

Role of Ultrasound in Diagnosis of Sjogren's syndrome

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Abstract

Background: Primary Sjögren's syndrome is a multisystem autoimmune disease characterized by hypofunction of salivary and lacrimal glands and possible multi-organ system manifestations. Over the past 15 years, three sets of diagnostic criteria have been proposed, but none has included salivary gland ultrasonography. However, recent studies support its role in the diagnosis and prognostic evaluation of patients with Sjögren's syndrome. **Aim:** The aim of the present study was to determine the role of ultrasound in diagnosis of Sjögren's syndrome and to assign scoring based on texture morphology of parotid gland seen on ultrasound in patients. **Subjects and Methods:** In this study, 30 patients of age between 20-60 years with Sjögren's syndrome referred for ultrasonography of neck to the department of radiodiagnosis, PDU Medical college and Civil hospital, Rajkot, Gujarat, over a period of 1.5 years from 01/05/2018 to 01/11/2019 are included. **Results:** SGUS was performed by the same investigator, an experienced radiologist, using a real-time scanner (LogiqP9; GE Healthcare) with a 12-MHz linear array transducer. Both the parotid and submandibular glands were scanned and echostructure of each gland was graded on a scale of 1 to 4. **Conclusion:** Our study contributes to the conclusion that ultrasonography can be used as a primary modality of imaging to early diagnose Sjögren syndrome in a non-invasive way.

Keywords: Sjögren's syndrome, Parotid gland, Ultrasound scoring system.

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Introduction

Primary Sjögren's syndrome (pSS) is a systemic autoimmune disease clinically characterized by oral and ocular dryness, reflecting lymphocytic infiltration and subsequent exocrine gland dysfunction. However, during disease progression, any organ or mucosal surface may be involved. Thus, pSS presents as a heterogeneous non-organ-specific autoimmune entity, encompassing a wide spectrum of clinical manifestations, serological abnormalities, and scattered complications.^[1] The incidence rate of pSS is estimated at about 6.92 per 100,000 person-years, with a female-to-male ratio in incidence data of 9:1.^[2] Prevalence rates vary widely between studies, but they are estimated at about 43.03 cases per 100,000 inhabitants when considering only population-based studies.^[2] The peak incidence of pSS is in women aged 55–65 years.^[2]

Because pSS is a systemic autoimmune disorder, several autoantibodies are detected in pSS patients,^[3] of which antinuclear antibodies (ANA) are the most frequently detected (in up to 80% of patients); however, the most relevant autoantibodies are directed against Ro/SSA or La/SSB antigens.^[3] SGUS is simple, non-invasive, widely available, non-irradiating and less expensive than other imaging techniques.^[4] Its use allows us to classify the echogenicity, homogeneity, degeneration, fibrosis and

calcification of the glandular parenchyma.

New indices have been developed to objectively assess systemic and symptomatic manifestations in patients with pSS. The European League Against Rheumatism (EULAR) Sjögren's syndrome disease activity index (ESSDAI) was developed in 2009 by consensus of a large group of worldwide experts from European and North American countries.^[5] The ESSDAI includes 12 organ-specific domains (cutaneous, pulmonary, renal, articular, muscular, peripheral nervous system, central nervous system, haematological, glandular, constitutional, biological, and lymphadenopathy), and scores < 5 indicate low activity, scores ≥ 5 and ≤ 13 indicate moderate activity, and scores ≥ 14 indicate high activity.^[6]

Aims and Objectives

The objective of the present study was to determine the role of ultrasound in diagnosis of Sjögren's syndrome and to assign scoring based on texture morphology of parotid gland seen on ultrasound in patients.

Subjects and Methods

Methodology

In this study, 30 patients of age between 20-60 years with Sjögren's syndrome referred for ultrasonography of neck to the department of radiodiagnosis, PDU Medical college and

Civil hospital, Rajkot, Gujarat, over a period of 1.5 years from 01/05/2018 to 01/11/2019 are included. All scans were carried out on high frequency linear transducer (12 MHz) on GE LOGIQ P9 USG Machine.

Type of study: Single center

Consent for participation in study: Yes.

Machine Used: GE LOGIQ P9 USG MACHINE

Patient selection:

Inclusion Criteria:

- The 2002 AECG Criteria were used to select patients.
- Patients of both sexes.
- Patients in the age group of 20-60 years.

Exclusion Criteria:

- Patient's negative consent
- Hepatitis B or C
- Sarcoidosis
- Other connective tissue disease.

Ultra-sonographic Examination

SGUS was performed by the same investigator, an experienced radiologist, using a real-time scanner (LogiqP9; GE Healthcare) with a 12-MHz linear array transducer. Both the parotid and submandibular glands were scanned and echostructure of each gland was graded on a scale of 1 to 4. According to this scale, grade 1 = small hypoechoogenic areas without echogenic bands, grade 2 = multiple hypoechoogenic areas measuring < 2 mm with echogenic bands, grade 3 = multiple hypoechoogenic areas measuring 2–6 mm with hyperechogenic bands, and grade 4 = multiple hypoechoogenic areas measuring > 6 mm or multiple calcifications with echogenic bands. Each patient received one score per gland and the highest score obtained was considered in the analysis.

