

## Sjogren's syndrome variant: A Case Report

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### ABSTRACT

Sjogren's syndrome is a part of the disease where moisture producing glands are affected by auto immune disease. There is variety of symptomatology depending upon the type and extent of the involvement of the underlying glands. There is dryness of the mouth when salivary glands are involved and dryness of the eyes when lachrymal glands are affected. We present 43-years old female who had subclinical and confusing symptoms of swelling and dryness of mouth. The radiological evaluation by ultrasound (US), non contrast computerized tomography (NCCT), contrast enhanced computerized tomography (CECT) and magnetic resonance imaging (MRI) studies clinched the diagnosis along with the background of clinical spectrum.

**Keywords:** Sjogren's syndrome; autoimmune; salivary gland ; ultrasound ;CECT;MRI.

### INTRODUCTION

Sjogren's syndrome was named after Henrik Sjogren, a Swedish ophthalmologist in 1933. He first time introduced the term of keratoconjunctivitis sicca. This syndrome affects the production of tears and saliva. 0.2-1.2% population is affected by this syndrome and the intensity and pattern may vary from person to person. There is predilection for the females with 9:1 as female male ratio.<sup>[1]</sup> There is always risk of lymphoma in 5 % of affected cases.<sup>[2]</sup>

### CASE REPORT

43-years old lady reported with the complaints of mild swelling of both the parotid gland region for the last three years. This swelling was off and on appearing with dull pain. Now for the last three months she was complaining of dryness of mouth. She had taken medication for symptomatic complaints which were subsiding and appearing again. On examination she was averagely built with normal physical parameters as per her age. Systemic examination was non contributory except mild enlargement of both the parotid regions [Figure 1 a,b and c]. Biochemical investigations were within normal limits. HIV status was negative. Ultrasound

examination had revealed bilateral multiple cystic anechoic areas within the parenchyma of parotid glands. No calcification or increased vascularity was noticed in the glands [Figure 2]. The patient was subjected to plain as well as contrast enhanced computerized tomography (CECT) of the face and neck regions. There were multiple well defined hypodense areas seen in both the glands involving superficial as well as deep lobes. Submandibular glands were not appreciated properly [Figure 3a and b], CECT had shown these hypodense areas as non enhancing areas as not part of the glandular parenchyma [Figure 4]



**Figure 1:** Photograph of 43-years old female having bilateral parotid swellings.(a) front view depicting both parotid regions (black arrows ). (b) left side view of left parotid swelling (white vertical hollow arrow) (c) right side view of parotid region (horizontal arrow)

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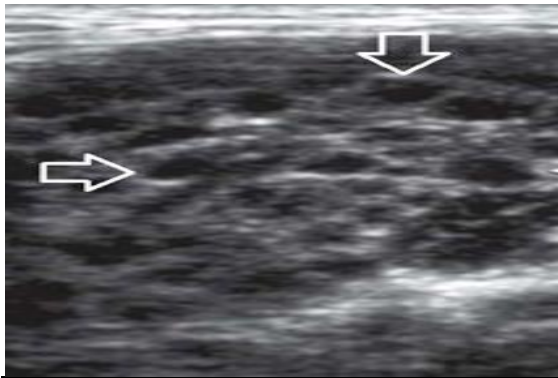


Figure 2: Ultrasound image of the right parotid gland. There are multiple discrete cysts seen in the glandular parenchyma with echogenic intercystic regions (white arrows).

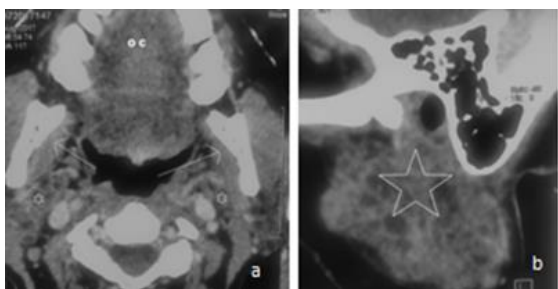
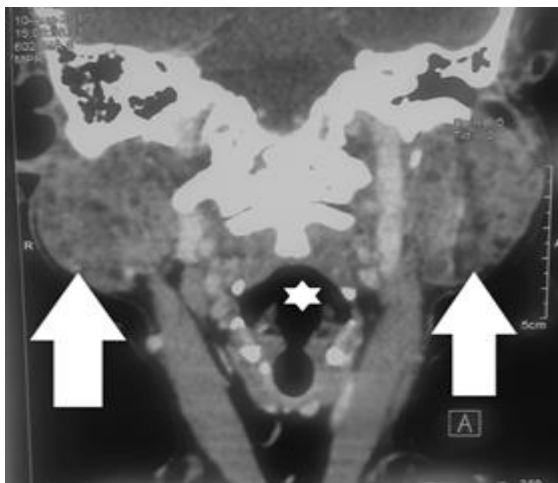


Figure 3: NCCT parotid region at oral cavity level.(a) Oral cavity seen anteriorly (oc) with deep part of both the parotid glands with multiple cystic areas (small stars).Both the submandibular glands appears to be small or atrophied (arrows).(b) Reformat oblique section of left parotid gland shows multiple cystic areas spread diffusely (white star).



