

A Case Report of Sjogren's Syndrome

Dr. Abinaya. R¹, Dr. Sai Shankar²

Aarupadai Veedu Medical College and Hospital, Pondicherry, India

Abstract: Sjogren's syndrome is an autoimmune disorder characterized by dry eyes (kerato conjunctivitis sicca), dry mouth (xerostomia) and glandular destruction (parotid and lacrimal glands). Sjogren's syndrome may be primary or secondary. On examination, parotid gland appears soft and swollen. Radiological imaging shows enlarged gland with multiple internal cystic areas. Histological examination of gland shows lymphocytic infiltration.

Keywords: Sjogren's syndrome, Sialography, Parotid enlargement

1. Summary

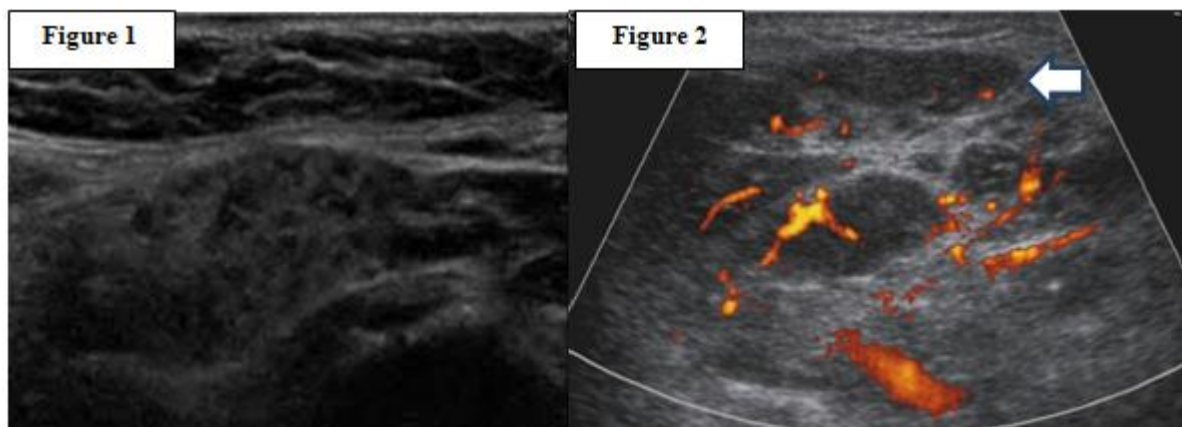
A 25 year old female known case of rheumatoid arthritis came with complaints of unilateral parotid swelling associated with low grade fever and dryness of mouth. On radiological imaging, bilateral parotid glands appear enlarged and heterogenous with internal cystic areas. On histopathological examination, multiple regions of focal lymphocytic, plasma cells and macrophages infiltration was appreciated confirming our diagnosis of secondary sjogren's syndrome. Hereby we describe our approach to the case of secondary sjogren's syndrome using various radiological investigations.

2. Case Presentation

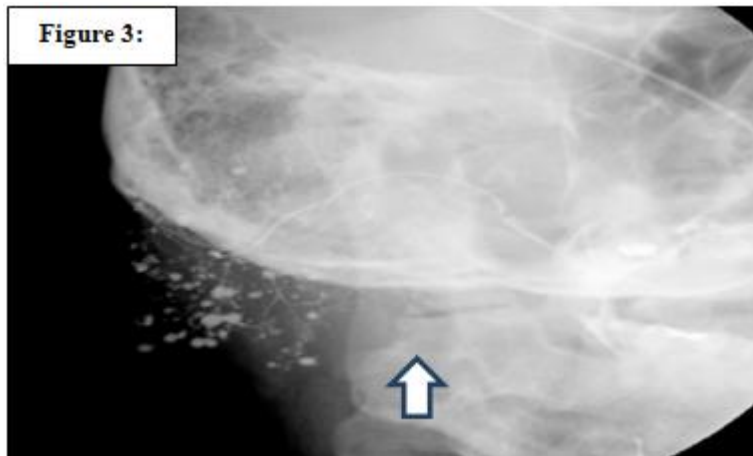
A 25 year old female came with complaints of unilateral parotid swelling for 5 days duration associated with low grade fever and mild joint pain. Patient also complained of dryness of mouth that increased in intensity early morning. Past history of recurrent episodes of parotid swelling was present and the patient was a known case of Rheumatoid arthritis on regular treatment (DMARDS). On local examination, bilateral parotid swelling was appreciated.

