Unexpected presentation of IgG4 related disease
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INTRODUCTION

- Immunoglobulin G4-related disease was first identified in the early years of the 21st century, when Japanese investigators recognized the patients with sclerosing pancreatitis (now known as type 1 IgG4 related autoimmune pancreatitis) have the same histopathological features in extrapancreatic organs as in the pancreas. Since then this disease has been reported in nearly every organ.
- IgG4-related tubulointerstitial nephritis is the most common IgG4-related disease in the kidney which can present as acute or chronic renal insufficiency, renal mass, or both. The hallmark of IgG4 related disease is characterized by the presence of abundant IgG4 plasma cells in affected tissue with interstitial (stornifibrosis),obliterative phlebitis and with or without elevated plasma levels of IgG4. It typically affects older adults, predominantly male. The incidence and prevalence of the disease are not known, because the only studies of patients with IgG4 related disease were thought due to multiple myeloma. It is another plasma cell related disease, has higher levels of incidence, associated with renal failure, bone manifestations (although they do not appear as lytic lesions), and anemia associated with chronic disease and renal disease with suppressed iron.

CLINICAL COURSE

- The patient, a 64-year-old male with past medical history of hypertension, remote smoking history, GSRD and recent C1 level (about 6 weeks ago) brought in by EMS to the ED for evaluation of generalized weakness resulting in multiple falls. The patient was admitted for further evaluation of renal failure of unclear etiology. The initial symptoms include interstitial inflammation and sclerosis noted in the CT of the abdomen and pelvis showed transmural stranding around the kidneys, with bilateral renal enlargement and without evidence of obstruction. The lymphadenopathy was also seen in abdomen and pelvis on CT scan. The CT of head was performed, showed a right frontal polar lesion. MRI on the head was done the next day which showed a lesion which was hypointense on T1 and hypointense in T2 which could represent brain tumor (glioma) vs myelopathy vs fluid level (2nd to renal failure). Contrast enhanced imaging was not performed due to renal failure.
- The hyperkalemia improved with insulin, bicarbonate and AKI. He was started on insulin and once was on insulin, bicarbonate and IVF. He was found to have RBC on urine microscopy, bilateral renal enlargement and moderate interstitial inflammation. Serum measurements of IgG4 were obtained at day 3 of his admission. The serologies were negative for ANA, total LG1, and IgG level was not obtained. IgG100, IgG101, negative for glomerular basement membrane antibodies and normal C3 and C4. He was noted to have elevated serum kappa/lambda light chains. He underwent renal biopsy which revealed IgG4 related disease with moderate interstitial fibrosis. The hemiparesis on CT and MRI showed no new lesion. The patient was placed on 40 mg prednisone daily to continue outpatient follow-up for the foreseeable future given the uncertainty of his long-term renal recovery.

BRIEF SUMMARY OF CASE

- We present you the case of older male who presented with renal failure of unclear etiology requiring hemodialysis.
- He was found to have RBC on urine microscopy, bilateral renal enlargement and lymphadenopathy which led to further work and ultimately diagnosed with IgG4 related kidney disease.
- Renal biopsy showed IgG4 related disease with moderate interstitial fibrosis.

LEARNING OBJECTIVES

- The early recognition of IgG4 related disease can prevent serious organ damage.
- Consider further work up in patient with an atypical presentation.
- IgG4 related disease has indolent and heterogenous nature which makes the diagnosis challenging.

DISCUSSION

- The patient’s lymphadenopathy, new CT of abdomen and pelvis, acute renal failure, and possibly electrolyte abnormalities, are thought to be secondary to IgG4 related disease. The patient’s unexplained elevated triglycerides of 380 mg/dl is concerning. Further imaging as well as acute kidney injury work up that was disappointing with lymphadenopathy may have been secondary to lymphoma, chronic inflammation, crush myopathy, paraneoplastic, or myeloproliferative disease. Although none of these diagnoses lymphoma with acute renal failure was now evident. There was no known source/exposure for potential parasitic infection, as well as no indication of a total 4001 protein of any cell line. In our patient’s case, the most probable cause of his IgG4 related disease was thought due to multiple myeloma. It is another plasma cell related disease, has higher levels of incidence, associated with renal failure, bone manifestations (although they do not appear as lytic lesions), and anemia associated with chronic disease and renal disease with suppressed iron.

CONCLUSION

In medicine we are most often confronted with what is referred to as “bread and butter” medicine and common conditions, and to think of them as horses rather than their more exotic counterparts’ zebras. However, we may come across a savannah with a difficulty in explaining constellation of symptoms and presentation with clear black and white stripes. Therefore, it is important to consider further work up in patient with an atypical presentation.

REFERENCES & ACKNOWLEDGEMENTS

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