Japanese Journal of Gastroenterology and Hepatology

Case Report

ISSN: 2435-1210 | Volume 10

Chronic Sclerosing Sialoadenitis (Kuttner Tumor) In A Patient with Igg4 Disease: About A Case

Téllez-Cervantes JA* and Castellanos-Ledesma B

Department of Gastroenterology, Puebla Specialty Hospital, Mexican Social Security Institute, Mexico

***Corresponding author:**

Téllez Cervantes Jaime Alberto,
Department of Gastroenterology, Puebla Specialty Hospital, Mexican Social Security Institute, Puebla, Puebla, Mexico, Tel: 2222329029.

Received: 26 Feb 2024
Accepted: 06 Apr 2024
Published: 12 Apr 2024
J Short Name: JJGH

Citation:

Téllez Cervantes JA. Chronic Sclerosing Sialoadenitis (Kuttner Tumor) In A Patient with Igg4 Disease: About A Case. J Gstro Hepato. V10(9): 1-3

1. Case Report

The igG4-related disease is a chronic relapsing multiorgan fibroinflammatory syndrome of autoimmune etiology [1], characterized by a distinctive histopathological pattern of IgG4 in affected tissues, imaging findings, increased serum IgG4 levels, synchronous multiorgan distress, and response to glucocorticoids. The most frequently affected organs are the salivary glands, orbital anexa, pancreas, kidney, and retroperitoneum, and any organ can be compromised [2].

We present the case of a 43-year-old woman with a history of type 2 diabetes mellitus, hypothyroidism, and dacryocystitis in the right eye with a 1-year account. She begins suffering from abdominal pain, jaundice, weight loss of 8 kg in a month, and unquantified fever. Physical examination revealed generalized jaundice, abdomen with muscular resistance in the right hypochondrium, and optimistic Murphy. Biochemical tests showed hyperbilirubinemia of 15 mg/dl dependent on direct bilirubin, the elevation of aminotransferases, mild anemia without elevation of tumor markers, concluding biliary pathology. An abdominal tomography was performed, reporting a pancreas with a regular location, a rounded and isodense image in the head, and adjacent and respecting the splenic vein, measuring 40x44 mm with heterogeneous enhancement after contrast administration. Her cholangioresonance shows a filiform bile duct without tumors. Surgical management is decided, showing a gallbladder of 12x5x4 cm, thickened walls, no stones, indurated pancreas, enlarged, cholestatic liver with multiple nodular lesions, round stony ligament, IgG4 is requested due to suspected disease associated with said immunoglobulin with levels higher than 200 mg/dl with negative AMA, concluding disease associated with IgG4 with biliary-hepatic-lacrimal-pancreatic involvement. Later with intolerance to the ingestion of solids, they detect a tumor depending on the left submandibular gland. Given the risk of malignancy, resection of the said lesion was decided with a histopathological report of salivary gland with predominantly periductal mononuclear inflammatory infiltrate, intense stromal fibrosis, acinar atrophy, and formation of large reactive lymphoid follicles immersed in the fibrous stroma (Figure 1a and 1b), macroscopically specimen irregular 7x4x3cm with a multinodular appearance with nodules of 4 to 1 cm, multiple gray-whitish nodular lesions, solid and soft (Figure 2), consistent with chronic sclerosing sialadenitis. He is currently under treatment with prednisone 5 mg daily, which has shown clinical and biochemical improvement. Küttner's tumor (sclerosing sialoadenitis) is a chronic benign pseudotumoral alteration with inflammation almost exclusively of the submandibular gland with a mean age of 43 years [3]. A pathogenic autoimmune mechanism, including the spectrum of IgG4-related sclerosing diseases, is suggested. It may be associated with sclerosing cholangitis, retroperitoneal fibrosis or sclerosing pancreatitis, and pathologies related to IgG4 [4]. Histologically, Küttner's tumor shows diffuse infiltration of lymphocytes, plasma cells, and periductal fibrosis. Variable degrees of atrophy and destruction of the acini are replaced by fibrotic tissue. The ductal architecture is interrupted in some cases [5]. The histological changes vary according to the stage in which said the process is found and are grouped according to Seifer in:

• Stage 1 (focal sialadenitis), chronic focal inflammation with nests of lymphocytes surrounding the dilated salivary gland ducts and scant intraluminal secretion

• Stage 2 (diffuse lymphocytic sialadenitis with glandular fibrosis),

diffuse inflammatory infiltrate with periductal lymphoid follicles and focal squamous metaplasia with the proliferation of the ductal epithelium, centrilobular fibrosis with moderate acinar parenchymal atrophy

• Stage 3 (chronic sclerosing sialadenitis with glandular sclerosis), inflammatory infiltrate [6], prominent with the diffuse formation of lymphoid follicles, parenchymal acinar atrophy, periductal hyalinization, and sclerosis

• Stage 4 (sialoadenitis with chronic progressive sclerosis and cirrhosis of the salivary gland), the final stage is known as type cirrhosis [7].

Treatment goals include remission induction and relapse prevention, achieved with a combination of steroids (at high and low doses), eight immunomodulators (azathioprine, mycophenolate), and B-cell depleting therapy with antibodies against CD20.

