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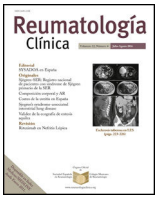

Letters to the Editor

Sjögren's Syndrome and Halitosis: A Case Report☆

Síndrome de Sjögren y halitosis: descripción de un caso clínico

To the Editor,

Halitosis is a common reason for consulting a physician that entails a broad differential diagnosis, as it can be a manifestation of extraoral or systemic diseases, like the case of Sjögren's syndrome (SS) that we present here.

The patient was a 36-year-old woman, with nothing remarkable in her medical or surgical history, who was referred from primary care with a 1-year history of persistent halitosis. She had been examined by dentists and had her teeth cleaned several times, but there was no evidence of oral disease to explain her condition. She had been studied in the ear, nose and throat and the gastroenterology departments, and had undergone computed tomography of the sinuses, rhinoscopy, upper gastrointestinal series, breath test and laboratory analyses, none of which had revealed signs of disease. She had tried mouthwashes, proton pump inhibitors, prokinetic agents and over-the-counter products, but the halitosis persisted, limiting her quality of life. In internal medicine, she reported having halitosis every day. It improved on eating and with chewing gum. She needed to drink water constantly, even at night, and had a feeling of dry mouth and frequent ocular discomfort, with pruritus, that she attributed to her job and stress. She had no complaints associated with organs or apparatuses. Physical examination only revealed evident halitosis when she exhaled through the mouth and dry tongue; the rest of the oral cavity was normal. The results of laboratory analyses with a complete blood count, tests for hemostasis, and kidney and liver function tests were normal. Indirect immunofluorescence only revealed an antinuclear antibody titer of 1/160 with a homogeneous pattern and the presence of anti-Ro. Chest radiography and abdominal ultrasound were normal (Table 1, *available in online supplementary material*). The Schirmer test in both eyes resulted in moisture of less than 5 mm in 5 min, and the score with rose Bengal staining was 4 points. As the patient refused to undergo salivary gland biopsy, gamma scintigraphy of parotid glands was performed. It revealed a grade II/IV bilateral diffuse uptake deficit. On the basis of the results obtained, a diagnosis of halitosis secondary to xerostomia associated with SS was established. The recommendations were that she eliminate caffeine consumption and use sugarless mint or lemon-flavored candy and alcohol-free mouthwashes. Frequent hydration was also indicated, as was the use of artificial tears and saliva. As the patient perceived a partial improvement in dry mouth and halitosis, treatment with pilocarpine was initiated, with incremental doses up to 5 mg/6 h, at which point, satisfactory symptom control was achieved. Progressive improvement in dry mouth and halitosis were observed in office visits and there have been no complications during follow-up.

Table 1
Differential Diagnosis of True Halitosis.

A) Physiologic halitosis (nonpathological oral factors)
B) Pathologic halitosis
Oral causes
Periodontal disease, stomatitis, pharyngitis, parotid gland dysfunction and neoplasms
Extraoral dysfunction
Perioral causes: nasal, paranasal, laryngeal (sinusitis, atrophic rhinitis, foreign bodies, malformations, epiglottitis)
Gastrointestinal diseases: Zenker's diverticulum, dyspepsia, gastroesophageal reflux, *Helicobacter pylori* infection, biliary disease and neoplasms
Respiratory diseases: pulmonary infections, bronchiectasis, abscess, tuberculosis and neoplasms
Neurological diseases: neurodegenerative diseases, epilepsy and neoplasms
Systemic diseases: diabetes mellitus, renal or hepatic failure, dehydration, poisoning, drugs, autoimmune diseases (Sjögren's syndrome, rheumatoid arthritis, systemic lupus erythematosus, scleroderma) and carcinomas

Halitosis is defined as an unpleasant odor in the exhaled breath, which can result in an important social problem, and may be the consequence mainly of dental disease.[1] In some cases, it is associated with extraoral disease (ear, nose and throat, gastrointestinal, hepatic, neurological, respiratory or systemic), which may require specific treatment and follow-up.[2] Sjögren's syndrome is a chronic autoimmune disease in which there is a lymphocytic inflammatory infiltrate in the exocrine glands and in certain extraglandular tissues. It causes a progressive destruction of the latter, producing xerostomia and xerophthalmia, among other symptoms.[3] Saliva is composed of water, electrolytes, proteins, glycoproteins, defensins, proteases, histatins and lysozymes, as well as other molecules with biological and biochemical properties that are essential in the maintenance of the oral physiology.

