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IgG4-Related Disease in a patient with Hodgkin`s Lymphoma in remission: A case report.

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Background:

IgG4 – related disease (IgG4-RD) is a multisystem fibroinflammatory disease affecting the kidney and other different organs. It affects the kidney through inflammatory interstitial nephritis and retroperitoneal fibrosis. The presentation of lymphoma and IgG4 disease in the same setting is not common. Most of the reported cases were mucosa-associated lymphoid tissue (MALT) lymphoma in IgG4 ocular disease. In this abstract, we are presenting a case with IgG4 disease in a patient who had Hodgkin lymphoma.

Method:

A male 68-year-old diabetic gentleman was referred urgently to our nephrology clinic with rapidly worsening renal function. He had been treated for stage IVB Hodgkin`s lymphoma - nodular sclerosing two years previously with ABVD (Doxorubicin/Adriamycin, Bleomycin, Vinblastine, Dacarbazine) and was in full remission. At the time of referral, he was under hepatobiliary team investigating painless obstructive jaundice. Bile duct biopsies showed 15% of IgG4-expressing plasma cells. On review in the renal clinic, he was asymptomatic and examination was unremarkable.

Results:

Serum creatinine had risen from 119 to 293 µmol/l. Calcium was normal. He had a urine protein-creatinine ratio of 55 mg/mmol. ANCA, ANA and myeloma screen were all negative. There was however a significant polyclonal gammopathy with an IgG of 39 (raised levels of IgG1, IgG3 and IgG4). C3 and C4 levels were very low. Renal ultrasound showed unobstructed but enlarged kidneys, the right measuring 143mm and the left 130mm, with suspicion of possible infiltration.

An urgent renal biopsy showed that the kidney was heavily infiltrated with predominantly plasma cells which stained significantly (>40%) for IgG4. Other stains for malignant cell markers were negative. No Hodgkin-Reed – Sternberg cells were identified morphologically or with the aid of CD30 immunohistochemistry. He was diagnosed with acute interstitial nephritis due to IgG4-related disease.

He was treated with high dose steroids, and after two weeks his creatinine fell from a peak of 314 to 158 µmol/l. He remained symptomatically well and reported no side effects except needing to increase his insulin. He continued on a weaning course of steroids.

Conclusion:

Several case reports described the incidence of the lymphoma in patients with a pre-diagnosis with IgG4-RD. Another study has described that lymphoma may precede IgG4-RD, rather than the opposite. In this

case, despite the fact that the IgG4-RD was diagnosed after lymphoma remission, we think that IgG4- RD preceded lymphoma diagnosis. We noticed that serum creatinine started to increase and eGFR to decline in October 2016 i.e. 4 months before Lymphoma diagnosis in January 2017. Most of the lymphoma cases described in case reports were either MALT or diffuse large B cell lymphoma (DLBCL). In this case, we describe a case with Hodgkin`s lymphoma. Increase in IgG M, in this case, is more related to the lymphoproliferative disease rather than to IgG4 disease. Nephrologists should be aware of the possibility of incidence of lymphoma in patients with IgG4- RD. Further studies are required to investigate the relationship between both diseases and which one is a risk factor for the other.