Acta Scientific Otolaryngology (ISSN: 2582-5550)

Volume 5 Issue 4 April 2023

Case Report

Sjögren's Syndrome: An Incidental Radiologic Finding in a Patient with Aural Symptoms

Ukamaka Nwankwo*[1], Ifedayo Ajayi[2], Junaid Ayinde[3] and Ibukun Abidoye[4]

[1]*ENT Surgeon, Evercare Hospital Lekki, Nigeria*

[2]*Consultant Radiologist, Department of Radiology, Evercare Hospital Lekki, Nigeria*

[3]*Consultant Radiologist, Department of Radiology, Evercare Hospital Lekki, Nigeria*

[4]*Consultant Radiologist, Department of Radiology, Evercare Hospital Lekki, Nigeria*

***Corresponding Author:** Ukamaka Nwankwo, ENT Surgeon, Evercare Hospital Lekki, Nigeria.

DOI: 10.31080/ASOL.2023.05.0551

Received: February 20, 2023

Published: March 21, 2023

Abstract

Sjögren's syndrome, an immune-mediated disorder of the exocrine glands (lacrimal and salivary) affects acinar cell serous production leading to complaints of dryness of the eyes and mouth. Radiological imaging has been used as a non-invasive modality for the diagnosis of this condition.

This is a case report of incidental diagnosis of Sjögren's syndrome in a 49-year-old female with left aural fullness and recurrent tinnitus. Incidental findings of scattered micro-calcifications and heterogeneity were seen in both parotid glands on computed tomography imaging and innumerable microcysts of low signal intensity on T1 and high signal intensity on T2/STIR, on magnetic resonance imaging of the temporal bone. These findings are pathognomonic for Sjögren's syndrome.

The meticulous review of computed tomography and magnetic resonance images help to detect silent lesions and the awareness of audio-vestibular involvement in Sjögren's syndrome can lead to prompt diagnosis and the monitoring of disease progression.

Keywords: Sjögren's Syndrome; Parotid Gland; Computed Tomography; Magnetic Resonance Imaging; Lacrimal Gland; Pneumatised Temporal Bone; Aural

Introduction

Sjögren's syndrome (SS) is a long-course, inflammatory, immune-mediated, clinical condition caused by lymphocyte exocrine gland invasion [1]. Exocrine glands involved are the lacrimal and salivary glands with complaints of dryness of the eyes (xerophthalmia) and mouth (xerostomia) [2,3]. There could also be bilateral enlargement of the parotid and lacrimal glands. SS can be of two types: primary and secondary. The secondary type is characterised by association with other immune- mediated diseases like rheumatoid arthritis, systemic lupus erythematosus, polymyositis or systemic sclerosis [4]. The frequency of SS is higher in females with double age peaks: 20 to 40 years and 50 to 60 years [5]. Periductal lymphocyte invasion and epimyoepithelial islands are seen on histological analysis which may become malignantly transformed giving rise to lymphoma. As the disease progresses, fatty degeneration of the gland is noted [4].

Aural complaints are early symptoms or the only presentation in some patients with SS. This is however not common. Sensorineural hearing loss is the most noted aural presentation [6] and almost 25 percent of patients with SS have hearing impairment affecting high frequencies (more than 9KHz in majority of them) [6,7] and since speech frequencies are not involved, hearing impairment may not be noticed. Vertigo, aural fullness and tinnitus can also occur in affected patients and may be misdiagnosed as Meniere's disease [8]. Audio-vestibular involvement is bilateral in most cases [8].

SS has no "gold standard" test for diagnosis [9,10] but diagnosis can be made in various ways. It may be diagnosed serologically, histologically (labial gland biopsy) or radiologically. X-ray sialography, histology and serology tests are invasive. There are dependable, non-invasive radiological investigations used for diagnosis [11]. These imaging studies include ultrasonography (USG), computed tomography (CT) imaging and magnetic resonance imaging (MRI). These investigations are also used for periodical evaluation, monitoring of disease progression and detection of possible complications.

