



Parotitis as the initial sign of juvenile Sjögren's syndrome

Catherine M. Flaitz, DDS, MS

Dr. Flaitz is professor, Oral and Maxillofacial Pathology and Pediatric Dentistry, Department of Stomatology, University of Texas at Houston Health Science Center Dental Branch. Correspond with Dr. Flaitz at cflaitz@mail.db.uth.tmc.edu

Abstract

Parotid swelling may be associated with a variety of glandular disorders in children. This case report describes the characteristic features of juvenile Sjögren's syndrome in an adolescent girl who presented with recurrent and bilateral parotid gland enlargement. Special emphasis is placed on an age-specific differential diagnosis for major salivary gland enlargements. (Pediatr Dent 23:140-142, 2001)

Sjögren's syndrome is a progressive autoimmune disorder of the exocrine glands, which affects primarily the salivary and lacrimal glands, resulting in xerostomia and xerophthalmia. Although Sjögren's syndrome is one of the more common rheumatic diseases in adult women, it is diagnosed infrequently during childhood. The disease spectrum is quite divergent, ranging from an organ-specific endocrinopathy to widespread systemic involvement that may result in musculoskeletal, pulmonary, gastric, hematological, vascular, dermatological, renal and neurological disorders.^{1,2} Sjögren's syndrome may occur alone and is referred to as the primary type or it may be associated with other autoimmune diseases and is known as the secondary type. The cause of this disease is unknown; however, there appears to be a genetic predisposition. Because the disease presentation is so variable and often nonspecific, the diagnosis of Sjögren's syndrome is often delayed for many years. The purpose of this case report is to describe the clinical findings of secondary Sjögren's syndrome in an adolescent girl. The classic head and neck manifestations will be discussed along with an age-appropriate differential diagnosis.

A 14 year-old African-American girl presented with bilateral parotid swelling and generalized tooth sensitivity, especially when drinking cold carbonated beverages (Fig 1). For the past 3 years, she has experienced an increase in dental problems and periodic enlargement of both the parotid and submandibular glands. Previous episodes of the facial swellings seemed to coincide with the occurrence of a variety of head and neck problems, including an odontogenic infection, chronic otitis media, chronic sinusitis and an upper respiratory infection. The patient had taken antibiotics for the treatment of these infections, which had not been beneficial in managing the swollen glands. Recently she complained of fatigue, fever, and joint pain, especially affecting the knees. Although the teenager did not complain about oral dryness, chronic gingivitis, generalized staining of the teeth with cervical enamel decalcification and numerous amalgam restorations with recurrent carious le-



Fig 1. Bilateral parotid and submandibular swelling and chapped lips.

sions were documented (Fig 2). In addition the lips were dry and chapped. Although adequate in quantity (1.1 ml/min for whole stimulated saliva), the quality of the saliva was stringy and bubbly. Other head and neck findings included a recent nose piercing, which had healed uneventfully except for an increase in nasal crusting in the decorated nares.

Clinical impression

Based on the episodic history of nontender enlargement of the parotid and submandibular glands, along with the high dental caries susceptibility, this teenager was referred for evaluation of juvenile Sjögren's syndrome (JSS). Although this disease is uncommon in children, it should be an important consideration when recurrent parotid swelling is observed. JSS has a marked predilection for females with the mean age of onset ranging from 7 to 14 years. In contrast to adults, children are less likely to develop components of the sicca complex (xerostomia or xerophthalmia). The most common extraglandular or systemic manifestation of this disease in children is leukopenia. Other manifestations observed in both children and adults include arthritis, arthralgia, purpura, interstitial lung disease, renal tubular acidosis, splenomegaly, gastrointestinal disease, and frequent upper airway infections.^{1,2}

Serologic studies are abnormal in children who have JSS and aid in the diagnosis. The most frequent findings include a positive rheumatoid factor (RF), antinuclear antibodies (ANA), anti-Ro (SSA) or anti-La (SSB) antibodies, elevated erythrocyte sedimentation rate (ESR), and hypergammaglobulinemia. Anemia, leukopenia and thrombocytopenia are additional hematological findings diagnosed in children.^{1,2}

Received December 12, 2000 Revision December 20, 2000



Fig 2. Chronic gingivitis and generalized staining of the teeth with cervical decalcification.

When JSS is found in association with autoimmune disorders, the most common diagnosis is rheumatoid arthritis, followed by systemic lupus erythematosus and mixed connective tissue disease. In most cases JSS precedes these other autoimmune diseases by several years.¹

Although sicca syndrome is uncommon in children as the presenting sign of the JSS, it often manifests later in the course of the disease. The most prominent symptom is dry mouth accompanied by painful, burning mucosa, sensitivity to spicy foods, taste aberrations and halitosis. These patients often develop widespread dental caries, difficulty swallowing and chewing foods, problems with speaking for long periods of time and an increased risk for candidal infection. In addition, the major and minor salivary glands may be enlarged and the saliva appears thick, sticky and foamy.³

Diagnosis

Following an extensive rheumatological evaluation this patient was diagnosed with juvenile rheumatoid arthritis and secondary SS. In order to help confirm the diagnosis of JSS, a labial lip biopsy was performed. Microscopic examination of the lobules of minor salivary glands revealed multiple aggregates of lymphocytes and plasma cells (≥ 50 or more chronic inflammatory cells) with destruction of the acinar structures. Although not obtained in this patient, salivary gland imaging, including sialography, magnetic resonance imaging and scintigraphy are valuable tools for providing functional and anatomical information.³

