Case Report

Facial tumor as a presenting complaint in a patient with primary Sjögren's syndrome

ABSTRACT

Sjögren's syndrome is a chronic autoimmune disorder of the exocrine glands with associated lymphocytic infiltrates of the affected glands. Dryness of the mouth and eyes results from involvement of the salivary and lacrimal glands. Up to one-half of affected individuals also develop extraglandular involvement in organs distinct from the salivary and lacrimal glands, including the joints, skin, lung, gastrointestinal tract, nervous system, and kidneys. The disease also occurs in conjunction with other autoimmune disorders, such as systemic lupus erythematosus and rheumatoid arthritis. We report a case of a 76-year-old woman who presented to our department with a swelling on the left cheek. Investigations revealed Sjögren's syndrome as the underlying cause of the facial tumor.

Keywords: Facial tumor, fine-needle aspiration, Sjögren's syndrome, ultrasound

INTRODUCTION

Primary Sjögren's syndrome is a systemic autoimmune disease, characterized by lymphocytic infiltration of the secretory glands. This process leads to sicca syndrome, which is the combination of dryness of the eyes, oral cavity, pharynx, larynx, and/or vagina.[1] Sicca syndrome is often accompanied by extraglandular manifestations, including cutaneous, musculoskeletal, pulmonary, renal, hematological, and neurological involvement. The pathogenesis of primary Sjögren's syndrome is currently not well-understood, but increased activation of B-cells followed by immune complex formation and autoantibody production is thought to play important roles.[2,3] The incidence of primary Sjögren's syndrome has been estimated to be 4/1000 patients per year, with an overall prevalence in Europe between 0.1% and 4.8%.[4,5] In a systematic review and meta-analysis of the epidemiology of primary Sjögren's syndrome, the pooled incidence rate was 6.92 (95% confidence interval [CI]: 4.98–8.86)/100,000 person-year, and the overall prevalence was 60.82 (95% CI: 43.69–77.94) cases/100,000 inhabitants, although incidence and prevalence rates vary widely around the world.[6] Furthermore, as many symptoms are nonspecific, the prevalence may be underestimated.

Primary Sjögren's syndrome is diagnosed using the American–European Consensus Group classification criteria,[7] which include the presence of ocular symptoms, oral symptoms, ocular signs (positive result of the Schirmer's test or Rose Bengal test), consistent histopathology, objective evidence of salivary gland involvement, and presence of autoantibodies to Ro (SSA) or La (SSB). There is no standard clinical presentation for primary Sjögren's syndrome, as many patients have various degree of systemic involvement at the time of disease onset. Symptoms are usually divided into sicca syndrome, general manifestations, and extraglandular manifestations. Classical symptoms of the sicca syndrome,

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such as xerophthalmia, xerostomia, pharynx and/or larynx dryness, nonproductive cough, dyspareunia, or skin dryness, occur in >95% of patients.[2,8] Fatigue is the most prevalent general symptom occurring in up to 70%–80% of patients with primary Sjögren's syndrome.[9] Extraglandular manifestations have been reported in approximately 71% of patients[10] including lymphoma, arthritis, interstitial lung disease, tubulointerstitial nephritis, and neurological symptoms, which frequently precede the diagnosis of Sjögren's syndrome.[11]

Here, we present a rare case of primary Sjögren's syndrome in an older woman presenting with a facial mass as the first symptom of the disease. As far as we are aware, no other similar cases have been previously reported in the literature.

CASE REPORT

A 76-year-old woman reported to the Outpatient Department of Oral and Maxillofacial Surgery of A Coruña University Hospital, with a chief complaint of painless swelling on the left cheek for the past 6 months. The swelling was initially smaller in size and had gradually increased to its present state. Her medical history included allergy to acetylsalicylic acid, arthrosis, osteoporosis, dyslipidemia, hypothyroidism, and high blood pressure controlled with antihypertensive medication. There was no history of any trauma or tooth extraction.

The clinical examination revealed a mass about 1 cm × 1 cm on the left cheek and anterior to the masseter muscle. The swelling was nontender and nonfluctuant and had well-defined margins. The overlying skin was not attached to the lesion and was freely movable. Intraoral examination was unrevealing. A fine-needle aspiration (FNA) was performed, the result of which was compatible with a diagnosis of lipoma [Figure 1]. Computed tomography (CT) scan revealed a partitioned hypodense mass in the subcutaneous cellular tissue of approximately 8.5 mm in diameter and adjacent to the left malar bone [Figure 2]. Density was not compatible with fat, but a tentative diagnosis of a lymph node enlargement or a tumor was made. An ultrasound-guided FNA biopsy showed mononuclear cells compatible with lymphoid tissue, without evidence of malignant or epithelial cells. At follow-up, the lesion increased in size, and a second ultrasound-guided FNA confirmed the diagnosis of lymphadenopathy. There were multiple anechoic and hypoechoic areas in the parotid gland corresponding to duct dilatation or small areas of inflammation [Figure 3]. Both submaxillary glands showed a heterogeneous structure [Figure 4]. These findings were compatible with Sjögren's syndrome. A minor salivary gland was biopsied under local anesthesia. Histopathologic examination disclosed nonspecific lymphadenitis without evidence of malignant cells together with acinar atrophy with fibrosis and foci of lymphoid cells clustered around

