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IgG4-related disease – 1 Case Report Patient

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Abstract

Background

The IgG4-related Disease is an auto-immune fibroinflammatory condition that can attack several organs. The diagnostic is based on the tendency of forming lesions in multiple sites and the characteristic histopathological appearance and, sometimes, high seric levels of IgG4. The base of treatment is immunosuppression, mainly using corticoids, it can be used rituximab too. In this article, we related a case of a 31-years-old patient of female sex with IgG4-Related Disease with cutaneous and systemic impairment, mainly in the Central Nervous System, Abdominal and Ocular Regions.

The IgG4-related Disease was first described as a clinic-pathological entity in 2003 in Kamisawa et al study in a case of auto-immune pacreatitis1. The IgG4-related Disease was an auto-immune systemic disease that can have as manifestations the impairment of pancreas, bile ducts, lymph nodes, prostate, retroperitoneum, salivary glands, lacrimal glands, orbital fabrics, kidneys, liver, meninges, lungs, aorta, pericardium, breast, thyroid and skin and others (1,2).

Epidemiologically, this affects more men, normally in the ages of sixties and seventies and the most common organ effect described was the pancreas (2).

Clinically, it was described as a disease with variable evolution, according to the acommeted organs in each patient (1,2). With the passing years, there is an increased risk to evaluate to neoplasias, such as stomach, lungers, prostate, colon, non Hodgkin's lymphoma, bile ducts and thyreoid (1,2,3). It can evaluate with kidney's chronic disfunction and kidney failure. The prognostic is not very well defined (2).

***Objective**: This article intends to relate a single case report of IgG4-related Disease with cutaneous and extra-cutaneous manifestations.*

***Keywords**: Cutaneous, Disease, Extra-cutaneous, IgG4, Manifestations, Related.*

Materials and Methodology:

In this article, it will be described a case report and a revise of articles in Pubmed and Scielo.

Single Case Report

Patient, woman, 31 years, she referred in specific systemic impairments, as esporadics fevers non-related to bacterial infections, malaise, back's, arms', legs' and abdominal's pains, articles' pain with no signal of arthritis and weakness. When she was 26 years old, she developed a generalized convulsive crisis and had a stroke at the same age as a sequel to a light weakness of the left arm. It was done investigation for Lupus and Antiphospholipid Antibody Syndrome and the exams were negative.

At the age of 27 years old, she had keratoconus and acute hydrops of fast evolution in the right eye, and she lost her vision on this side. After three months, she had the same problem in the left eye and received corneal transplantation on this side.

Concomitantly, the Computadorized Tomography of Orbital Tecid has the signal of myositis muscle, thickening of orbital fat, and lacrimal glands. After three years of transplantation, she developed a cataract of the left eye, she is waiting for surgery to correct this.

At the age of 29 years old, she had an erythematous, itchy and swollen plaque in the lateral region of the left eye, after using 60 mg of prednisone, it turned to a light-brown macle in this region.

Concomitantly with the skin lesions, she has had to worsen of abdominal's pain, and she has done another tomography and has several abdominal lymph nodes.

She has done biopsies of the lacrimal's left gland and abdominal's lymph node. And in both of these, the result was abundant linphoplasmocitary infiltrate, presence of eosinophils, areas of focal fibrosis and plasmacytosis with the immunohistochemistry with the proportion o IgG4+/iGg+> 80%. I was compatible with de the IgG4-related disease. And the seric level of IgG was 79.000 Ui/ml.

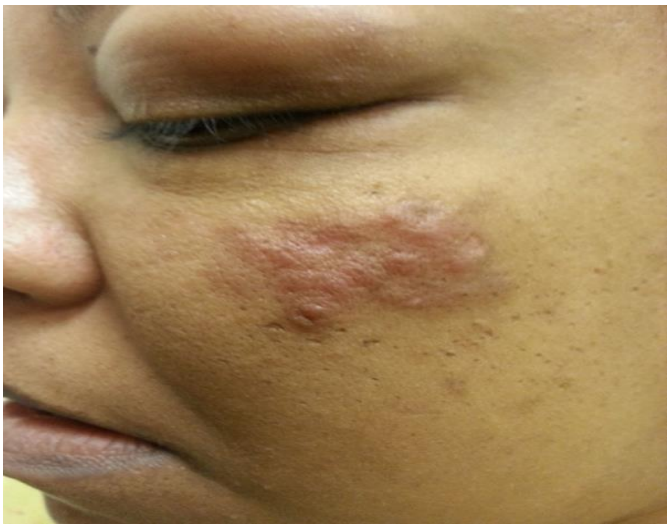

Illustration 1: Skin lesions, initially. Erythematous and multiple papules in malar left region.

