Imaging of Sjogren syndrome in salivary glands

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Learning objectives

- The knowledge of the findings in different imaging modalities about Sjogren Syndrome (SS) in salivary glands.

Fig. 1: Salivary Glands

References: R. BECERRA ULLOA; Mexico City, MEXICO

Background
In the category of systemic disorders that affect the major salivary glands, there are a number of autoimmune diseases. HIV related cysts and nodules, Sjogren and sarcoidosis fall within the general rubric of autoimmune disorders with salivary gland manifestations. [2]

**Sjogren syndrome (SS)** is characterized by keratoconjunctivitis and xerostomía, that result from autoimmune destruction of the lacrimal and salivary glands, respectively. [1]

The disease affects predominantly **women older than 40 years**. [1]

It can occur as an **isolated syndrome** (primary SS, previously known as Mikulicz disease) or **in conjunction with associated autoimmune disorders** (secondary SS). [1,2]

Histologic análisis typically reveals lymphocytic and plasma cell infiltration and loss of normal gland architecture. When autoimmune disease affects the parotid gland, nonobstructive sialectasia occurs; it is initially punctate and progresses (in some) to globular, cavitary, and destructive changes. Whether these changes actually represent the underlying histopathologic characteristics of dilated hypertrophic ducts and acini surrounded and eventually occluded and destroyed by lymphocytic infiltration, or in fact reflect extravasation secondary to weakened duct walls and complications of secondary infection, remains a matter of some controversy. [1,7]

Conventional diagnostic methods used to detect SS-associated structural glandular changes in the exocrine glands include **ultrasonography (US)**, **x-ray sialography**, **CT sialography**, **MR sialography**, **magnetic resonance (MR)**, **scintigraphy and gland biopsy**. Most of these diagnostic methods are invasive, other techniques such as US and MR imaging have been used to reliably characterize and diagnose SS and offer promise as replacements for the earlier invasive and potentially harmful techniques. [1]

The knowledge of the findings in different imaging modalities about Sjogren syndrome in salivary glands it is important to suspect it. If we can do early diagnosis we will save money, time y often pain in others procedures.

**Imaging findings OR Procedure details**

The range of studies needed to assess salivary gland lesions spans a wide gamut. The algorithm for imaging the salivary glands depends on the clinical scenario with which the patient presents to the clinician. [2]
In most cases, the diagnosis of Sjogren disease is made clinically based on the sicca syndrome and the connective tissue disorder (eg, rheumatoid arthritis) combined with serology of antinuclear antibodies. [2]

**Scintigraphy**

H. M. Markusse et al. studied the diagnostic value of salivary gland scintigraphy in patients suspected of having primary SS. They studied in 149 patients. [3]

Salivary gland scintigraphy was abnormal in 19 out of 26 patients with primary SS but abnormal scintigrams were also found in 57 of the 123 patients without primary SS and in five of the controls. This resulted in a positive predictive value of an abnormal salivary gland scintigram of 25% and a negative predictive value of a normal investigation of 90%. [3]

From the high number of false positive test results in the patient population studied it is concluded that salivary gland scintigraphy has only a limited discriminatory value for the diagnosis of primary SS. [3]

E-K Tensing et al. found that the results of salivary scintigraphy can be predicted by diagnosis and autoimmune findings; psychological characteristics added 20% to this predictive value and distinction between SS and patients with sicca symptoms but without SS is difficult, but in addition to autoantibodies, salivary scintigraphy can be used for this purpose. [4]

The results of the study show that visual evaluation of scintigraphies is both sensitive and specific for the submandibular glands, with 85.3% sensitivity and 77.8% specificity. These results are similar to those described earlier by other groups using different evaluation methods. [4]

Winn Aung et al. evaluated quantitatively in healthy volunteers and in 70 patients with Sjogren's syndrome with salivary gland scintigraphy and labial biopsy. Using 99mTcsodium pertechnetate, dynamic scintigraphy and time-activity curves for the oral cavity and four major salivary glands were generated. They concluded that quantitative oral activity indices together with maximum accumulation and uptake ratio of the submandibular gland were sensitive enough to distinguish the disease severity of Sjogren syndrome. [5]
These quantitative variables may be used to identify the stage of Sjogren syndrome, and thereby replace the labial gland biopsy, or they can be used to determine the clinical stage in equivocal cases.[5]

**Fig. 2 on page 8  Fig. 3 on page 10**

**Sialography**

Sialography is used primarily to stage disease. For those cases in which the diagnosis is in question, some would favor performing a lip biopsy of the minor salivary gland tissue over sialography [2]

Clubbing is a cylindrical dilatation of a most peripherally placed duct and is the most common radiological finding in SS, present in 55% of the cases, this finding has great importance in early diagnosis of Sjogren's syndrome. [6]

Second sign is the punctate ectasis (in 28%). The remainder of the peripheral patterns is present in between 8 and 12% of the cases. [6] **Fig. 4 on page 10  Fig. 5 on page 11**

The other different patterns noted are the following: Peripheral sialectasis, punctate, globular, cavitary, cylindrical, fusiform. [6]

Sialography may be useful in staging Sjogren syndrome. [2]

**Ultrasound**

Sjögren’s syndrome can produce some punctate hypoechoic lesions with eventual cavitation and destructive changes. The gland can be normal or become enlarged and hyperechoic in the early processes. [8] **Fig. 6 on page 12  Fig. 7 on page 13**

In the late stage, the sonographic manifestations of a multicystic or reticular pattern in an atrophic gland are characteristic. The severity of parenchymal damage correlates with glandular vascularity on color-flow imaging. Patients with Sjögren’s syndrome require careful follow-up to detect possible association with lymphoma. [8]

Ultrasonography, underutilized in most North American sites, may supplant the role of CT and MR Imaging for superficial salivary gland lesions when experienced sonographers use the technique. [2]
Fig. 11: US images show Sjogren syndrome in the parotid gland.