A meta-analysis of 14 studies revealed collective sensitivity of 75% and specificity of 93% in precise diagnosis of SS with USG [12]. Ultrasound findings are mixed or heterogenous echogenicity with numerous hypoechoic points in the affected exocrine gland [2]. Fibrous proliferation, noted as multiple hyperechoic regions are found in advanced cases [13]. Detection of hypoechoic regions has been determined to be the most distinguishing feature in USG of affected salivary gland [14]. These findings are similarly noted in long standing sialadenitis [4] and this may lead to missed diagnosis in some cases. USG is also skill dependent and therefore, reports may be biased [15]. Despite this, USG is used in a lot of settings because it is affordable, accessible and easy to perform [16].

The density of parotid gland in SS is higher than normal glands on CT imaging in early involvement of the gland, signifying lymphocytic infiltration. Tiny sized, fluid density, cystic lesions are also seen. As the disease progresses, density reduces due to the presence of atrophied or destroyed tissues and accumulation of fat cells [17]. This is responsible for the heterogeneous appearance on CT imaging. Macrocytic alterations and lymphoid aggregations are also noted as solid enhanced nodules [18].

MRI is regarded as an objective diagnostic radiological imaging. It is costly and not readily affordable. Numerous points of high intensity are seen in T2 weighted imaging and MR sialography while fatty degeneration is the hallmark in T1 weighted imaging [2].

Fatty degeneration on MRI and hyperechoic regions found in USG are characteristic of impaired flow of saliva. This suggests impaired salivary gland function and is commonly noted in progressive disease [2].

Case Report

A 49-year-old female presented at the ENT clinic in Evercare Hospital, Lekki, Nigeria with three weeks history of left aural fullness and recurrent non-pulsatile tinnitus. Otoscopy revealed normal findings and hearing assessment was found to be normal. A computed tomography (CT) scan of the temporal bone was done which revealed bilateral, symmetrical, pneumatized temporal bone apices; normal variant (Figure 1 and 2).

Figure 1: Bilateral pneumatization of temporal bone apices.

Figure 2

Interestingly, scattered micro-calcifications and heterogeneity were incidentally seen in both parotid glands on CT imaging (Figure 3).

Figure 3

Magnetic resonance imaging was done for better soft tissue delineation. This also confirmed natural pneumatization of both temporal bone apices (Figure 4). Innumerable microcysts were noted to be disseminated in both parotid glands, of low signal intensity on T1 (Figure 5) and high signal intensity on T2/STIR, giving it a "honeycomb" appearance (Figure 6 and 7).

Figure 4: MRI depiction of temporal bone pneumatization.

Figure 5: MRI T1- weighted image of parotid gland.

Figure 6: T2/STIR imaging of parotid gland.

Figure 7: T2/STIR imaging of parotid gland.

The lacrimal glands were found to be normal sized and homogenous in appearance on MRI (Figure 8).

Figure 8: Lacrimal gland T2/STIR imaging.

On further inquiry, patient revealed that she has been experiencing dryness of both eyes but no dryness of the mouth, joint pain or swelling. She was then referred to see the Rheumatologist.

Discussion

In SS, disorder of the exocrine glands (lacrimal and salivary) affects acinar cell production of serous secretion be it tears or saliva. This leads to complaints of dryness of the eyes and mouth. At disease onset, lymphocyte invasion of exocrine glands causes connective tissue thinning and breakage of duct walls but in later stages, continuous invasion by lymphocytes which surrounds serous acini, causes atrophy and destruction [19], leading to impaired saliva production [20]. This subtle autoimmune disorder is associated with a sixteen to forty percent increased risk for lymphoma [1,21]. Therefore, early detection of this condition is vital.

In the early stages of the disease, the affected parotid gland may appear normal on imaging and diagnosis may be missed. The duration between inception and accurate diagnosis ranges from four to ten or more years [22]. That is why there is no lone test for the diagnosis of SS to minimise missed diagnosis. Serologic detection of SS-A and SS-B antibodies are positive mainly in the active period of the disease. Serologic negative forms are occasionally seen and mostly in males [4]. Labial exocrine gland biopsy for histological diagnosis is a standard test but it is not without its disadvantages which include pain, unrepresentative tissue diagnosis in early stage of the disease [23] and sometimes, tissue findings in the normal parotid gland of the elderly are similar to that in individuals with SS [24].

