The Journal of Laryngology & Otology
February 2003, Vol. **117,** pp. 151–152

Bilateral parotid cysts as presentation of Sjögren's syndrome

G. Plaza, M.D., Ph.D., M. P. Domínguez, M.D., Ph.D.[*], A. Bueno, M.D.[†]

Abstract
We present a case of a 40-year woman with bilateral parotid salivary gland enlargement as presentation of primary Sjögren's syndrome. Computed tomography (CT) and magnetic resonance imaging (MRI) showed parotid cysts, suggestive of cystic benign lymphoepithelial lesions. A sub-labial biopsy confirmed the syndrome. After 24-month follow-up, the left parotid cysts remain the same, whereas other cysts have appeared in the right parotid gland. Parotid involvement in Sjögren's syndrome is discussed.

Key words: Salivary Gland Diseases; Sjögren's Syndrome; Parotid Gland

Case report

A 40-year-old woman presented with a six-month history of bilateral parotid enlargement. It was a painless and progressive swelling. She did not refer to any other symptoms. On physical examination a diffuse enlargement of the left parotid gland was noted, with a palpable 2-cm node, whereas the right parotid was also enlarged, without palpable nodes. No other abnormalities were found in the upper respiratory tract. Systemic examination was normal.

A cranial CT scan showed a 18 mm left parotid cyst (Figure 1(a)), while the other parotid gland was also enlarged. Fine needle aspiration of the left parotid gland did not reveal any significant cells. Later, MRI confirmed the presence of multiple cysts in both parotid glands, the bigger (2 cm) within the left parotid, with a high protein content, shown as hyperintense in T2 (Figures 2 and 3).

As Sjögren's syndrome was suspected, an immunological screen was done showing raised immunoglobulins (IgG type, 35.10 g/l), positive antinuclear antibodies (at a 1/320 dilution), as well as high rheumatoid factor (141 UI/ml), and positive anti-Ro (SSA) and anti-La (SSB). The erythrocyte sedimentation rate (ESR) was 75 mm/hour. Chest X-ray was normal. Human immunodeficiency virus (HIV) and hepatitis C virus serology were negative. A sub-labial biopsy confirmed the diagnosis of the syndrome, showing diffuse lymphocyte infiltration of the glands with benign lymphoepithelial lesions. Latent membrane protein-1 immunohistochemistry and *in situ* hybridization for Epstein-Barr virus (EBV)-encoded non-polyadenylated RNAs (EBERs) did not show the presence of EBV within myoepithelial sialadenitis (MESA) islands. Parotid gland biopsy or cyst excision was not considered at that time.

After 24-month follow-up, the parotid cysts have remained the same in the following MRI and the patient has developed only moderate xerostomia. Although the patient is very concerned about the possible development of lymphoproliferative diseases of the parotid gland, we are following a wait and see schedule so far.

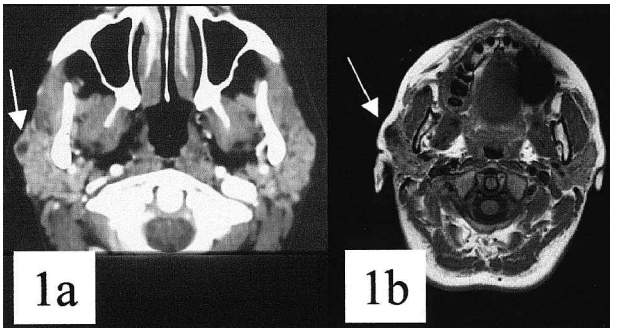

Fig. 1

(a) Axial contrast-enhanced CT scan: Enlarged and dense parotid glands, with a small rounded hypodense cyst in the left parotid gland (arrow), and a very small cyst within the right parotid gland. (b) Axial T1-weighted MR scan (TR 600, TE 11): Small hypointense rounded well-defined lesions within both parotid glands (arrow).

