증례는 다른 침범장기의 생검과 혈청 IgG4 level을 통해 IgG 연관된 후복막 섬유증으로 인한 급성 신손상을 진단 후 치료한 사례로 IgG 연관 질환의 다장기 침범에 대한 이해가 필요할 수 있음을 보여주었다.



A. Bilateral hydronephrosis B. Ill defined soft-tissue in periaortic area, which extends to the iliac vessels and envelop the both ureters



The biopsy specimen(C) show lymphoplasmacytic infiltration with septal fibrosis. IgG4 and IgG immunohistochemical staining
Immunostaining of IgG4 (E) and IgG (D). IgG4/IgG-positive plasma cell ratio is more than 40%

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Immunoglobulin G4-related kidney disease presenting as a renal infarction

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Introduction: Immunoglobulin G4-related disease is a systemic immune-mediate disorders shared particular clinicopathologic feature. Tubulointerstitial nephritis (TIN), most common form of IgG4-related kidney disease, present characteristic radiologic findings may be confused other renal disease. We report a case of IgG4 related TIN, initially misdiagnosed as a renal infarction. **Case presentation:** A 67-year-old man was consulted to the nephrology department because of azotemia. He was diagnosed as chronic pancreatitis 5 years ago. He was followed up with laboratory tests and computed tomography by a gastroenterologist. Recently, his serum creatinine was elevated from creatinine level of 1.10mg/dl to 2.05 mg/dl over the last 8 months. He complained no other symptoms except chronic fatigue. On computed tomography, multifocal low attenuation lesions with atrophic changes in both kidneys were revealed. However, renal arteriography demonstrated no abnormal finding in both renal main arteries and intrarenal arteries. Laboratory findings disclosed: ANA positive, complement C3: 70mg/dL, C4: 3 mg/dL, serum IgG4: >340mg/dL, anti-phospholipid antibody IgM/IgG: 162.7/18.2 U/mL, BUN/Cr 23/2.22 mg/dL. Histopathologic examination showed severe tubular atrophy and diffuse interstitial fibrosis with dense infiltration of lymphocytes, plasma cells and neutrophils. The IgG4+/IgG ratio was over 50%. He was treated with 30 mg of prednisolone. Prednisolone tapered gradually and oral mycophenolate mofetil was added. serum creatinine level decreased and C3, C4 and IgG4 levels were normalized progressively. **Conclusion:** In the case presented here, the well demarcated, wedge-shaped poor enhanced cortical areas were detected upon imaging evaluation, thus mimicking renal infarction. Conventional imaging has limitations in diagnosis of IgG4-TIN. A multidisciplinary approach, especially kidney biopsy, is needed for correct diagnosis.

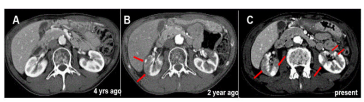

Figure 1. Time series of contrast enhance abdominal Computed Tomography (CT). Multifocal low attenuation lesions in both kidneys were gradually progressed. 4 years ago (A), 2 years ago (B) and present (C).

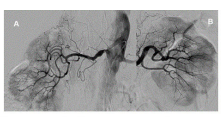

Figure 2. Renal angiography. There was no abnormal finding in right main renal artery and intrarenal arteries (A) as well as left (B).

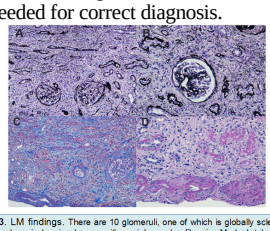

Figure 3. LM findings. There are 10 glomeruli, one of which is globally sclerotic. Some glomeruli show ischemic change with periglomerular fibrosis. Marked tubular atrophy, interstitial inflammation and diffuse fibrosis, consistent with chronic interstitial nephritis. Nephrosclerosis, moderate. Methenamine silver (A,B), Masson's trichrome (C), PAS (D).

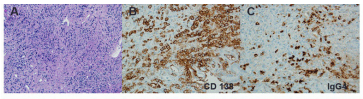

Figure 4. Dense interstitial infiltration of lymphocytes and plasma cells are shown (A, PAS). Representative IHC findings : Many plasma cells (CD138 +) are infiltrated in the fibrotic interstitium (B), >102 IgG4 (+) cells per high power field are seen (C).

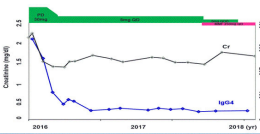

Figure 5. Time series of serum creatinine and IgG4 level. He was treated with 30 mg of prednisolone. Prednisolone tapered gradually and oral mycophenolate mofetil was added. His creatinine and IgG4 level have remained stable.

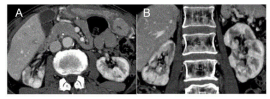

Figure 6. Follow up CT findings after 36 months. Markedly decreased multifocal cortical low attenuation lesions in size and extent with atrophic changes in both kidneys were demonstrated on transverse section (A) and coronal section (B) compared to initial CT findings (**Figure 1**).