

Short Case Report

Sjögren syndrome hidden by previous parotidectomy and cervicofacial radiotherapy

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Abstract – Introduction: Sjögren's syndrome (SS) is a chronic inflammatory auto-immune disease of the exocrine glands whose incidence increases with age. Sex ratio is 9.9 female for 1 male. **Observation:** A 67-year-old female patient presented for xerostomia leading to major functional impairment. The medical history of the patient was pulmonary sarcoidosis, with joint and adenoid involvement, considered as cured; left partial parotidectomy, with a diagnosis of acinous carcinoma, followed 7 years later by a total parotidectomy due to tumor recurrence, and followed by external radiotherapy (70 Gy). Arterial hypertension, hypothyroiditis and diabetes mellitus were also noticed. She reported xerostomia and xerophthalmia. Seric anti-SSA antibodies were positive and histologic findings were compatible with SS. **Commentaries:** Despite other co-morbidities, the diagnosis was hidden by previous parotidectomy and cervical radiotherapy. Thus, SS should not be underdiagnosed in patients with complex medical history.

Case report

A 67-year-old female patient presented for xerostomia leading to major functional impairment. The medical history of the patient was pulmonary sarcoidosis, with joint and adenoid involvement, considered as cured; left partial parotidectomy, with a diagnosis of acinous carcinoma, followed 7 years later by a total parotidectomy due to tumor recurrence, and followed by external radiotherapy (70 Gy). Arterial hypertension, hypothyroiditis and diabetes mellitus were also noticed.

The patient described asthenia due to alterations of sleep because of dry mouth and phlegm. She said that xerostomia was increased since 1 year. There was no joint pain. Xerophthalmia was also noticed. Seric anti-SSA and anti-SSB antibodies were investigated and anti-SSA antibodies were positive (over 8). A salivary gland biopsy was performed and revealed a preservation of the parenchyme's architecture and a lymphoplasmocystic infiltrate. The Chilsom and Mason score was 4. Histologic findings were compatible with Sjögren's syndrome (SS).

A complete examination was performed, which confirmed the diagnosis of primary SS. There was no other location of the syndrome. A symptomatic treatment for xerostomia

was instated, consisting in pilocarpine chlorhydrate (10 mg, 3 times a day). Artificial tears were prescribed for eye dryness.

Discussion

SS is a chronic inflammatory auto-immune disease of the exocrine glands that concerns 0.05–4.8% of adults. Its incidence increases with age with a peak between 55 and 64 years. Sex ratio is 9.9 female for 1 male [1]. The diagnosis is made on ocular dryness, mouth dryness, histologic features with focal lymphocytic sialadenitis, salivary alterations on scintigraphy and specific auto-antibodies (anti-SSA, anti-SSB). The association of SS with diabetes, thyroiditis, or lymphoproliferative diseases are well described [1].

Previous head and neck radiotherapy or parotidectomy may interfere with the clinical presentation and complicate the diagnosis [2]. This case report aims to highlight that the clinician must not eliminate the diagnosis of SS despite other risk factors for hyposalivation.

In her medical history, the patient suffered from different diseases that are more frequently described in SS patients than in the general population. Nonetheless, previous parotidectomy and radiotherapy on the parotid zone render the diagnosis more difficult. Head and neck radiotherapy causes

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hyposalivation for radiation doses over 40 Gy. The patient received 70 Gy on the left parotid, therefore, saliva secretion was ensured by the right parotid, sublingual glands and accessory glands until SS appeared. The patient described previous hyposalivation as “moderate”, and “severe” the last year.

Sarcoidosis is a multiorgan granulomatous disease and is currently associated with SS. Some authors [3] consider that it should be the main differential diagnosis with SS in case of pulmonary involvement. The salivary gland biopsy is a reliable way to differentiate the two diseases. Many cases of secondary SS are described in sarcoidosis patient [3].

The incidence of diabetes mellitus in SS is controversial: in some studies, patients with SS present twice more often with diabetes, whereas other studies do not consider any relation between these diseases.

Many authors have described a higher incidence of SS in patients presenting with auto-immune thyroiditis or Hashimoto’s disease or a thyroid dysfunction [1].

Alterations of vascular endothelium in SS may lead to infraclinical atherosclerosis and secondary hypertension.

Long-term secondary diseases are mainly represented by the 18.8 times higher risk (IC: 95%) of developing non-Hodgkinian lymphoma, like MALT lymphoma. These lymphomas occur mostly in 55–60 female patients, with a time interval between SS and lymphoma ranging from 6 to 12 years [4]. Previous radiotherapy seems to increase this risk. Thus, a close follow up has been instituted in this patient.

The treatment proposed for this patient was only symptomatic, due to the only ocular and salivary involvement of the SS. If systemic alterations should be diagnosed, rituximab could be proposed, but results seem to be inconstant [5].

Close follow-up is necessary to ensure early diagnosis of possible lymphoma.

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