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Case Study
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IgG4 Odyssey: Unraveling a Complex Disease Presentation Through Comprehensive Diagnosis and Interdisciplinary Management

Montoya P and Barrientos W*

Department of Internal Medicine, Clínica Vida, Medellín, Colombia

Abstract

This case report outlines a unique presentation of IgG4-related disease, a 53-year-old female with a history of non-oxygen-dependent Chronic Obstructive Pulmonary Disease (COPD) presented with bilateral eyelid edema, nasal congestion, and posterior discharge. A blepharoplasty revealed reactive lymphoid follicular hyperplasia, and subsequent imaging, including orbital MRI, CT of the neck-chest, and abdominal MRI, demonstrated extensive involvement of various anatomical structures.

Clinical History

The patient, a 53-year-old female with a background of non-oxygen-dependent Chronic Obstructive Pulmonary Disease (COPD), presented five years ago with a constellation of symptoms, including bilateral eyelid edema, nasal congestion, and posterior discharge. Notably, the patient underwent an ambulatory blepharoplasty, during which a biopsy was performed, revealing reactive lymphoid follicular hyperplasia.

Imaging Findings

Subsequent investigations included an orbital MRI, which unveiled a soft tissue mass exhibiting marked hypointensity in T2-weighted sequences. This mass not only affected the orbital cavities but also extended to various anatomical structures, encompassing frontal epicranial soft tissues, the nasal cavity (particularly around the septum), premaxillary spaces, Bucco-maxillary spaces, portions of masticatory spaces, and the right temporal epicranial region. These findings were indicative of IgG4-related disease [1-5].

Referral and Initial Examination

The patient was promptly referred from the primary care provider to an oncology clinic, emphasizing the need for further evaluation. On admission, routine blood chemistry and a complete blood count reported within normal limits. Evaluation by the internal medicine service highlighted bilateral bi-palpebral edema, erythema, and a distinctive brown discoloration. Additionally, a scant serous conjunctival secretion and cervical adenopathy's, predominantly in station II on the left, measuring approximately 1 cm, were noted (Figure 1).

Further Radiological Assessment

In response to the diagnostic suspicion, a comprehensive neck-chest CT was performed, revealing thickening of periorbital soft tissues and involvement of bilateral malar and right cervical regions. Bilateral parotid gland enlargement, pansinus involvement, and adenomegalies in cervical stations I, II, III, IV, and V was evident. Notably, a thoracic CT demonstrated peribronchovascular thickening, septal lobular thickening, bilateral bronchial wall thickening, mediastinal adenopathy's, and proximal esophageal wall thickening.

To further delineate the extent of the disease, an abdominal MRI was conducted. The findings suggested the primary possibility of IgG4-related disease with initial involvement of the biliary tract. A pattern indicative of cholangitis sclerosant, autoimmune pancreatitis, periaortitis with initial retroperitoneal fibrosis, subcentimeter bilateral renal pseudotumors, and reactive abdominopelvic lymph nodes, with signs of chronic thrombosis at the splenic hilum, were discerned [6-12].

Laboratory Results

Extensive laboratory investigations were conducted, including a panel for viral markers (HVB,

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Correspondence:

Pablo Montoya, Department of Internal Medicine, Clínica Vida, Medellín, Colombia,

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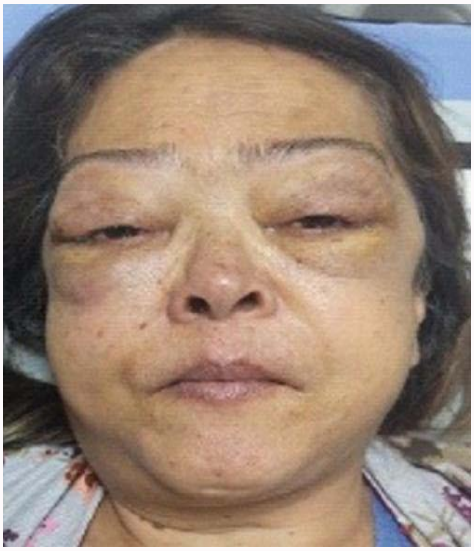

Figure 1: Bilateral palpebral edema and bi-palpebral brown discoloration.

HVC, RPR) yielding negative results. Autoimmunity profile for ANA, ANCA, ASMA, and AMA was also negative. Notably, the patient exhibited elevated IgG4 levels (4900 mg/dL), along with suboptimal vitamin D levels (17 ng/mL) and normal vitamin B12 levels (195.54 pg/mL).

Diagnostic Conclusion

Given the suggestive imaging findings and elevated IgG4 levels, the patient was diagnosed with IgG4-related disease. The management plan included a course of pulsed steroids (methylprednisolone 500 mg for 3 days), Ursodeoxycholic Acid (UDCA) for cholangitis sclerosant, anticoagulation for chronic splenic hilum thrombosis, a 7-day antibiotic regimen for pansinusitis, and supplementation for vitamin D and vitamin B12 deficits.

Clinical Outcome

Following the completion of the steroid pulse regimen, the patient exhibited a significant improvement in her overall condition. Consequently, she was discharged with a comprehensive outpatient management plan, including high-dose oral steroids, UDCA, and anticoagulation. Ambulatory referrals were made for further evaluation by rheumatology and internal medicine specialists.

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