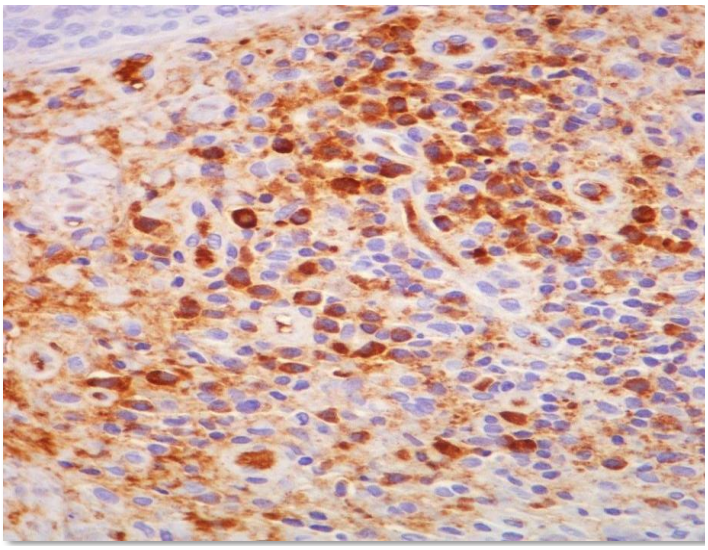

Illustration 2: Immunohistochemistry of skin lesion for phenotipage of phlasmocytes.

Results

The IgG4-related disease has a large variety of clinical spectrum (1,2,5).

In the period of 4 to 7 October in 2011, there had been a symposium in Boston to make a Consense about the necessary criteria to establish the diagnostic of IgG4-related disease. In this consense, there has been established that besides soiled lesions in multiple sites, it is necessary the presence of two other histopathological major criteria, between these, are: soft lymphoplasmocitary's infiltrated, focal's fibrosis and obliterative phlebitis. These major criteria are harder to be found in lymph nodes, lungers' tecids, saliva's and lacrimal's glands. Between the minor criteria are: phlebitis without obliteration of lumen and an increased number of eosinophiles (5). Other relevant criteria to addition the diagnostic would be the relation of IgG4+ > 40 % in any tecid, It can have elevated seric levels of IgG or not (1,2,5).

Among the presented manifestations of this case, the report is important to know that skin lesions can occur later and rarely preceded the systemic manifestations. And these lesions are described as papules, plaques or nodes, usually erythematous and itchy in head or neck areas that improve with immunosuppression. There have never been described macles or bullies as initial lesions (7).

The ophthalmic impairment can present a large variety of manifestations and can englobe more frequently lacrimal's glands. But, the patient can present uveitis, episcleritis, muscular, nervous and osseous impairment and eyelids (8).

The lymph node impairment is very frequent, mainly in abdominal and aortic regions, and can simulate neoplasias and compress structures around (1,2).

The impairment of the nervous system is not very described and it is postulated that can be a consequence of expansive's lesions. And it can have consequences paralyzes and paraplegias (9).

The base of treatment is immunosuppression, normally with prednisone in high doses as 1-1,5 mg/kg per day. And rituximabe can be necessary. And according to the acommeted organs, it can be necessary complementary treatments. After the diagnostic, the following and the trace of lesions in kidneys, eyes and neoplasias are indispensable (1,2,5,7).

Conclusion

Besides being a rare disease, it is well established the diagnostic criteria. And it has to be thought in patients that have other systemic manifestations to program a better approach and proportionate a multidisciplinary attend to the patient.

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