

Sjögren Syndrome with Bilateral Multicystic Enlargement of the Parotids and Submandibular Glands

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DOI: [10.36347/sjmcr.2022.v10i02.021](https://doi.org/10.36347/sjmcr.2022.v10i02.021)

| Received: 09.01.2022 | Accepted: 14.02.2022 | Published: 19.02.2022

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Abstract

Case Report

Sjögren's syndrome is an autoimmune disease. This means that the immune system attacks parts of your own body by mistake. It attacks the glands that produce tears and saliva. We report the case of a 51-year-old woman who presented with bilateral parotid gland enlargement as the initial manifestation of primary Sjögren syndrome.

Keywords: Sjögren's syndrome, immune system, submandibular glands.

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INTRODUCTION

Sjögren's syndrome (SS) is an autoimmune systemic disease characterized by ocular and oral dryness due to affection of exocrine glands, but may also present extraglandular manifestations that are more active and severe and conditions the prognosis of long-term disease.

The first case of Sjögren's syndrome was reported in 1888 by Johann Mickulicz [1], linked at that time with obvious diseases, such as sarcoidosis and lymphoma, after that and exactly In 1933, the Danish ophthalmologist Henrick Sjögren summarized 19 female patients with Keratoconjunctivitis sicca, two of which had swelling of major salivary glands [2].

The disease predominantly affects postmenopausal women, with an incidence rate of 5 cases/100,000 inhabitants. The prevalence is around 0.6-3.3% depending on the criteria used to retain the diagnosis [3].

CASE REPORT

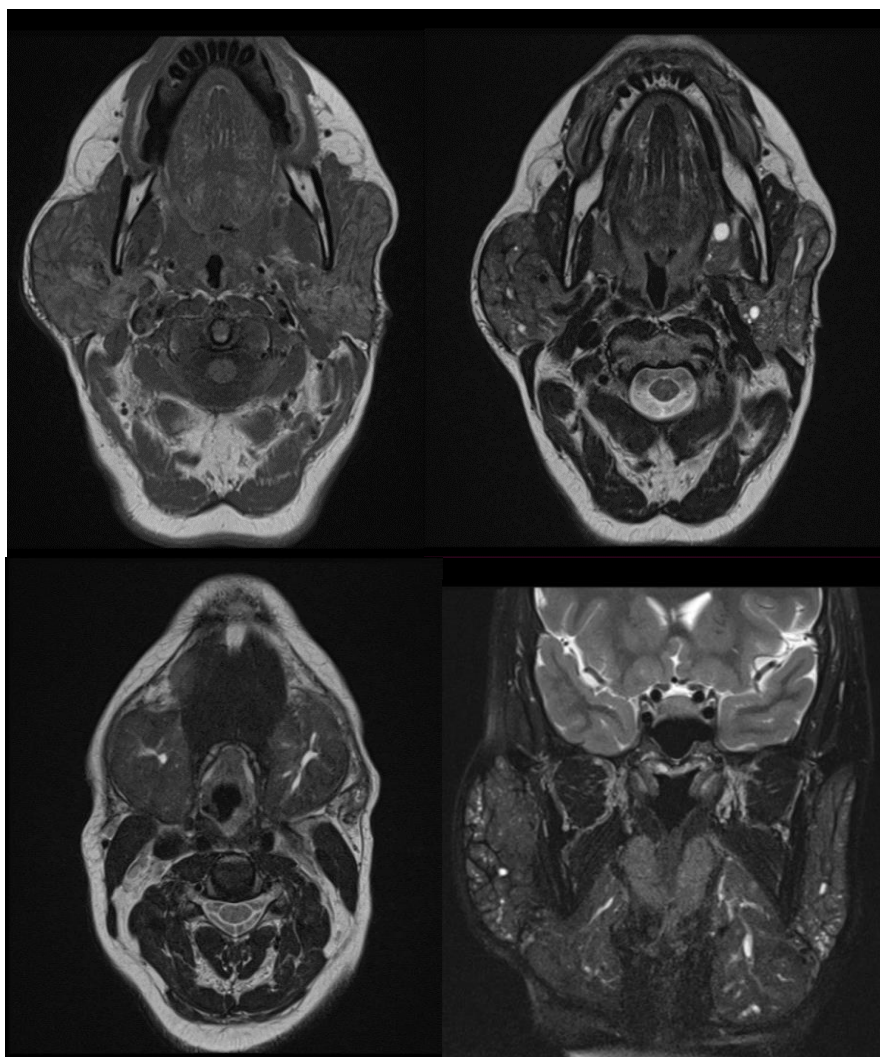
A 48-12 months-old lady was addressed to the Oto-rhino-laryngology Department for assessment of a 6-week history of bilateral parotid gland swelling.

There was no family history. The physical exam confirmed both parotid glands' bilateral, diffuse expansion with ocular dryness and rheumatism pain. The blood laboratory test was normal, however, the erythrocyte sedimentation was 48 mm/hr.