Results

Table 1: Scoring of Sjögren Syndrome according to different variables

Variables	Score 1-2	Score 3-4
ANA :		
Positive	6	16
Negative	5	3
ANTI Ro/SSA :		
Positive	7	12
Negative	8	3
ANTI La/SSB :		
Positive	6	4
Negative	7	13
RF		
Positive	3	17
Negative	3	7

Table 2: Characteristics of the study population

Characteristics	Females	Males
Age(years), mean	47	53
Symptom duration(years), mean	2	2.5
Sex, number	22	8
Xerophthalmia, number	20	6
Xerostomia, number	21	7
Abnormal ocular tests, number	20	6

The results of the study as represented in [Table 1 & 2] are summarised in the following paragraph.

[Table 1] shows 6 ANA positive patients had score 1-2 and 16 had score 3-4; 7 ANTI Ro/SSA positive had score 1-2 and 12 had 3-4; 6 Anti La/SSB positive had score 1-2 and 4 had score 3-4; 3 RF positive had score 1-2 and 17 had score 3-4.

[Table 2] shows mean age of Sjögren's syndrome in females is 47 and that in males is 53. Also the mean duration of symptom in females is 2 years and that in males is 2.5 years. Also most of the patients in the study cohort are females. 20 female and 6 male patients had xerophthalmia; and 21 female and 7 male had xerostomia. 20 females and 6 males had abnormal ocular test.

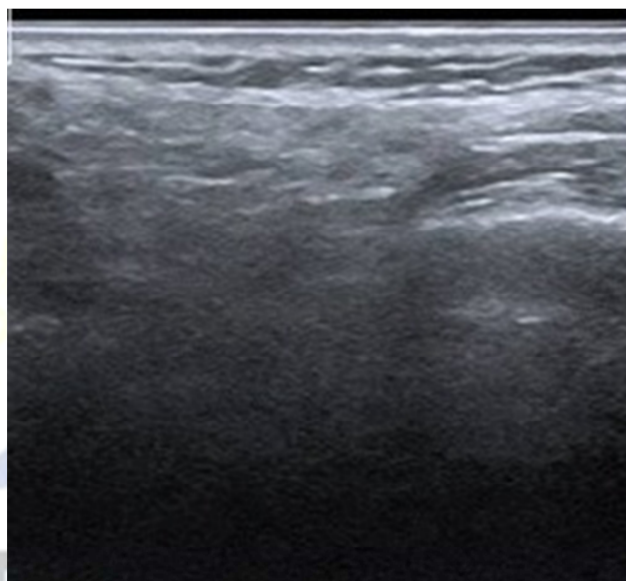


Figure 1: Parotid gland with mild unspecific inhomogeneity (grade 1).

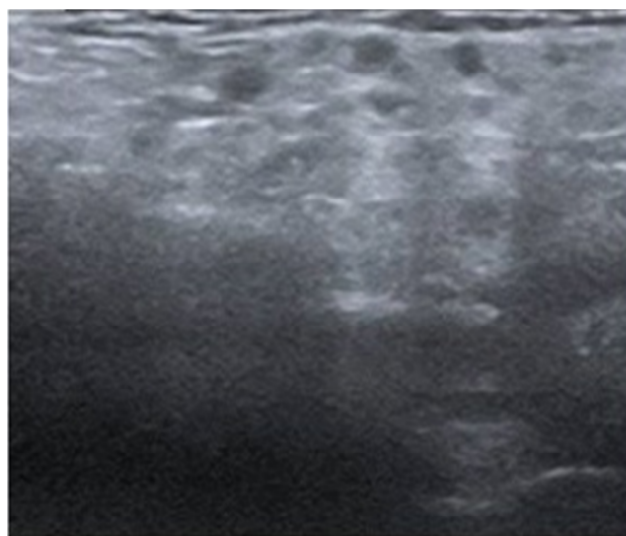


Figure 2: Parotid gland with evident inhomogeneity (grade 2)

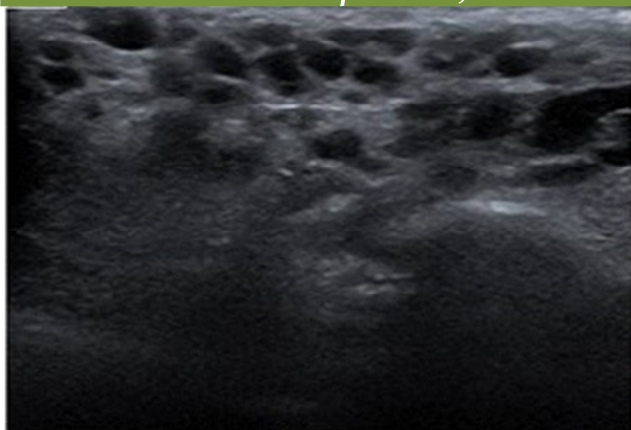


Figure 3: Parotid gland with gross inhomogeneity (grade 3)

Discussion

Abnormalities in sonographic findings in patients with Sjögren's syndrome were characterized by Bradus et al. as bilateral cystic changes. We found multiple hypochoic areas in the affected glands, consistent with the cystic changes that Bradus et al.^[7] proposed. The hypochoic areas were larger in the patients with definite than with probable Sjögren's syndrome. Furthermore, in advanced stages, the hypochoic areas were irregularly demarcated. At present, we cannot confidently indicate the pathologic features relevant to these sonographic changes.