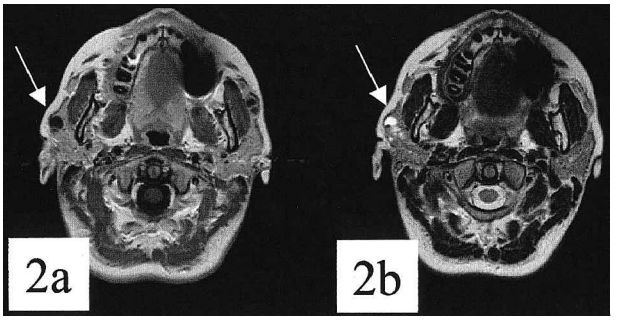

Fig. 2

(a) Axial T1-weighted contrast-enhanced MR scan (TR 600, TE 11): The parotid lesions are not enhanced with gadolinium, and therefore are more defined against the glandular enhancement. (b) Axial T2-weighted MR scan (TR 4400, TE 96): The parotid lesions are hyperintense (as CSF) and homogeneous, suggesting a cystic nature.

From the Departments of Otolaryngology, Pathology[*] and Neuroradiology[†], Fundación Hospital Alcorcón, Madrid, Spain.
Accepted for publication: 1 October 2002.

https://doi.org/10.1258/002221503762624675 Published online by Cambridge University Press

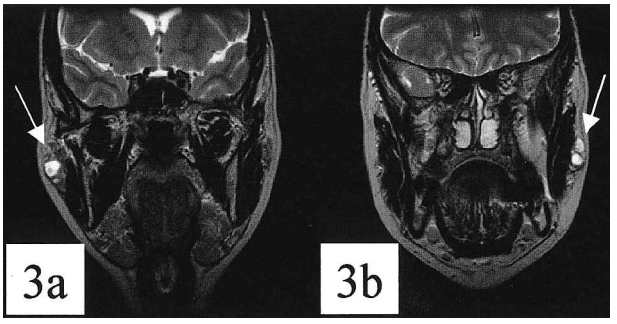

FIG. 3

(a) Coronal T2-weighted MR scan (TR 4400, TE 96): Left parotid hyperintense cysts. (b) Coronal T2-weighted MR scan (TR 4400, TE 96): Right parotid hyperintense cysts.

Discussion

Sjögren's syndrome is an autoimmune disease characterized by inflammation and destruction of exocrine glands, mainly the lacrimal and salivary glands. The syndrome is considered as primary when it appears in the absence of other autoimmune diseases, such as rheumatoid arthritis or systemic lupus erythematosus.[1]

Major salivary gland enlargement occurs in 25–66 per cent of patients with primary Sjögren's syndrome.[1] It may be recurrent and episodic, or chronic and fixed. Benign lymphoepithelial lesions are the most common presentation of such parotid swelling. Although multiple cysts of varying size in the parotid gland have been reported in Sjögren's syndrome, rarely presentation may be as a large parotid cyst.[2] In this case other cystic conditions should be excluded such as cystic Warthin's tumour, human immunodeficiency virus (HIV)-related parotid cyst, branchial cyst or lymphoepithelial cyst on clinical, immunological and histological grounds.[2,3]

CT can aid in the diagnosis of Sjögren's syndrome, define the nature and extent of salivary gland involvement, localize optimal regions for biopsy and detect complications such as lymphoma presentation.[4] MRI shows a multiple mixed hypointense and hyperintense foci (salt and pepper appearance) which can be considered as strongly suggestive of Sjögren's syndrome. Those hyperintense foci in T2-weighted images may correspond with the dilated ductal system (sialectasis) while those hypointense in T2-weighted images may be due to focal lymphocytic aggregation.[5] Our case showed cysts that were hyperintense in T2-weighted images, suggesting the presence of a benign lymphoepithelial cyst in the context of Sjögren's syndrome.