There are diverse imaging modalities used to examine the structure of salivary glands in SS. USG is mostly used due to its non-invasiveness, availability and affordability [16], then MRI. CT imaging and contrast X-ray sialography are not frequently used. Findings on USG of affected salivary glands can enhance diagnostic performance of SS classification criteria [25] but the drawback of USG is that it is operator dependent and distinguishing SS from other diseases like lymphoma may be difficult. USG finding may be normal in the early stage of the disease and diagnosis can be missed [26].

Micro or spotted calcifications in the parotid glands bilaterally on CT imaging has a low sensitivity but high specificity for the diagnosis of SS [27]. This makes it a dependable, radiological diagnostic tool in SS. Heterogeneousness, scattered microcalcifications and atypical fat deposition are distinct findings in SS on CT imaging [27]. In this patient, scattered micro-calcifications, fatty deposition and heterogeneity were noted which are typical findings seen in the intermediate stage of this disease condition. She was referred

to the rheumatologist for further assessment and commencement of treatment to limit the progression of this disease. This diagnosis may be missed on CT imaging as the above stated typical findings are like those found in long standing sialadenitis. Low sensitivity and radiation exposure has limited the use of CT imaging for the diagnosis of SS.

Homogeneous signal intensity (MRI) is seen in non-diseased parotid gland on T1 and T2 weighted imaging which is lost in SS [28]. Multiple, distinct, diffusely spread nodules of varying sizes are characteristic findings on MRI [29]. Bilateral parotid gland enlargement may also be seen on MRI due to excess fat deposition. High signal is noted on T2-weighted imaging. These findings give the affected parotid gland a "honeycomb" or "salt and pepper" appearance. In this patient, innumerable microcyst were seen of low signal intensity in T1 and high signal intensity on T2/STIR imaging with the typical "honeycomb" or "salt and pepper" appearance further confirming CT imaging suggestion of SS. MRI sialography was not done to assess the ductal system. As this patient is yet to experience xerostomia, it is likely that there are still functioning acinar cells and salivary ducts in the parotid gland bilaterally.

In MRI sialography, T2-weighted sequences with fat suppression is done [18]. Saliva serves as contrast in MRI sialography and highlights constrictions, expansions or strictures involving the ductal system in parotid and submandibular glands [4]. Strictures of the ducts appear as "string of beads" in MRI sialography. MRI and MR sialography findings in patients with SS have been found to tally with histology findings in minor salivary gland biopsy [30]. The appearance of numerous high signal intensity dots in MR sialography is being advocated as the prime diagnostic marker for SS [31,32]. Sialography in MRI, a non-invasive procedure with no exposure to radiation, is by far, a better alternative to X-ray sialography. In X-ray sialography, salivary gland duct is cannulated for introduction of contrast material, and this can be a painful process. There is also, the exposure to radiation and possible perforation of the duct. Sialectasis due to accumulation of contrast material and sparseness of ducts are findings that may be noted in affected glands [11].

Izumi and his colleagues [33] in their study categorized lacrimal glands in patient with SS according to size: normal, hypertrophied and atrophied. Lacrimal gland serous acinar function in hypertrophied glands was noted to be moderately reduced, that of atrophied glands, markedly reduced and that of normal sized glands ranged from mild to severe reduction. The varied function in those with normal sized glands may depend on the stage of disease. Lacrimal function is normal in disease onset but reduces as disease progresses despite that fact that the gland may still appear normal on imaging. They also noted that lacrimal function in patients with normal sized glands with homogeneity on MRI was better than those with heterogeneous appearance. They proposed that the homogeneous appearance on MRI was typical of early onset of disease. Fat cell deposition was found to be more in the glands of patients with SS. Accelerated accumulation of fat cells may lead to alteration in the size of affected lacrimal glands on MRI [26]. The lacrimal gland finding on MRI in this patient was normal and homogeneous, but she has been experiencing dryness of the eyes which suggests impaired acinar serous production due to disease progression and probably atrophy and destruction of acinar cells and ducts. This tallies with intermediate disease suggestion of MRI and CT imaging findings in the parotid gland of this patient.