References: R. BECERRA ULLOA; Mexico City, MEXICO

Gian Marco Giuseppetti et al. using a contrast-enhanced US imaging of the parotids with a second-generation contrast agent moreover analysis of time-intensity curves at rest and during salivary stimulation, found that contrast-enhanced US imaging allowed to discriminate Sjogren's from non-Sjogren's sicca patients with 87.5% sensitivity, 85% specificity and 86.7% accuracy and the primary from the secondary syndrome with 78.2% sensitivity, 70.5% specificity and 75% accuracy. [10]

**Computed Tomography (CT)**

Mainly CT is a poor technique for SS diagnosis but beyond of it´s sensibility, can orient the disorder. The imaging features of SS in CT keep relation with the other techniques depending the stage, in general a multicystic or reticular pattern in an atrophic gland. Fig. 8 on page 14 Fig. 9 on page 14

Sjogren syndrome and sarcoidosis predispose to stone formation. CT is probably the best way to image patients with systemic disorders since calculi may be at the root of or a byproduct of the acute symptoms related to the systemic disorder. [2]
**Magnetic resonance**

The normal parotid gland show homogeneous signal intensity on T1-weighted MR images. On the other hand, the parotid gland in patients with SS is characterized by a loss of homogeneity in signal intensity on T1-weighted MR images, with a resultant granular appearance. However, the parotid gland in patients with parotid inflammation also showed irregular signal intensity on T1-weighted MR images which, on occasion, is indistinguishable from that of the parotid gland in SS patients, especially when inflammation persisted. [9] Fig. 10 on page 15

Therefore, Masahiro et al. assessed changes in signal intensity patterns on T1-weighted MR images by calculating standard deviations of signal intensities for the parotid gland. [9]

Standard deviations of signal intensity for the panotid gland in patients with probable SS partially overlapped that for the panotid gland in normal subjects, but the difference is highly significant (p < .001). On the contrary, they found no significant difference in the standard deviations of signal intensities for the parotid glands in normal subjects and patients with panted inflammation. [9]

Caleb et al. used dynamic contrast enhanced MR imaging tracer kinetic model, and they estimated the volume of extracellular extravascular space to be greater in patients with SS than in healthy volunteers. They observed a higher degree of microvascular heterogeneity in the parotid glands of patients with SS compared with that in the parotid glands of healthy volunteers. [1]

They conclude that dynamic MR imaging has potential as a diagnostic tool for discriminating patients with SS from healthy individuals. [1]

SS increases the risk of parotid lymphoma by more than 4,400%. Therefore, any dominant mass in a Sjogren-affected parotid gland must be considered lymphoma and requires aspiration or biopsy. MR imaging has been particularly helpful with identifying dominant masses within glands affected by SS. [2]

**MR sialography**
The results of a prospective study comparing MR sialography with the standard of reference conventional sialography showed an agreement rate of 89% \((P < .001)\) in staging the disease. [2]

For identifying stage I or greater disease and stage II or greater disease, MR sialography was 100% and 91% accurate, respectively. Punctate, globular, and destructive patterns may be discerned with MR sialography. [2]

MR sialography is used primarily to stage disease. For those cases in which the diagnosis is in question, some would favor performing a lip biopsy of the minor salivary gland tissue over sialography. [2]

**Histopathological**

Biopsy of the parotid gland has a higher yield than a labial biopsy since the sensitivity for Sjogren disease with labial minor salivary gland biopsy is 58% compared to 100% for parotid biopsy. [2]

**Images for this section:**
Fig. 2: Scintigram showing normally functioning parotid and submandibular glands.

Fig. 3: Scintigram showing seriously damaged functioning of all parotid and submandibular glands.
**Fig. 4:** Conventional sialography. Parotid gland with punctate ecstasies, the extent of the ductal system and its tributaries is well seen on this conventional sialogram in a patient who has Sjogren disease.
Fig. 5: Sialography. Parotid gland with clubbing and punctate ectasis
**Fig. 6:** Gray-scale and Doppler. US images show advanced-stage Sjogren syndrome in the parotid gland. The gland has an inhomogeneous structure with multiple small, oval, hypoechoic areas and increased blood flow.
Fig. 7: Gray-scale and Doppler. US images show advanced-stage Sjogren syndrome in the parotid gland. The gland has an inhomogeneous structure with multiple small, oval, hypoechoic areas and increased blood flow.

Fig. 8: Sagital and Coronal CT images of parotid gland in 48-year-old woman with Sjogren’s syndrome. CT images shows heterogeneous appearance and characteristic focal low-density areas.
**Fig. 9:** Sagital and Coronal CT images of parotid gland in 48-year-old woman with Sjogren's syndrome. CT images shows heterogeneous appearance and characteristic focal low-density areas.
Fig. 10: Sjogren disease. While both parotid glands (arrowheads) show cystic changes in and enlargement of the gland on this coronal T2-weighted SE (4,000/80) MR image, the left side also shows periparotid adenopathy (arrow). This pattern may be seen with Sjögren disease or HIV-related lymphoepithelial cysts and nodules.
Conclusion

The range of studies needed to assess salivary gland lesions spans a wide gamut. The algorithm for imaging the salivary glands depends on the clinical scenario with which the patient presents to the clinician.

Knowledge of the various utilities that can offer imaging tools in the diagnosis of SS improved the accuracy of the radiologist and the best use of resources moreover the health of the patient.

Personal Information

References