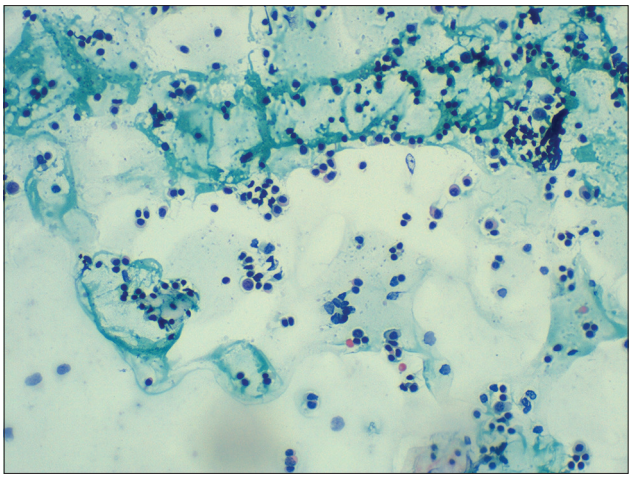

Figure 1: Nondiagnostic cytology: lymphocytes and plasma cells

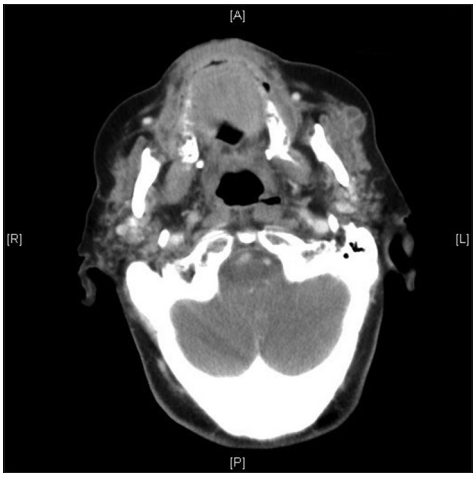

Figure 2: Computed tomography: Partitioned hypodense node in the subcutaneous cellular tissue, adjacent to the left malar bone, of 8.5 mm

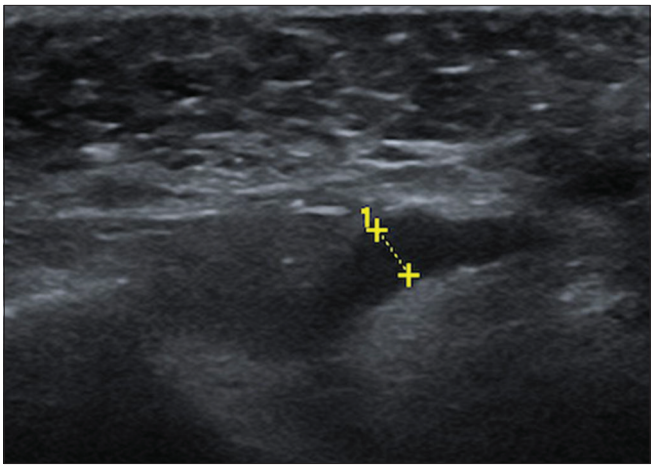

Figure 3: Ultrasound scan: Parotid duct dilatation

the ducts [Figure 5]. The magnetic resonance imaging (MRI) scan disclosed a heterogeneous nodule, 17 mm × 13.7 mm, compatible with cystic degeneration and necrosis [Figure 6]. The left parotid gland was decreased in size too. Based on all these findings, the patient was diagnosed with primary Sjögren's syndrome and was referred to the Department of Rheumatology of our institution for further workup studies, treatment, and follow-up.

DISCUSSION

We report an unusual case of facial mass as a presenting clinical sign of Sjögren's syndrome in a previously healthy woman without symptoms of the sicca syndrome and in the absence of general or systemic manifestations of the disease. The diagnosis was established by suggestive findings of lymphadenopathy in the FNA of the nodule and typical images of lymphocytic infiltration surrounding the excretory ducts in combination with destruction of the acinar tissue in the biopsy of a minor salivary gland.[12] Different studies have shown good performance of biopsy of the minor salivary glands for the diagnosis of primary Sjögren's syndrome.[13,14] Furthermore, salivary gland ultrasound is a useful method in visualizing glandular structural changes as a first-line imaging tool in the diagnosis of the disease.[15] It has been suggested that evaluation of salivary gland involvement by means of ultrasonography can replace other diagnostic techniques, such as sialography or salivary scintigraphy.[16] CT of the parotid gland has also been shown to be accurate and reliable in the diagnosis of Sjögren's syndrome, with heterogeneity, abnormal diffuse fat tissue deposition, and diffuse punctate calcification as specific findings for the disease.[17] Recently, the heterogeneous signal intensity distribution on T1- and T2-weighted images; the multiple high-signal-intensity spots on magnetic resonance sialograms; and the reduced volume of the parotid, submandibular, and sublingual salivary glands have been considered good diagnostic MRI indicators for Sjögren's syndrome.[18]