High-dose steroid therapy (40 mg/day for four weeks, followed by tapering to 5 mg/week until patients are off medications) is an acceptable regimen for prednisolone. Response rates to therapy with steroids are 79-97%, with relapse rates to maintenance therapy being 31-60% [8]. The presence of chronic sclerosing sialadenitis is essential for diagnosing IgG4-associated disease since it is an important manifestation of said pathology, which the clinician must take into account when approaching it. Histopathology is the key to diagnosis. Tissue biopsy is the gold standard; serum IgG4 concentrations are useful for screening but are not reliable as a diagnostic marker. The challenge is to have clinical suspicion. Other drugs used in the treatment of IgG4-RD are azathioprine, methotrexate, tacrolimus, 6 -mercaptopurine, cyclophosphamide, and rituximab, and other clinical entities present with increased serum IgG4 such as asthma, atopic dermatitis, pemphigus or Castleman's disease [9], etc. Therefore, in the face of such a finding, we must consider a differential diagnosis with them. Although this disease has been compared to a crow flying in the dark of night [10], they have once diagnosed, the prognosis of life of patients changes since it is good in most of them.

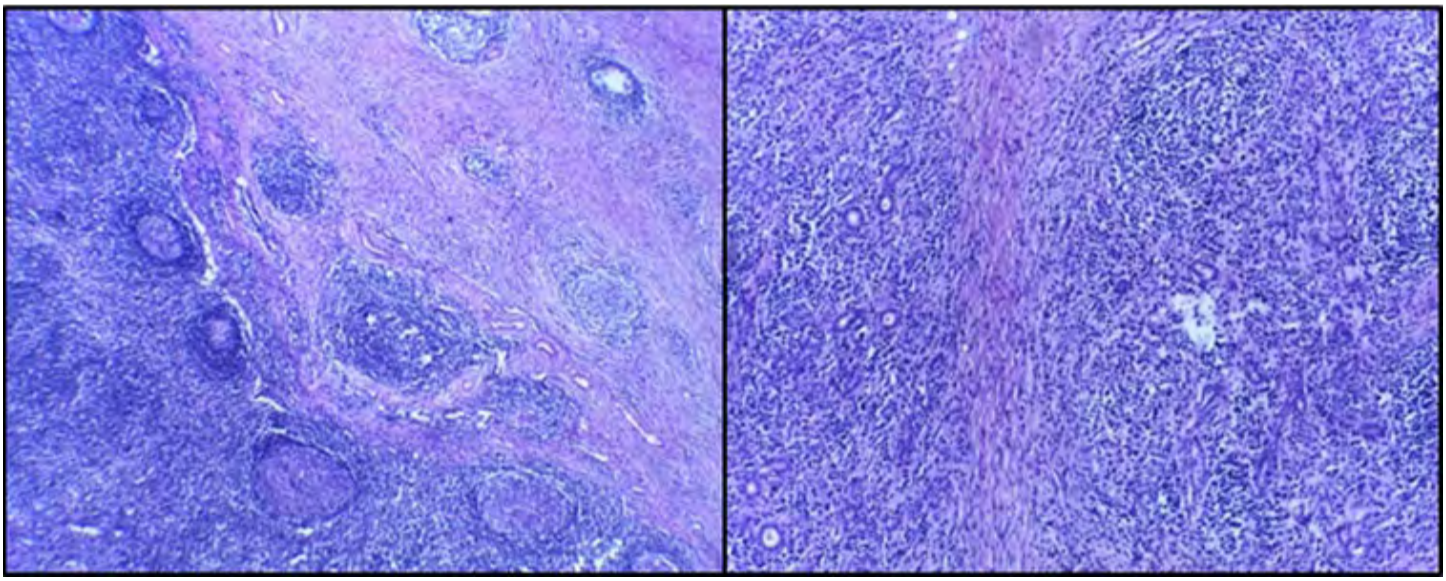

Figura 1a y 1b. 1a) Aumento 4x de glándula submandibular. Configuración lobulillar con prominente infiltrado inflamatorio crónico así como folículos linfoides asociados con fibroesclerosis y atrofia severa. 1b) Aumento 10x. Infiltrado inflamatorio prominentemente periductal, esclerosis y atrofia intense.