The multiple hypochoic areas represent enlarged parotid lobules replaced by lymphocytic infiltration. The multiple echogenic bands, which appeared in the higher stages of our grading (grades 3-4), may represent fibrous connective tissues proliferating in and replacing the gland lobules. The noninvasive diagnostic method might be beneficial to patients with Sjögren's syndrome, who are expected to endure a long progression of the disease.

This study aimed to assess the diagnostic accuracy of SGUS for the early detection of Primary Sjögren's Syndrome (pSS).

Our study specifically limited enrollment to patients at the onset of the disease. Hence, rather than addressing the duration of the disease, we focused on the duration of the symptoms reported by the patients.

Intriguingly, by using a scoring system mainly focused on salivary gland inhomogeneity, we were able to demonstrate that changes in the salivary gland parenchymal echostructure appeared relatively early in the course of the disease. In line with these data and despite the different scoring systems adopted, all the available studies have highlighted parenchymal gland inhomogeneity as the most important distinctive feature for differentiating pSS from other salivary gland disorders.^[8] The SGUS score for inhomogeneity was also the parameter in which interobserver agreement between the radiologists was the highest, reinforcing the reliability and the feasibility of SGUS in daily clinical practice. Therefore, SGUS appeared to mirror dysfunction of the salivary glands and inflammation even in the early stage of the disease. In

patients who have declined biopsy but have a positive SGUS score this may be helpful in routine care, additionally supporting the value of SGUS for non-invasive diagnosis of the disease.

Conclusion

We evaluated 30 patients with Sjögren's Syndrome using ultrasonography.

Our study contributes to the conclusion that ultrasonography can be used as a primary modality of imaging to early diagnose Sjögren syndrome in a non-invasive way.

Our results may further evidence the usefulness and the opportunity of including SGUS among the tests for the diagnosis of the disease.

The use of ultrasound is easily available, low cost and widespread that can easily diagnose Sjögren's Syndrome.

With the use of ultrasound we have classified the severity of Sjögren's syndrome from score 1 to 4.

Abbreviations:

1. pSS: Primary Sjögren's syndrome
2. SGUS: Salivary gland ultrasonography
3. ANA: Antinuclear antibodies
4. RF: Rheumatoid factor
5. ESSDAI: European League Against Rheumatism (EULAR) Sjögren's syndrome disease activity index
6. AECG: American-European Consensus Group
7. MHz: Megahertz

References

1. Luciano N, Valentini V, Calabro A, Elefante E, Vitale A, Baldini C, et al. One year in review 2015: Sjogren's syndrome. *ClinExpRheumatol.* 2015; 33: 259–271. pmid:25896475
2. Qin B, Wang J, Yang Z, Yang M, Ma N, Huang F, et al. Epidemiology of primary Sjogren's syndrome: a systematic review and meta-analysis. *Ann Rheum Dis.* 2015; 74: 1983–1989. pmid:24938285
3. Bournia VK, Vlachoyiannopoulos PG. Subgroups of Sjogren syndrome patients according to serological profiles. *J Autoimmun.* 2012; 39: 15–26. pmid:22575069
4. Cornec D, Jousse-Joulin S, Pers JO, Marhadour T, Cochener B, Boisrame-Gastrin S, et al. Contribution of salivary gland ultrasonography to the diagnosis of Sjogren's syndrome: toward new diagnostic criteria? *Arthritis Rheum.* 2013; 65: 216–225. pmid:23108632
5. Seror R, Ravaud P, Bowman SJ, Baron G, Tzioufas A, Theander E, et al. EULAR Sjogren's syndrome disease activity index: development of a consensus systemic disease activity index for primary Sjogren's syndrome. *Ann Rheum Dis.* 2010; 69: 1103–1109. pmid:19561361
6. Seror R, Bootsma H, Saraux A, Bowman SJ, Theander E, Brun JG, et al. Defining disease activity states and clinically meaningful improvement in primary Sjögren's syndrome with EULAR primary Sjögren's syndrome disease activity (ESSDAI) and patient-reported indexes (ESSPRI). *Ann Rheum Dis.* 2016; 75: 382–389. pmid:25480887
7. Bradus RJ, Hybarger P, Gooding GAW. Parotid gland: US findings in SjOgren syndrome. *Radiology* 1988; 169:749-751
8. Tzioufas AG, Moutsopoulos HM. Ultrasonography of salivary glands: an evolving approach for the diagnosis of Sjogren's syndrome. *Nat ClinPractRheumatol.* 2008;4:454–5. doi: 10.1038/ncprheum0864.

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