MRI was performed showing a bilateral and diffuse enlargement of both lobes superficial and deep of the parotid glands, and also the submandibular glands, which contain multiples cystic foci hyposignal T1 and hyper signal T2 there was a combination of punctate regions of calcification and fatty replacement realising the salt and pepper appearance figures. The Immunoglobulins found in the patient's blood IgG level of 6.7 g/L, IgM: 2.1 g/L, and IgA: 5g/L, and the serology object the positivity of anti-La/SSB but the absence of anti-Ro/SSA antibodies The confirmatory diagnosis of Sjögren's syndrome was made after histopathological evaluation of the biopsy specimens of the submandibular glands which shows lymphoepithelial infiltration by a mixed population of B and T lymphocytes without evidence of malignancy

After a discussion of the surgery and its risks, the medical staff jointly decided to adopt a A wait-and-see policy by a repeated MRI with possible second biopsy if there is any change of the aspect of the glands.

Citation: Essaber Hatim, Daoud Ali Mohammed, Firdaous Touarsa, Amina El Khamlichi, Ech-cherif El Kettani Najoua, Fikri Meriem, Jiddane Mohammed. Sjögren Syndrome with Bilateral Multicystic Enlargement of the Parotids and Submandibular Glands. Sch J Med Case Rep, 2022 Feb 10(2): 131-133.



MRI images on axial T1 and T2 and coronal T2 fat-suppressed: demonstrating both parotid and submandibular glands which are enlarged with multiples cystic lesions in hyposignal T1 hypesignal T2 as expected in Sjorgen syndrome

DISCUSSION

The etiopathogenesis of Sjögren's syndrome is multifactorial. Nowadays, We retain the theory of the lymphocytic infiltration of the glandular tissue with lymphoepithelial lesions.

Most (as many as 66%) patients with primary Sjögren syndrome experience major salivary gland enlargement [4].

The enlargement may be constant or it can be intermittent with variable periods of remission. Clinically, there might also be unilateral or bilateral; the parotid gland is most of the time affected by benign lymphoepithelial cysts as a common manifestation.

Other less common cystic benign lesions can occur in the parotid gland like mucocele, branchial cyst or cystic Warthin tumor, or malignant including mucoepidermoid cysts, adenoid cystic carcinomas, and undifferentiated carcinomas.

MRI has become an increasingly more beneficial tool in detecting inflammatory modifications and in diagnosing tumours of the parotid gland. In our patient, MRI objective numerous foci of low signal intensity on T1 WI and hyper signal on T2 WI; and also punctate calcification on hyposignal T1 and T2 WI the "salt and pepper look," and it's far suggestive of Sjögren syndrome.

Histologic confirmation is an important factor in the process of diagnosing Sjögren syndrome. The fine needles aspiration cytoprotection "FNAC" of the involved gland can demonstrate the presence of numerous lymphocytes combined with follicular centre cells, plasma cells, and histiocytes, which imply a benign lymphoepithelial infiltration consistent with Sjögren syndrome.

Regular observe-up of patients with Sjögren syndrome is important for early detection of signs and symptoms suggestive of lymphoma. During each follow-up, the clinicians should examine the neck to

search for signs of lymphadenopathy and of the parotid gland to look for asymmetry and changes in size. Additionally, they have to look for associated B symptoms such as unexplained prolonged fever, night time sweats, constant fatigue, and unexplained weight loss. If a malignancy is suspected, the affected person ought to undergo a repeat MRI followed by FNAC.

Differential diagnoses include:

- **Chronic Infectious or Obstructive Parotitis:** with Irregular dilatation and stenosis of ducts containing sialoliths with oblong sharp without Lacks solid masses Multiple calculi
- **Benign Lymphoepithelial Lesions of HIV:** there is Mixed cystic and solid lesions enlarging both parotids, it is associated with tonsillar hyperplasia and cervical reactive adenopathy usually we note the lack of glandular calcifications
- **Warthin Tumor;** only 20% are multiple; may be unilateral or bilateral with mural nodules if cystic
- **Parotid NHL Nodes:** Solid masses in parotid usually without cystic change and cervical adenopathy
- **Parotid Metastatic Disease:** Primary malignancy like skin cancers of the scalp, face, external ear
- **There is a** Unilateral or bilateral, single or multiple parotid masses with invasive margins
- **Parotid Sarcoidosis:** is a rare manifestation of sarcoidosis associated with cervical & mediastinal lymph nodes

The treatment of Sjögren syndrome is symptomatic with acid sugar-free food to increase

salivary secretion, and oral hygiene tears artificial and ointments, and by avoiding infection among others or if systemic disease.

CONCLUSION

There are a lot of etiologies of multicystic parotid gland enlargement, but some clinical features like xerostomia and dry eyes can suggest Sjögren's syndrome. The recommended diagnostic investigations are immunology, radiology especially MRI, and histology are important in the process of diagnosis and exclude other severe similar pathologies such as HIV infection and lymphoma. A lengthy-term and careful follow-up are important, especially in younger patients, to detect early any degeneration.

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