3. Investigations

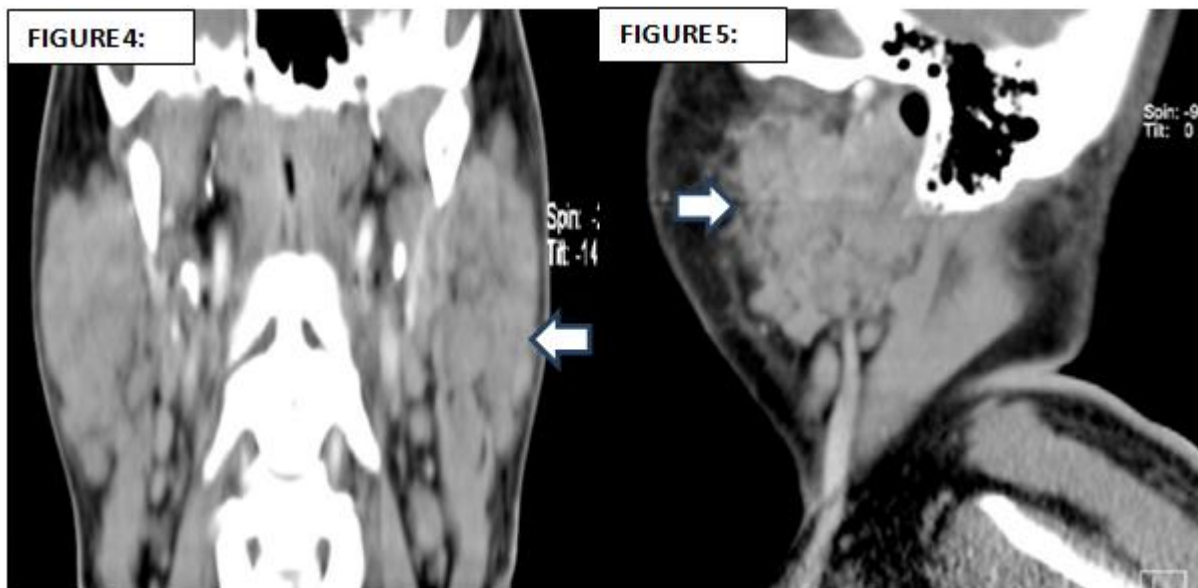
Routine blood investigations showed elevated ESR levels, mild normocytic normochromic anemia and mild leukopenia. Rheumatoid factor was positive. The patient then underwent ultrasound of the salivary glands which revealed enlarged bilateral parotid glands with multiple well defined hypoechoic areas seen within the gland. On colour Doppler, both parotid glands show increased vascularity on colour Doppler. The patient was then subjected for conventional sialography which showed punctate ectasis of the parotid gland ducts as well as the entire extent of the ducts involved. Then we took the patient for CT neck which revealed enlarged bilateral parotid glands with multiple internal hypodense areas and cervical lymphadenopathy. MRI Neck of the same patient showed enlarged parotid gland with internal T2 hyperintense areas likely to be cystic areas within the gland. The patient was then subjected for ultrasound guided biopsy which revealed multiple regions of focal lymphocytic, plasma cells and macrophages infiltration.



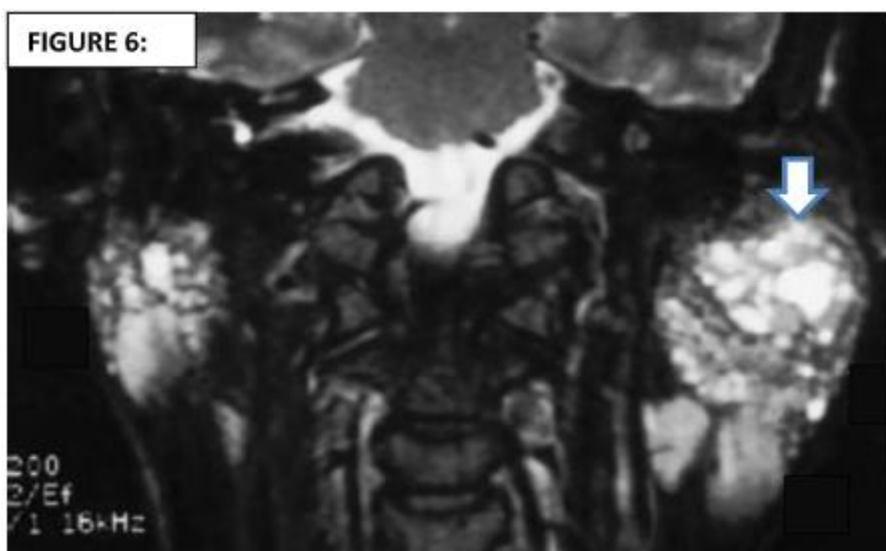
Gray-scale and Doppler USG images 1 and 2: show enlarged parotid gland with multiple small internal hypoechoic areas (white arrow). The gland shows increased vascularity on colour Doppler.



