Abstract

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Lymphoproliferative Disease in Retroperitoneal Fibrosis: a Case Report

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Background: Idiopathic retroperitoneal fibrosis, named Ormond disease, is a rare, connective tissue disease. In the idiopathic form, in over 70% of the cases, the growth of the fibrous tissue involves mediastinic site, orbital tissue, biliary tract, thyroid. In more one third of the cases retroperitoneal fibrosis is due to several causes: infections, trauma, radiation, drugs, neoplasms. The idiopathic form is due to IgG4 related disease, a sistemic fibro-inflammatory disease characterized by infiltration of IgG4 positive plasma cells in the affected tissues, with or without elevated plasma levels of IgG4.

Case history: We report a clinical case of a patient 70 years old, affected by dilatative cardiomyopathy with sistolic disfunction, diabetes, hypertension. He is hospitalized for dyspnea. In the chest X ray left pleural effusion, so for that reason he did a CT scan that showed us mediastrinic and retroperitoneal fibrosis and multiple district lymphadenomegaly. At blood chemistry tests mild anemia, mild leukopenia with lymphocytosis, increase of tumor markers. At immunofixation monoclonal component IgG lambda in gamma region. The flow cytometry with monoclonal antibodies showed a lymphoproliferative disease.

Conclusion: This is a rare case of secondary fibrosis due to lymphoproliferative disease although monoclonal component IgG. Differential diagnosis is very hard.