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IGG4-RELATED DISEASE : ANALYSIS OF TWO CASES PRESENTING WITH ABDOMINAL PAIN AND ACUTE INFLAMMATORY RESPONSE

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BACKGROUND

Ig-G4 related disease (IgG4-RD) is a recently recognized immune-mediated systemic disease that can affect any organ or system, most often the pancreas, biliary tract, salivary and lacrimal glands and the aorta. It is a fibroinflammatory condition that often presents as a tumefactive lesion, with an IgG4 plasma cell organ infiltration and often, but not always, elevated levels of circulating IgG4. Generally, patients with IgG4-RD have a subacute clinical presentation, with only less than 10% presenting with acute symptoms, increase of acute phase reactants and other manifestations of an acute inflammatory response. The histological hallmarks for IgG4-RD include lymphoplasmacytic infiltration, storiform fibrosis and obliterative phlebitis. The elevation of IgG4+/IgG+ ratio and of IgG4+ cells per high powered field is highly suggestive of IgG4-RD. Diagnosis relies on the coexistence of various clinical, laboratory and histopathological findings, although none is pathognomonic.

CASE REPORT

We present two patients admitted last year to our Rheumatology department for constitutional syndrome, with asthenia, fatigue and weight loss. They presented with increased levels of acute phase reactants, mainly erythrocyte sedimentation rate (ESR) and C-reactive protein, as well as microcytic hipocromic anaemia. Both patients presented with abdominal pain – the first, a male of 48 years old, had pain predominantly in the left hypochondrium aggravated in the postprandial period, and the second, a female of 43 years old, had colicky pain in the right iliac fossa. In both cases, other etiologies such as cancer, pancreatic or bile duct inflammation were excluded. On computerized tomography (CT) there was evidence of several upper abdominal adenopathies in the male patient and a large adenopathy next to the cecum in the female patient. The biopsies showed lymphoplasmocytic infiltration, with an increased population of IgG4 plasmocytes and a raise in the IgG4+/IgG+ plasmocytes ratio of 50%, which associated to the elevation of serum IgG4. These results, combined with the clinical manifestations, allowed for the diagnosis of IgG4-RD to be made.

CONCLUSION

The authors emphasize the importance of theses cases, both with an atypical presentation – with abdominal pain but without evidence of pancreatic or biliary inflammation and exuberant acute inflammatory response. Although IgG4- RD is a rare condition, increasing awareness has been developing towards it in the scientific community. Therefore, it should be considered as a differential diagnosis of abdominal pain in the context of abdominal lymphadenopathy. The authors also highlight the importance of searching for typical histological findings, when there is a clinical suspicion.