The typical histological finding in Sjögren's syndrome is a massive periductal lymphocytic infiltration of glandular tissue with atrophy or destruction of the glands.[1] Salivary ductal epithelium may proliferate producing epimyoepithelial islands and often glandular atrophy, so called MESA.[6] In current practice minor salivary gland biopsy has found a widespread consensus as an essential histopathologic finding for the diagnosis of Sjögren's syndrome.[6]

The predisposition for the development of non-Hodgkin's lymphoma in patients with Sjögren's syndrome has been clearly established as an over 30 times greater relative risk for the development of lymphoma.[7] Those patients with persistent parotid gland enlargement, persistent fever, splenomegaly and lymphadenopathy, as well as monoclonal cryoglobulinemia are at higher risk. It may develop in about five to 10 per cent of patients with Sjögren's syndrome, more commonly in younger ones. They are usually marginal zone B-cell neoplasms that arise against a background of MESA when monoclonality has emerged.[7]

In patients with Sjögren's syndrome presenting with persistent parotid gland enlargement, a localized lymphoma in the parotid gland must be excluded.[1,7] CT and MRI are useful for determining the cystic nature of the parotid swelling, but should also be periodically performed to show changes in benign lymphoepithelial lesions within the parotid.[8] Furthermore, serial examination of salivary gland tissue to detect monoclonality can also be used in order to prompt recognition of lymphomas. We think that a wait and see policy can also be followed, as only 10 per cent of patients with Sjögren's syndrome will develop lymphoma, and up to 40 per cent will be low-grade lymphoma that can be managed conservatively.[7] Careful clinical and imaging follow-up can avoid parotid scars and allow prompt recognition of lymphomas.

References

1 Manoussakis MN, Moutsopoulos HM. Sjögren's syndrome. *Otolaryngol Clin North Am* 1999;**32**:843–60
2 Ahmad I, Ray J, Cullen RJ, Shortridge RTJ. Bilateral and multicystic major salivary gland disease: a rare presentation of primary Sjögren's syndrome. *J Laryngol Otol* 1998;**112**:1196–8
3 Som PM, Brandwein MS, Silvers A. Nodal inclusion cysts of the parotid gland and parapharyngeal space: a discussion of lymphoepithelial, AIDS-related parotid, and branchial cysts, cystic Warthin's tumors, and cysts in Sjögren's syndrome. *Laryngoscope* 1995;**105**:1122–8
4 March DE, Rao VM, Zwillenberg D. Computed tomography of salivary glands in Sjögren's syndrome. *Arch Otolaryngol Head Neck Surg* 1989;**115**:105–6
5 Niemela RK, Paakko E, Suramo I, Takalo R, Hakala M. Magnetic resonance imaging and magnetic resonance sialography of parotid glands in primary Sjögren's syndrome. *Arthritis Rheum* 2001;**45**:512–8
6 Kondratowicz GM, Smallman LA, Morgan DW. Clinicopathological study of myoepithelial sialadenitis and chronic sialadenitis (sialolithiasis). *J Clin Path* 1988;**41**:403–9
7 García-Carrasco M, Ramos-Casals M, Cervera R, Font J. Síndrome de Sjögren primario y linfoproliferación. *Med Clin (Barc)* 2000;**114**:740–6
8 Sato K, Kawana M, Sato Y, Takahashi S. Malignant lymphoma in the head and neck associated with benign lymphoepithelial lesion of the parotid gland. *Auris Nasus Larynx* 2002;**29**:209–14

Address for correspondence:
Guillermo Plaza, M.D., Ph.D.,
Unidad de Otorrinolaringología
Fundación Hospital Alcorcón,
Av/Budapest 1,
Alcorcón 28922, Madrid,
Spain.

Fax: 34 91 6219409
E-mail: gplaza@fhalcorcon.es and gmayor@ctv.es

G. Plaza, M.D., Ph.D. takes responsibility for the integrity of the content of the paper.
Competing interests: None declared