Though sensitivity and specificity for SS is better in MRI and MR sialography compared to other radiological investigations, its use is limited due to its high cost, limited accessibility and claustrophobia. It is contraindicated in patients with metallic implants, pacemakers and cerebrovascular cramps [34].

MRI findings correlate with serological and clinical findings in SS making it easy to diagnose SS radiologically. There is also good correlation with USG findings [27]. MRI has been found to have the best sensitivity and specificity for imaging of the salivary glands compared to CT imaging and USG [35], it also highlights surrounding structures better than CT imaging and USG [34,36]. MRI helps to determine the stage of the disease and distinguish between SS and other causes of xerostomia [18]. MRI and CT imaging can also be used to observe response to therapy and screen for lymphoma. This patient will benefit from yearly imaging to monitor disease progress or possible malignant transformation.

Pneumatization of temporal bone apices seen both on CT imaging and MRI in this patient is a normal variant. Pneumatization has been found to be present in 9 to 30 percent of people and the

correlation between extent of mastoid air cells pneumatization and that of temporal bone apex is positive [37]. These pneumatized air cells are prone to disease of other pneumatized areas of the temporal bone like infection, inflammation and fluid accumulation [37,38].

This extensive temporal bone pneumatization has been suggested to be associated with some complaints [39]. While some studies have noted the likely association between tinnitus and well pneumatized temporal bone and apex [40-42], others have found no relationship [37]. It has been proposed that pneumatization between the cochlea and internal carotid probably acts as amplifiers, increasing vascular supply to the cochlea and subsequently, tinnitus [37]. Tinnitus in this patient is only limited to the left ear and heard occasionally. Both apices were found to be symmetrically pneumatized on CT imaging and MRI. No abnormality was detected. Therefore, tinnitus experienced by this patient may be due to other factors and not pneumatization of petrous bone apex.

The incidence of aural presentation in SS ranges from 22 to 46 percent [8]. This may actually be more due to missed diagnosis or ignorance of its occurrence in those with SS. The trigger and process of inner ear involvement in SS is not known and it has been suggested that accumulation of autoantibodies and immune mediated cytotoxicity may lead to disruption of inner ear function and audio-vestibular complaints [43]. This accumulation of immune complexes reduces the diameter of vessels impairing blood supply, causing destruction of hair cells and spiral ganglion [44].

The accumulation of immune complexes reduces the diameter of vessels impairing blood supply, causing destruction of hair cells and spiral ganglion [44].

This patient presented with left aural fullness and recurrent tinnitus. There was no complaint of vertigo. Hearing assessment was found to be normal. This may not exclude hearing impairment in this patient as high frequencies more than 9KHz are involved in most people with SS [6,7]. These very high frequencies were not assessed in this patient because they are not routinely done. The auditory complaints in this patient were the indication for radiological investigation which led to the detection of this disease. Though audiovestibular presentations are said to be bilateral in most cases [8], unilateral presentation can occur as in this patient. The diagnosis of SS would have been missed in this patient if not for the incidental radiological finding. She regarded the dryness of her eyes as being insignificant. The recovery of aural function would have been affected as the wrong treatment would have been instituted. Therefore, awareness should be created on the aural presentation of SS as this is a rare manifestation. This will greatly help the early diagnosis of this disease and its management. A high index of suspicion should also be raised in those with immune-mediated disorders and aural complaints.

Conclusion

CT and MR imaging are beneficial for the diagnosis of Sjögren's syndrome. They are very useful for incidental detection, local staging, monitoring of the disease process and early detection of malignant transformation. The meticulous review of these images is vital for incidental diagnosis of disease conditions. The awareness of aural involvement in SS could lead to prompt diagnosis and recovery of function if the right treatment is commenced. It can also be used for the monitoring of disease progression or recovery.