Xerostomia is among the classification criteria of the disease, and is the complaint most widely reported by patients, among other oral problems caused by the reduced salivary flow. Our patient met 5 of the 6 American-European criteria established in 2002.[4,5] The microbiological composition of the saliva plays an essential role since, in patients with reduced salivary flow, as in SS, there is a modification in the bacterial flora. This circumstance is related to an increase in the concentration of microorganisms like *Lactobacillus acidophilus*, *Streptococcus mutans* and *Candida albicans*, among others, which favors the development of caries, infections (candidiasis), burning mouth syndrome, glossodynia, dysphagia, halitosis and oral lesions.[6,7] Halitosis is a result of this reduced salivary flow. However, it is rarely mentioned as a major manifestation leading to the perception and suspicion of a diagnosis of SS, as occurred in our patient.[6] The treatment consists of multiple hygienic and

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dietary measures that favor oral hydration, the use of artificial saliva and, in the most severe cases, systemic therapy, for example, with cholinergic agents.[8,9]

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Case of familial mediterranean fever presenting with constant abdominal pain

Caso de fiebre mediterránea familiar con dolor abdominal constante

Dear Editor,

Familial Mediterranean fever (FMF) is a disease characterized by sporadic, serosal inflammation and unpredictable attacks of fever. This condition is thought to be hereditary and autosomal recessive. Patients often consult with fever, joint pain and intermittent abdominal pain, which progresses as an attack that does not last more than 3 days.[1] We discuss a case, rarely reported in the literature, in which the presenting symptom was continuous abdominal pain. An extensive study led to a diagnosis of FMF.

A 49-year-old man of Turkish origin came to our outpatient clinic with isolated, persistent abdominal pain. He had a 10-year history of abdominal pain in the form of continuous shooting pain in left upper quadrant. The attacks of abdominal pain were not very severe, but would last all day and, over the past 5 years, he had noted that the severity did not change upon eating or drinking. The patient had undergone examination with all the advanced radiological techniques, including exploratory laparoscopy focusing on possible sources of the abdominal pain, but no diagnosis had been reached.

The patient's vital signs on admission included a body temperature of 36.3 °C, pulse rate of 85 bpm, blood pressure of 140/90 mm Hg and respiratory rate of 13 breaths/min. There were no notable abdominal findings in the physical examination except tenderness on deep palpation in the left upper quadrant. Tests performed to determine the etiology included complete blood count, routine biochemistry, markers of hepatitis, urinalysis, stool microscopy and culture, thyroid function tests, anti-extractable nuclear antigen antibody profile, culture for *Salmonella* and *Brucella*, and tumor markers, and the results were normal or negative. Erythrocyte sedimentation rate was 25 mm/h (normal range, 0–20) and C-reactive protein level was 9 mg/L (0.2–5). In radiological examinations using advanced techniques, the findings in abdominal ultrasonography and computed tomography, esophagogastroscopy and colonoscopy were normal. Although the patient's clinical presentation was not suggestive of FMF, genetic testing was carried out with this disorder in mind. As a result, a homozygous R202Q mutation was detected. A Tru-cut biopsy taken from the rectum during the colonoscopy revealed AA amyloidosis. The patient was diagnosed with FMF on the basis of abdominal pain, the positive genetic test result and AA amyloidosis. The patient was started on colchicine 3 times daily. After 3 weeks of treatment, the patient's abdominal pain had completely resolved.

In FMF, patients often present with peritonitis, pleurisy, synovitis and skin lesions such as erysipelas. However, approximately 95% of the patients complain of localized abdominal pain. The pain, local at first, progresses to rigidity, adynamic ileus and rebound tenderness, and ultimately spreads to the whole abdomen. The attacks often last up to 3 days.[2] Familial Mediterranean fever is caused by a *MEFV* gene mutation,[3] which often occurs in exon 2 or 10. While the prevalent mutation (47–94%) is M694V in exon 10, previous genetic studies have shown that M680, E148Q, V726A, A744S, R202Q, R761H and T267 are also frequent mutations.[4] R202Q is a mutation that can be detected quite often in the Turkish population. In studies carried out in Turkey, it has been shown that heterozygous forms produce no symptoms and do not cause amyloidosis, but homozygous forms are associated with the development of symptoms and progression to amyloidosis.

Our patient was admitted to the hospital with a 10-year history of persistent isolated left upper quadrant pain. A homozygous R202Q mutation was detected in the genetic analysis and rectal biopsy revealed AA amyloidosis. The patient responded well to treatment with colchicine.

Thus, FMF should be considered in patients presenting with abdominal pain that is not characteristic of this disorder.

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Conflict of interest

The authors declare that they have no conflict of interest.

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