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INFLAMMATORY PSEUDOTUMOR RELATED TO IGG4: DIFFERENTIAL DIAGNOSIS OF LYMPHOPROLIFERATIVE DISEASES

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BACKGROUND

IgG4-related disease (IgG4-RD) is a systemic autoimmune disorder that affects multiple organs with clinical presentation in four phenotypes: pancreate-hepato-biliary, retroperitoneal fibrosis/ aortitis, limited to head and neck, and systemic involvement. The aim of this article was to describe a case of DR-IgG4 and contribute to a better understanding of this disease.

CASE REPORT

Female patient, 44 years old, with lower back pain and weight loss (10kg in 4 months). Abdominal Computed tomography revealed attenuation of soft parts tissue, anterior to the abdominal aorta, with expansive involvement. The abdominal magnetic resonance imaging rshowed periaortic tissue with expansive aspect, suggesting neoplastic lymphoproliferative injury. After exploratory laparotomy, the presence of infra-umbilical lymph node of 2cm diameter, small ascites and palpable retroperitoneal mass (7cm) was reported. The mass was biopsied and revealed Storiform myofibroblasts or fibroblasts proliferation in fibrous stroma with extension to perinodal tissues, mixed vascular proliferation and inflammatory infiltration consisting of plasma cells, lymphocytes, histiocytes and neutrophils characterizing aInflammatory myofibroblastic tumor (inflammatory pseudotumor). Immunohistochemistry: Presence of Antibodies Ki-67 <1%, CD138, IgG4 (MRQ-44-30 cells / CGA), IgG, being histologically IgG4 related disease. Serum IgG4 dosage: 68.7 (Reference Value: 3 - 201), ESR: 79. The patient was treated with prednisone 1mg/ kg/ day and rituximab evolving with clinical and laboratory improvement.

CONCLUSION

The pathogenesis of IgG4-RD is unclear. There is a decrease in Thelper1/ Thelper2 cells and

increase in TRegulators, with co-activation of B cells expressing IgG4, but their role is unknown, probably play an anti-inflammatory role in response to a trigger.

Clinical manifestations may occur in almost all organs or systems. Patients often have a mass on the affected organ, such as a retroperitoneal mass or diffuse organ enlargement. Retroperitoneal fibrosis is one of the most common presentations of IgG4-RD, responsible for the most cases of retroperitoneal fibrosis previously considered "idiopathic".

Diagnosis is based on histopathological, clinical, serological and radiological findings. However, when a retroperitoneal fibrosis manifests, it is essential to make the differential diagnosis with lymphoproliferative diseases, due to the similarity in the radiological findings. Histopathology shows dense lymphoplasmacytic infiltrate, fibrosis, and obliterative phlebitis is somewhat characteristic and the serum increase of IgG4 is present in only 60% of the cases.

The treatment of IgG4-RD may change according to the affected organ, and glucocorticoids are the first line, but reports indicate long-term benefits with rituximab.