Bibliography

1. Skarlis C., *et al.* "Sjögren's syndrome: Recent updates". *Journal of Clinical Medicine* 11.2 (2022): 399.

2. Takagi Y., *et al.* "Comparison of salivary gland MRI and ultrasonography findings among patients with Sjögren's syndrome over a wide age range". *Rheumatology* 61.5 (2022): 1986-1996.

3. Van Ginkel MS., *et al.* "Imaging in primary sjögren's syndrome". *Journal of Clinical Medicine* 9.8 (2020): 2492.

4. Danchaivijitr N., *et al. "*MRI of the parotid glands in a patient with Sjögren's syndrome". *Eurorad* (2006).

5. Fox RI. "Sjögren's syndrome". *Lancet* 366 (2005): 321.

6. Okawa Y and Ihara K. "Sensorineural hearing loss in sjögren's syndrome". *International Journal of Molecular Sciences* 23 (2022): 11181.

7. Tucci M., *et al.* "Sjögren's syndrome: an autoimmune disorder with otolaryngological involvement". *ACTA Otorhinolaryngologica Italica* 25.3 (2005): 139-144.

8. Ralli M., *et al.* "Audiovestibular symptoms in systemic autoimmune diseases". *Journal of Immuno Research* 2018 (2018): 5798103.

9. Vitali C and Del Papa N. "Classification and diagnostic criteria in sjögren's syndrome: A long-standing and still open controversy". *Annals of the Rheumatic Diseases* 76 (2017): 1953-1954.

10. Bootsma H., *et al.* "Toward new classification criteria for sjögren's syndrome?" *Arthritis and Rheumatology* 65 (2013): 21-23.

11. Baldini C., *et al.* "Imaging in primary sjögren's syndrome: The "obsolete and the new". *Clinical and Experimental Rheumatology* 36 (2018): 5215-5221.

12. Zhou M., *et al.* "Diagnostic accuracy of salivary gland ultrasonography with different scoring systems in Sjögren's syndrome: a systematic review and meta-analysis". *Scientific Report* 8.1 (2018): 17128.

13. Carotti M., *et al.* "Diagnostic value of major salivary gland ultrasonography in primary sjögren's syndrome: The role of grey-scale and colour/power doppler sonography". *Gland Surgery* 8 (2019): S159-S167.

14. Mossel E., *et al.* "Scoring hypoechogenic areas in one parotid and one submandibular gland increases feasibility of ultrasound in primary sjögren's syndrome". *Annals of the Rheumatic Diseases* 77 (2018): 556- 562.

15. Cornec D., *et al.* "Salivary gland ultrasound to diagnose Sjögren's syndrome: a claim to standardize the procedure". *Rheumatology (Oxford)* 54.1 (2015): 199-200.

16. Afzelius P., *et al.* "Imaging of the major salivary glands". *Clinical Physiology and Functional Imaging* 36.1 (2016): 1 -10.

17. Izumi M., *et al.* "Premature fat deposition in the salivary glands associated with Sjögren's syndrome: MR and CT evidence". *American Journal of Neuroradiology* 18 (1997): 951-958.

18. Lemon SI., *et al.* "Salivary gland imaging in Sjögren's syndrome". *Future Rheumatology* 2.1 (2007): 83-92.

19. De Vita S., *et al.* "Current limitations and ongoing iniatives to evaluate the activity, damage and outcome of primary Sjögren's syndrome". *Clinical and Experimental Rheumatology* 37.118 (2019): 24-26.

20. Sreebay LM. "Saliva in health and disease: an appraisal and update". *Pathology - Research and Practice* 50.3 (2000): 140-161.

21. Theander E and Mandl T. "Primary Sjögren's syndrome: diagnostic and prognostic value of salivary gland ultrasonography using a simplified scoring system". *Arthritis Care and Research* 66.7 (2014): 1102-1107.

22. Peck AB and Ambrus JC. "A temporal comparative RNS transcriptome profile of the annexon gene family in the salivary versus lacrimal glands of the sjögren's syndrome susceptible C57BL/6.NOD-Aec2 mouse". *International Journal of Molecular Sciences* 23 (2022): 11709.