Conventional sialography of parotid gland Figure 3: shows punctate ectasis of the ducts (white arrow) and also the extent of the ductal system involved.



CT Coronal and Sagittal images of parotid gland 4 and 5: Enlarged bilateral parotid gland (white arrows)with internal low-density areas and cervical lymphadenopathy.



MRI T2WI CORONAL SECTION FIGURE 6: shows enlarged bilateral parotid glands with internal T2 hyperintense cystic areas (white arrow).

Treatment:

Supportive treatment and counselling was given for the patient. The patient was advised for regular follow up and investigations.

4. Discussion

Sjogren's syndrome is one of the common autoimmune diseases that affects multiple organs. Sjogren's syndrome is mainly characterized by dry eyes (Kerato conjunctivitis sicca), dry mouth (Xerostomia) and enlarged parotid and lacrimal glands. As many as 1 to 2 million persons in the United states are affected ; the reported prevalence is between 0.05 and 4.8 percent of the population⁽¹⁾. The disease affects predominantly women older than 40 years. Sjogren's syndrome may be primary or secondary with primary syndrome having no antecedent cause and secondary syndrome occurring along with rheumatoid arthritis / scleroderma / systemic lupus erythematosus ⁽²⁾. Patients with sjogren's syndrome have more chance of developing lymphoma and hence continuous follow up is mandatory to rule out lymphoma.

Clinical Features

Eye symptoms include dryness of eyes, irritation, foreign body sensation and recurrent infections in eye. Due to dryness of eyes and recurrent irritation, the patient may develop corneal and conjunctival ulcers. Oral symptoms include dryness of mouth, halitosis, recurrent oral ulcers and dental caries. Reduced secretions in the respiratory tract leads to recurrent bouts of dry cough and recurrent respiratory tract infections. Reduced secretions of skin leads to dry skin and ulcers. Bilateral parotid glands may be soft and swollen.

Pathology

The pathogenesis of sjogren's syndrome still remains a mystery. Serologic studies show an association between primary sjogren's syndrome and HLA-DR haplotypes ⁽³⁾. Some say that the etiology is environment oriented which stimulates an autoimmune reaction in genetically susceptible individuals⁽¹⁾. Various etiology lead to B lymphocyte hyperstimulation which leads to accumulation in the target organs namely eyes, mouth, salivary glands and exocrine glands. The histology of the affected glands are characterized by lymphocytic and plasma cell infiltration that lead to gland degeneration and necrosis of the gland. Involvement of parotid gland is characterized by non obstructive sialectasia. This duct ectasia may be due to dilated hypertrophic ducts and acini surrounded and then occluded and destroyed by lymphocytic infiltration, or due to the extravasation caused by weakened duct walls remains controversial till now⁽⁴⁾.

5. Imaging Findings

The imaging modalities used to diagnose sjogren's syndrome include ultrasonography, conventional sialography, CT sialography, MR Sialography and

scintigraphy. On ultrasound, the gland may be normal or enlarged in size with presence of internal hypoechoic areas that represent glandular destruction.

On conventional sialography, dilatation of ducts can be seen. The most common finding is the dilatation of the most peripherally placed duct which is present in 55% of positive cases⁽⁵⁾. The other signs include punctate ectasis (seen in 28% of cases ⁽⁵⁾) peripheral sialectasis, punctate, globular, cavitary, cylindrical and fusiform⁽⁵⁾.

CT scan is a poor imaging modality for the diagnosis of sjogren's syndrome and can be used to rule out presence of stones within the ducts.

In MR imaging, T1 and T2 weighted images were taken into consideration. Normal parotid gland appears homogenous and are of intermediate signal intensity in T1WI and T2WI⁽⁶⁾. In patients with sjogren's syndrome, the glands appears heterogenous with high and low intensity areas on T1WI in which the high intensity areas indicate fat lobules; the glands show hyperintense cystic areas on T2WI⁽⁶⁾.

