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# Sjögren Syndrome: Description of 4 Cases Guinean

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**Abstract:** ***Sjögren syndrome is an auto-immune inflammatory epithelitis with female predominance, characterised by dry mouth and eyes, worsened quality of life and rarely by systemic complications. We report 4 cases presented in Ignace Déen University Hospital. The diagnosis was reached by clinical and laboratory findings, according to 2002 American-European consensus criteria and ACR 2012 classification criteria. Cases of 3 women and 1 man with a mean age of 41 years were collected. They all suffered from oral and eye dryness and the most common extra glandular manifestations was articular involvement. Anti-SSA and anti-SSB were positive in all patients. Minor salivary gland biopsy was performed in 2 patients. Treatment included steroids, Plaquenil and Methotrexate. In conclusion Sjögren syndrome, although rare in Africa, may represent a real therapeutic challenge.***

**Keywords:** Sjögren syndrome,antiSSA and SSB, Gunea

## 1. Introduction

Sjögren syndrome is a chronic, systemic, auto-immune inflammatory disease, characterised by lymphocytic infiltration of the exocrine glands, especially salivary and lacrimal glands, causing mouth and eye dryness [1,2]. Ss is 10 times more frequent in women with a peak incidence between the ages of 50 and 80 years[3]. Less frequently the disease may affect the skin, lungs, GI tract, central and peripheral nervous system and musculoskeletal system. The disease may be primary when it occurs alone or secondary when associated with other auto-immune diseases as rheumatoid arthritis, SLE, systemic sclerosis [1,2,4]. Ss is rarely described in sub saharan Africa. We report 4 cases of Ss in Guinea.

## 2. Observation

**Case 1:** We report a case of a 33 years old patient, without a past medical history, who presented with chronic polyarthralgia in the last 4 years, characterised by exacerbations and remissions, involving, symmetrically hands, wrists, ankles and MTP joints. She also mentioned dry mouth necessitating frequent water ingestion with meals, and the inspection revealed a mouth ulcer. She reported also foreign body sensation in the eyes and diminution of tears. Dryness of the skin and the vagina were also revealed. The patient declined taking any medication ( benzodiazepines, anti depressants, neuroleptic, calcium inhibitors) or smoking. Clinical examination showed a patient in a good general status, synovitis of the right wrist and pain in active movement of left wrist, ankles and the squeeze test was positive in MTP joints. The rest of the examination was unremarkable except a swelling of the parotid glands.

Laboratory tests showed a normal blood count, increased phase reactants with ESR 31mm/h and CRP at 10.6mg/l. ANA were positif in a titre of 1/640 in Hep-2 cells. Double stranded anti DNA were negative. ENA were also positive with a strong positivity for anti- SSA. Anti-SSB, anti RNA, anti Sm, Jo-1, Scl 70, anti centromere were negative. Rheumatoid factor, anti-CCP and anti-phospholipid antibodies were also negative. No complement consumption or deficiency was revealed. All tests for the possibility of an infection were negative.

X-rays of the chest, hands and feet did not show any abnormalities.

Schirmer test was 5mm/5min bilaterally and the salivary flow without stimulation was < 1.5ml/15 min. Minor salivary gland biopsy was positive for Ss with a score of grade III by Chisholm and Mason. The diagnosis of Ss was made according to 2002 American-European consensus criteria and ACR 2012 classification criteria. Treatment included hydroxychloroquine and low dose steroids.

**Case 2:** A 55 years old female was addressed by her ophthalmologist due to eye dryness ( Schirmer test <4mm/5min) leading to artificial tears application 5 times per day evolving over the past 3 years. Clinical examination showed a symmetric polyarthritis involving wrists, right elbow, and knees associated to mouth and eye dryness, rosacea, and impression of depression. Laboratory tests showed a normal blood count, increased ESR (35mm/h) and a normal CRP. All auto-antibodies (ANA, ENA, anti-CCP) were negative. Tests for hepatitis C, HIV were also negative. X-rays of the chest, hands and feet were unremarkable.

Biopsy of minor salivary glands showed a grade IV score by Chisholm and Mason. The diagnosis of Sc was reached after clinical manifestations, Schirmer test and

the results of the biopsy. Treatment with Plaquenil 200mg was initiated.