23. Vitali C., et al. "Parotid sialography and minor salivary gland biopsy in the diagnosis of Sjögren's syndrome: a comparative study of 84 patients". *Journal of Rheumatology* 15 (1998): 262-267.

24. Fox RI., et al. "Sjögren's syndrome: proposed criteria for classification". *Arthritis and Rheumatology* 29 (1986): 577-585.

25. Cornec D., *et al.* "Contribution of salivary gland ultrasonography to the diagnosis of Sjögren's syndrome: Havard new diagnostic criteria". *Arthritis and Rheumatology* 65.1 (2013): 216-225.

26. Ching A and Ahuja A. "Resolution on sonography of the submandibular space: Anatomy and abnormalities". *AJR American Journal of Roentgenology* 179.3 (2002): 703-8.

27. Sun Z., *et al.* "Diagnostic accuracy of parotid CT for identifying Sjögren's syndrome". *European Journal of Radiology* 81.10 (2012): 2702-2709.

28. Izumi M., *et al.* "MR imaging of the parotid gland in Sjögren's syndrome: a proposal for new diagnostic criteria". *AJR American Journal of Roentgenology* 166.6 (1996): 1483-1487.

29. Swiecka M., *et al.* "Imaging methods in primary sjögren's syndrome as potential tools of disease diagnostics and monitoring". *Rheumatology* 57 (2019): 336-342.

30. Niemala RK., *et al.* "Ultrasonography of salivary glands in primary Sjögren's syndrome. A comparison with magnetic resonance imaging and magnetic resonance sialography of parotid glands". *Rheumatology* 43.7 (2004): 875.

31. Andre R., *et al* "Comparison of clinical characteristics and magnetic resonance imaging of salivary glands with magnetic

resonance sialography in sjögren's syndrome". *Laryngoscope* 131.1 (2021): E83-E89.

32. Kojima I., *et al.* "Diagnostic performance of MR imaging of three major salivary glands for Sjögren's syndrome". *Oral Disease* 23.1 (2016): 84-90.

33. Izumi M., *et al.* "MR features of the lacrimal gland in sjögren's syndrome". *AJR* 170 (1998): 1661-1666.

34. Freling NJ. "Imaging of salivary gland disease". *Seminar on Roentgenology* 35.1 (2000): 12-20.

35. Freling N., *et al.* "Staging and follow-up of high-grade malignant salivary gland tumours: The role of traditional versus function imaging approaches-a review". *Oral Oncology* 60 (2016): 157-166.

36. Nishimura DA., *et al.* "Magnetic resonance imaging of salivary glands: a literature review". *Clinical and Laboratorial Research in Dentistry* (2019): 1-6.

37. Sayit AT., *et al.* "Temporal CT of the pneumatized petrous apex: Associated anomalies and the relationship with tinnitus and vertigo". *İstanbul Medical Journal* 19.3 (2018): 225-230.

38. Chapman PR., *et al.* "Petrous apex lesions: Pictorial review". *AJR American Journal of Roentgenology* 196.3 (2011): WS26-WS37.

39. Dong-Hee L., *et al.* "Anatomical factors influencing pneumatization of the petrous apex". *Clinical and Experimental Otorhinolaryngology* 8.4 (2015): 339-344.

40. Sosen E., *et al.* "Is there a relationship between subjective pulsatile tinnitus and petrous bone pneumatization?" *Journal of Craniofacial Surgery* 24.2 (2013): 461-463.

41. Topal O., *et al.* "Subjective pulsatile tinnitus associated with extensive pneumatization of temporal bone". *European Archives of Otorhinolaryngology* 265 (2008): 123-125.

42. Tuz M., *et al.* "Subjective pulsatile tinnitus associated with extensive pneumatization of temporal bone". *Auris Nasus Larynx* 30 (2003): 183-185.

43. Ruckenstein NJ. "Autoimmune inner ear disease". *Current Opinion in Otolaryngology and Head and Neck Surgery* 12.5 (2004): 426-430.

44. Di Stadio A and Ralli M. "Systemic lupus erythematosus and hearing disorders: literature review and meta-anslysis of clinical and temporal bone findings". *The Journal of International Medical Research* 45.5 (2016): 1470-1480.