Once the diagnosis of primary Sjögren's syndrome was established in our patient, she was referred to the Department of Rheumatology for serological studies, treatment, and follow-up. Treatment is mainly symptomatic for the relief of xerophthalmia and xerostomia. Systemic treatment is indicated when general symptoms cannot be managed with local treatment or adjustment of the patient's lifestyle and in case of organ involvement, including renal disease, interstitial pneumonitis, peripheral neuropathy, or systemic vasculitis. Therapy-resistant primary Sjögren's syndrome with proven organ damage is an indication to start biologicals, with the B-cell as the most promising target based on the etiopathogenesis of the disease.[2] Patients with primary Sjögren's syndrome should be closely monitored to assess the effect of treatment and the development or progression

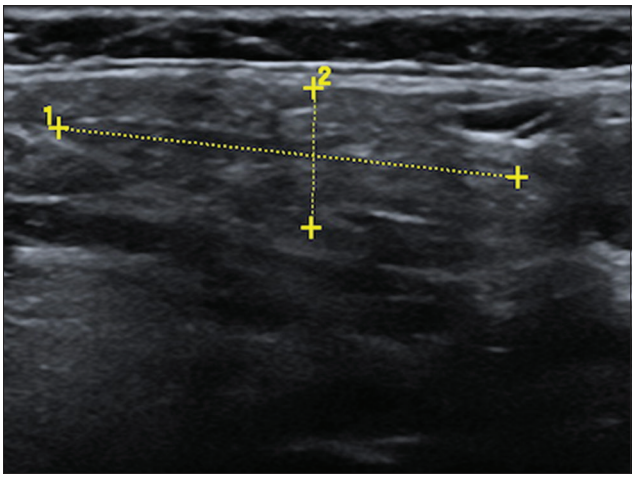

Figure 4: Heterogeneous structure of both the submaxillary glands

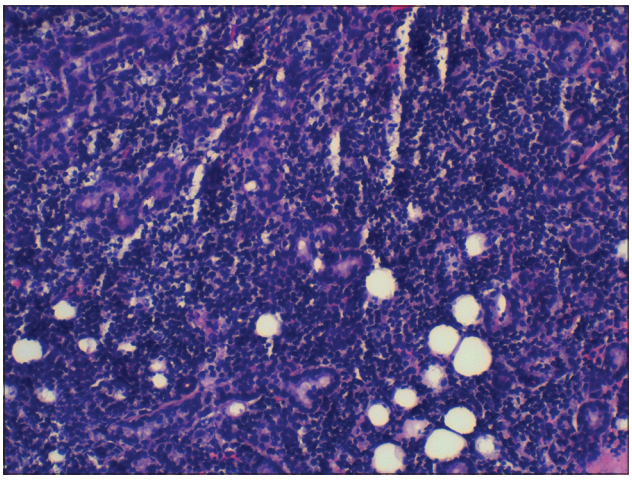

Figure 5: Nonspecific lymphadenitis without evidence of malignant cells. Acinar atrophy with fibrosis and lymphoid accumulates

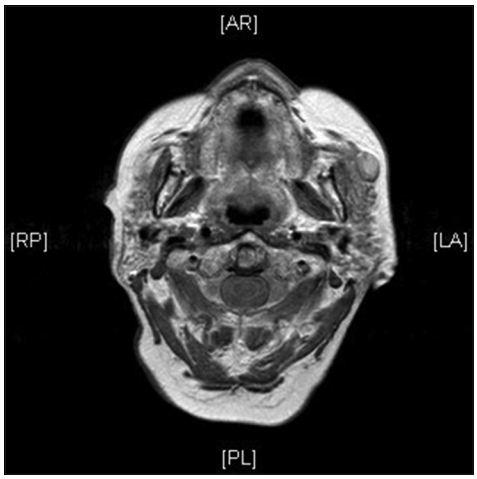

Figure 6: Magnetic resonance: heterogeneous node measuring 17 mm × 13.7 mm compatible with cystic degeneration or necrosis

of systemic manifestations, including an increased risk for lymphoma and lymphoproliferative disorders.[19]

CONCLUSION

The presence of a facial tumor, in the present case as a painless nodule on the subcutaneous cellular tissue of the cheek, can be added to the list of very unusual initial clinical manifestations of primary Sjögren's syndrome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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