**Case 3:** A 35 years old female patient was presented dut to mouth dryness, difficulty in swallowing, taste disturbance associated with bilateral parotid gland swelling. She also reported foreign body and burning sensation in the eyes. Clinical examination of the oral cavity showed a dry tongue with fissures at the edges. Laboratory tests were positive for the ANA, anti-SSA, anti-SSB, while the anti-dsDNA, anti-CCP and rheumatoid factor were negative. Schirmer test was <5mm/5min and non stimulated salivary flow was <1.5ml/15min. Ultrasound of salivary glands showed multiple cysts. The diagnosis was reached after the dryness syndrome, positivity of antibodies anti-SSA and anti-SSB. Treatment consisted of Plaquenil 200mg and Medrol 4mg.

**Case 4:** A 42 years old male with diagnosed rheumatoid arthritis since 2012 according to 2010 ACR/EULAR criteria presented himself in January 2014 with ocular dryness and a foreign body sensation, mouth dryness associated with synovitis of the elbows, wrists and hands. He was tested positive for the rheumatoid factor, the anti-CCP, anti-SSA and anti-SSB. All tests for a possible infection were negative. Non stimulates salivary flow was <1.5ml/15min.

Beyond the diagnosis of rheumatoid arthritis, based on the dryness syndrome, the antibodies positivity and the reduces salivary flow a diagnosis of secondary Sc was reached. He was treated with Methotrexate, Plaquenil et low-dose steroids.

**Table 1:** Clinical and Paraclinical Characteristics of Sjogren's Syndrome of 4 Patients

| Observations | 1 | 2 | 3 | 4 |
|---|---|---|---|---|
| Sex | Female | Female | Female | Male |
| Age | 33 years | 55 years | 35 years | 42 years |
| Dry syndrome oral | yes | yes | yes | yes |
| Dry syndrome ocular | yes | yes | yes | yes |
| ESR (mm/h) | 31 | 35 | 20 | 22 |
| CRP (mg/l) | 10,6 | negative | negative | 20,2 |
| Anti-SSA or SSB | SSA / Ro ++ | negative | SSA/Ro++ SSB/La ++ | SSA/Ro++ SSB/La ++ |
| RF/Anti-CCP | negative | negative | negative | Ant-CCP >250UI/ml |
| Non stimulated salivary flow | < 1,5 ml / 15 minutes | Non fait | < 1,5ml /15 minutes | < 1,5ml /15 minutes |
| Schirmer test | 5 mm/5 minutes | 4mm/5 minutes | <5mm/5 minutes | Not done |
| Minor salivary gland biopsy | stage III | stage IV | Not done | Not done |

## 3. Discussion

Sjögren syndrome is an auto-immune epithelitis, characterises by lymphocytic infiltration the exocrine glands, frequently under diagnosed, which affects 1-3% of the general population with a female predominance ( sex ratio:9/1)[6]. In the African literature the disease is mostly reported in Norther Africa[7]. In subsaharan Africa, most of the reports come from case studies, especially in Senegal[8]. Our case series present primary Sjögren syndrome in 3 women and a secondary one in a man. It is considered primary when it runs alone and secondary when follows another auto-immune disease like SLE ( 15-36%), rheumatoid arthritis ( 20-32%) and systems sclerosis (11-24%) and less frequently multiple sclerosis, hepatitis and auto-immune thyroiditis[9].

Sjögren syndrome is of unknown aetiology[3]. We may assume a multifactorial origin coming from genetic and epigenetic factors like infections, endocrine alterations, metabolic or psychological factors orchestrating together [10]. Whatever the cause, epithelial cells become capable to synthesise cytokines, chemokines, auto-antibodies present in laboratory analysis. Consequently the epithelitis is followed by lymphocytic infiltrations which may facilitate the formation of pseudo-follicules where auto-reactive B lymphocytes reside[11].

In our case series antibodies were positive in 3 out of 4 patients.

Anti-SSA and ant-SSB antibodies are present in 75% of patients with Ss[12]. Absence of antibodies does not rule out the disease as 5-30% of patients are seronegative[13]. Furthermore anti-SSA and anti-SSB are not disease specific, as they may be encountered in other auto-immune diseases ( SLE, inflammatory myopathies), chronic viral infections, and sometimes in healthy individuels.

Biopsy of salivary glands is not always accessible and was performed in 2 out of 4 of our patients.

In case of Ss suspicion we have to rule out an HIV, Hepatitis C, HTLV infection or a monoclonal gammopathy which may present with a similar clinical picture[14].

## 4. Conclusion

Sjögren syndrome is an auto-immune epithelitis with a female predominance, characterised by ocular and mouth dryness and worsening of quality of life. The disease represents a real therapeutic challenge in African hospitals.

## 5. Conflicts of Interest

None

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