According to the criteria proposed by Tonami et al, the MR sialographic stages of SS were determined as follows ⁽⁷⁾in which stage 0 = normal. Stage 1 = punctate (diffuse, spherical areas of high-signal intensity, 1 mm or less in diameter and uniform in size, are distributed evenly throughout the gland). Stage 2 = globular (the spherical areas of high signal intensity increase to 1 to 2 mm in diameter). Stage 3 = cavity (with further disease progression, the areas of high intensity coalesce and enlarge further, more than 2 mm in diameter). Stage 4 = destructive (marked dilation of the main duct with an irregular diameter, as well as irregular branching). MR Sialography gives more information about the ducts but MR imaging gives even more information about the glandular parenchymal changes⁽⁸⁾.

^{99m}Tc-pertechnetate salivary gland scintigraphy can also be done which shows reduced uptake of radiotracer by the salivary glands in patients with sjogren's syndrome ⁽⁹⁾. However this finding can be present in patients with obstructive disorders. Hence abnormal finding in salivary scintigraphy should not be considered pathognomonic of sjogren's syndrome and other clinical criteria should be taken into consideration

6. Treatment

There is no definite cure for sjogren's syndrome and hence only symptomatic treatment prevails till now. For dry eyes, artificial tears or methyl cellulose eye drops can be used. A proper oral hygiene has to be maintained and the use of antibacterial mouth wash has to be initiated. Rituximab which is an anti – CD20 monoclonal antibody may be used for severe inflammatory manifestations of sjogren's syndrome as it reduced B lymphocyte levels ⁽¹⁰⁾.

References

- [1] Paul kruszka, Robert J. O'brian. Diagnosis and management of sjögren syndrome. *Americal family physician*. 2009 (mar)15;79(6):465-470.
- [2] Alex S.L. Toh, Stephen J. Broomfi eld, Lee-Suan Teh, et al. Bilateral multicystic parotid masses in primary Sjögren syndrome. *ENT Journal*. 2011 (oct), 90:10
- [3] Gottenberg JE, Busson M, Loiseau P, et al. In primary Sjögren's syndrome, HLA class II is associated exclusively with autoantibody production and spreading of the autoimmune response. *Arthritis Rheum*. 2003 Aug;48(8):2240-5.
- [4] R. Becerra ulloa, D. Pachuca gonzalez, I. F. Alva lopez, et al. Imaging of Sjogren syndrome in salivary glands. *ECR 2012 / C-2568*
- [5] Leopold Gonzalez, Allen H. Mackenzie, Riaz A. Tarar. Parotid Sialography in Sjögren's Syndrome. *Radiographics*. 1970 oct,97(1).
- [6] Yukinori Takagi, Misa Sumi, Tadateru Sumi, Yoko Ichikawa and Takashi Nakamura. MR Microscopy of the Parotid Glands in Patients with Sjögren's Syndrome: Quantitative MR Diagnostic Criteria. *American Journal of Neuroradiology*. May 2005, 26 (5) 1207-1214.
- [7] Yan-De Ren, Xiang-Rong Li, Jing Zhang, et al. Conventional MRI techniques combined with MR sialography on T2-3D-drive in Sjögren syndrome. *international journal of clinical and experimental medicine*. 2015 Mach; 8(3):3974-3982.
- [8] Danchaivijitr N, Tamburini S, Hesselink JR. MRI of the parotid glands in a patient with Sjogren's syndrome. *Eurorad*. 2006; case 1856.
- [9] Wing Hang Luk, Jessie Tse Hang Yeung, Eliza Po Yan Fung, Chiu Ming Lok, and Yuet Ming Ng. Salivary Gland Scintigraphy in Patients with Sjogren's Syndrome: A local Experience with Dual-tracer. *AOJNMB*. 2017; 5(1):56-65.
- [10] E. William St Clair, Marc C. Levesque, Eline T. Luning Prak, et al. Rituximab Therapy for Primary Sjögren's Syndrome: An Open-Label Clinical Trial and Mechanistic Analysis. *Arthritis Rheum*. 2013 Apr; 65(4): 1097–1106.
- [11] Stuart S. Kassan, Haralampos M. Moutsopoulos. Clinical Manifestations and Early Diagnosis of Sjögren Syndrome. *JAMA Internal Medicine*. 2004;164(12):1275-1284.
- [12] Chiara Baldini, Nicoletta Luciano, Gaia Tarantini, et al. Salivary gland ultrasonography: a highly specific tool for the early diagnosis of primary Sjögren's syndrome. *Arthritis research and therapy*. 2015; 17(1): 146.