Figures 4: Coronal reformat section of both parotid glands. There are multiple non enhancing cystic areas within the enhancing parenchymal component of both the glands (white solid arrows).Oral cavity is seen in the centre occupied by the air (solid star).

Patient was subjected to Magnetic resonance imaging (MRI) of the salivary gland regions. There were similar multiple hypointense well defined lesions in both the glands on T1WI and seen as hyperintense on T2WI [Figure 5 a and b].

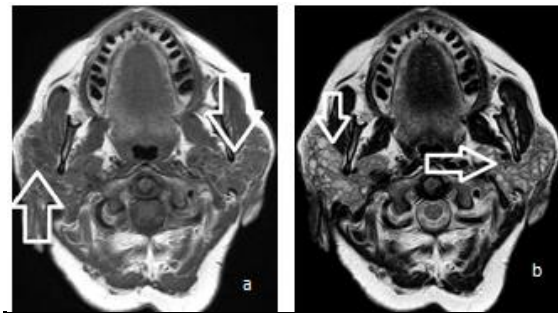


Figure 5: Axial sections of both parotid glands.(a) T1W image shows multiple hypointense lesions spread diffusely in both glands (hollow white arrows).(b) T2W image shows multiple hyperintense lesions spread diffusely in both the glands(hollow white arrows)

Saturation tau inversion recovery (STIR) images clearly highlighted the hyperintense nature of the lesions in superficial and deep lobes of both the parotid glands [Figure 6]

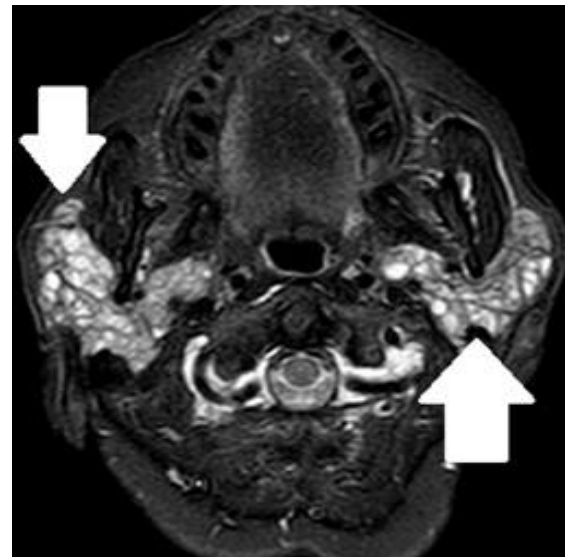


Figure 6: Saturation Tau Inversion Recovery (STIR) axial section of parotid gland level. Multiple cysts can be seen evenly spread in superficial as well as deep lobes of both the parotid glands (solid white arrows). The major part of parenchyma has been replaced by the cystic lesions (non-functional).

The patient was diagnosed as a case of sjogren's syndrome variant keeping in view of the clinical features and radiological evaluation. She had been kept on symptomatic treatment with regular monthly follow up.

## DISCUSSION

Sjogren's syndrome is of long time autoimmune disease where the moisture producing glands are affected. Xerostomia and xerophthalmia are the most common features of the syndrome.<sup>[3]</sup> Other features are in the form of dry skin, vaginal dryness, cough, numbness in the limbs with muscle and joint

pains.40% cases are presented as isolated cases . Genetic and environmental background leads to most of these cases.<sup>[4]</sup> The affected females are usually above 40 years and sometimes it is mistaken as that of age related changes. Rose Bengal and Schirmer's test are done to confirm the findings. There are two kinds of the entity as:

a) Primary type.

b) Secondary connected to other types of connective tissue disorders. Various associated conditions are celiac disease, SLE (systemic lupus erythematosus), fibromyalgia, MS(multiple sclerosis) and malignancies.

The hormonal factor as that because of oestrogen leads to cell mediated immune response. The underlying inflammation destroys the glands and affects the functioning.<sup>[5]</sup> Ultrasound examination shows early hyperechoic appearance but shows multicystic appearance in long standing cases as was our case. Cross sectional imaging by CECT and MRI delineate more tissue characterization. The diagnosis can be made by either biopsy or by blood test for specific types of circulating antibodies .The mainstay of the treatment is symptomatic.<sup>[6]</sup>

## CONCLUSION

Sjogren's syndrome can be isolated with only of mild complaints as was our case where there was only mild xerostomia was present.This could be confusing as this is regarded as age related outcome. But when the patient undergoes radiological evaluation by US, CECT and MRI,the diagnosis is unfolded. If not treated properly then rare complications can arise sometimes in the form of malignant lymphomas.

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