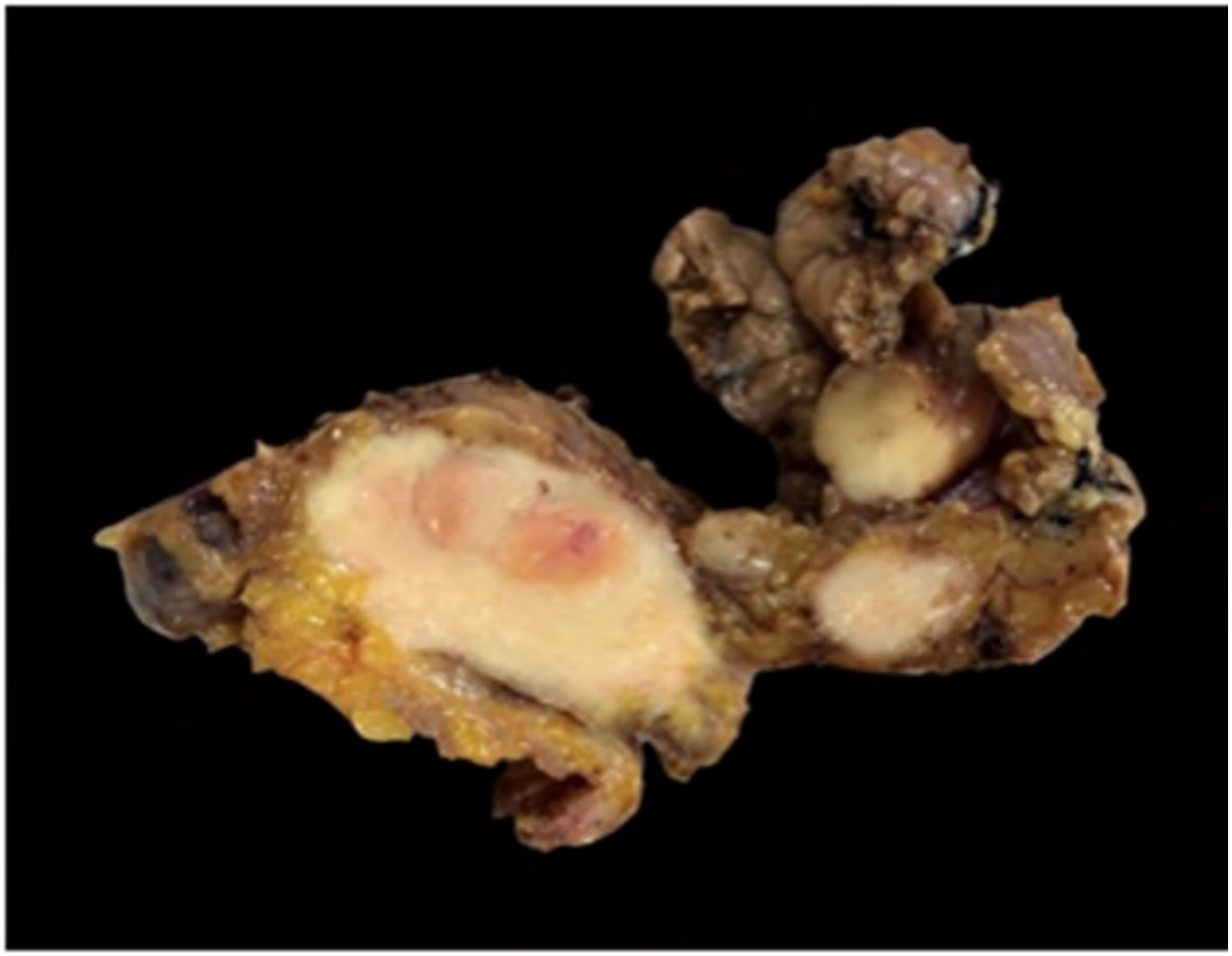

Figura 2. Una lesión multinodular de superficie heterogénea al corte con agrandamiento difuso, correspondiente a tumor de Kuttner.

Reference

1. Romero FJ, Zetina VV and Marquez RML. Chronic sclerosing sialadenitis (Küttner tumor) in submandibular glands related to IgG4 disease. Rev Mex Cir Bucal Maxillofac. 2012; 8(3): 92-97.
2. Cardenas MI and Ortiz MI. Immunoglobulin G4-related disease: Imaging findings. Ann Radiol Mex. 2019; 18: 36-45.
3. Mesquita MA, Viana AL, de Freitas JC. Küttner's tumor with parotid gland involvement. Arch Head Neck Surg 2017; 49:1-4.
4. Martínez N, Baquera J, Sánchez R. Küttner's Tumor (Chronic Sclerosing Sialadenitis). Clinical, Pathological, and Immunohistochemical Study in 8 Cases of a Little-Known Entity. Act Otorrinolaringol Esp. 2007; 58(1): 25-30
5. Miyabe K, Zen Y, Cornell LD. Gastrointestinal and Extra-Intestinal Manifestations Of Igg4–Related Disease. Gastroenterol. 2018; 155: 990-1003.
6. Agazzoni M, Barrenecha M, Bolaños S et al. Igg4-Related Disease: A Diagnostic Challenge; hematol. 2017; 21 1: 71-76.
7. Backhus J, Neumann C, Perkhofer L. A Follow-Up Study Of A European Igg4-Related Disease Cohort Treated With Rituximab. J. Clin. Med. 2021; 10: 1329.
8. Deshpande V. Igg4-Related Disease Of The Gastrointestinal Tract. Arch Pathol Lab Med. 2015; 139:742-749.
9. Carrillo R and Echeverría JA. Igg4-related disease. Med Int Mex. 2013; 29 (1): 53-61.
10. Kamisawa T, Zen Y, Pillai S. IgG4-related disease. Lancet. 2015; 385: 1460–71.