Treatment and prognosis

This autoimmune disease was treated with low-dose systemic corticosteroids for the management of the parotitis and the arthritis in this patient. In addition, a comprehensive program for prevention of caries and periodontal disease was instituted. Because the patient did not complain about a dry mouth and the measurements of the whole stimulated saliva were within the normal range, salivary substitutes and systemic sialogogues were not recommended at the present time. Besides oral health concerns, this patient is at increased risk for developing non-Hodgkin's lymphoma. Patients at greatest risk for developing lymphoma are those with persistent salivary gland enlargement, lymphadenopathy and peripheral neuropathy. Recently a chromosomal translocation, t(14;18), has been detected in the lymphocytes from the peripheral blood and bone marrow in adults with SS, which is a marker for this malignancy.⁴

Differential diagnosis

Enlargement of the parotid glands may be seen in a number of pediatric diseases, including inflammatory, infectious and idiopathic conditions. Another uncommon disease of the parotid gland that must be differentiated from JSS is juvenile recurrent parotitis (JRP).⁴ This later condition is characterized by multiple episodes of painful parotid swelling that is usually unilateral and typically lasts for several days. There is a male predisposition with the age of onset usually between 3 and 6 years of age. Typically this disease undergoes remission by puberty, although it may persist into adulthood. A variety of etiologic factors have been associated with this disease including allergies, upper respiratory infection, and congenital and anatomical defects of the salivary gland. The major salivary gland swelling is of acute onset and may be accompanied by fever and general malaise. Xerostomia is a complaint during the acute attack and a mucopurulent discharge can be milked from the parotid duct. The treatment of this disease is conservative management since most cases spontaneously resolve by puberty. The use of sialogogues, massaging the gland, increased fluid intake, oral antibiotics and analgesics have been recommended to promote resolution of the swelling. Surgery is reserved for severe cases that persist into adulthood. Diagnosis is made by ultrasound and supplemented by sialography, which also may be beneficial in the management of this disease. In contrast to JSS, there is an absence of detectable autoantibodies and there is no lymphocytic infiltration within the salivary gland tissue.

Bilateral parotid swelling may be a sign of an adolescent with an eating disorder. This condition is referred to as sialadenosis, a noninflammatory disorder of the major salivary glands that is associated with an underlying systemic condition. The increase in the gland size is gradual and is due to the hypertrophy of the acinar cells. Other head and neck manifestations of eating disorders include dental erosions, chronic ulcers in the soft palate and tonsillar pillar region, palatal petechiae, fissures involving the commissures of the mouth, dry skin and limpness of the scalp hair.⁵ Management of the salivary gland disease is difficult because it involves controlling the underlying eating disorder.

Infectious causes for parotitis in children include mumps, streptococcal and staphylococcal infections, human immunodeficiency virus (HIV), Epstein-Barr virus, cytomegalovirus, parainfluenza virus, influenza virus and enterovirus. Of these infectious diseases, it is diffuse infiltrative lymphocytosis syndrome (DILS) associated with HIV disease that is indistinguishable clinically from JSS.⁶ Besides a positive history of HIV infection, these children are likely to have concurrent pulmonary involvement and lack of autoantibodies. However, similar to JSS these immunocompromised children are at increased risk for developing non-Hodgkin's lymphoma.

Rarely, local trauma to the buccal mucosa, including the papilla overlying Stensen's duct may be responsible for tender parotid enlargement. Localized edema or ulceration at this site, such as recurrent aphthous ulcers or irritation from orthodontic appliances, may obstruct the salivary flow and cause a mild facial swelling. Typically, the enlargement is unilateral and resolves shortly after the cause is identified.

References

1. Anaya J-M, Ogawa N, Talal N: Sjögren's syndrome in childhood. *J Rheumatol* 22:1152-8, 1995.
2. Bartunkova J, Sediva A, Vencovsky J, Tesar V: Primary Sjögren's syndrome in children and adolescents: proposal for diagnostic criteria. *Clin Experiment Rheumatol* 17:381-6, 1999.
3. Fox PC, Brennan M, Pillemer S, Radfar L, Yamano S, Baum BJ: Sjögren's syndrome: a model for dental care in the 21st century. *JADA* 129:719-28, 1998.
4. Takacs I, Zeher M, Urban L, Bachmann M, Szegedi G, Semsei I: Frequency and evaluation of t(14;18) translocation in Sjögren's syndrome. *Ann Hematol* 79:444-8, 2000.
5. Chirte VV, Premchandra DJ: Recurrent parotitis. *Arch Dis Child* 77:359-63, 1997.
6. Schulze UM, Pettke-Rank CV, Kreienkamp M, Hamm H, Brocker EB, Wewetzer C, et al.: Dermatologic findings in anorexia and bulimia nervosa of childhood and adolescence. *Pediatr Dermatol* 16:90-4, 1999.
7. Chetty R, Vaithilingum M, Thejpal R: Epstein-Barr virus status and the histopathological changes of the parotid gland lymphoid infiltrates in HIV-positive children. *Pathol* 31:413-7, 1999.

ABSTRACT OF THE SCIENTIFIC LITERATURE



CONTINUITY OF MEDICAL CARE AND TIMELY VACCINATION

Poor access to primary medical care is a known barrier to timely achievement of up-to-date immunization status for children. This study looks beyond having a usual source of care in order to evaluate the impact of continuity of medical care with a specific primary care provider on the achievement of up-to-date status for measles-mumps-rubella vaccination at 15 months of age. Study subjects consisted of children born between January 1, 1993 and August 31, 1997 and enrolled continuously in a large health maintenance organization during the first 15 months of life. Continuity of care was based on an established index that measures the degree to which children were treated by their assigned primary care provider. The authors evaluated the effect of greater continuity of care on the likelihood of a child receiving his/her first MMR vaccination by age 15 months. After adjusting for factors such as gender, year of birth and Medicaid status, it was found that children with high continuity of care were 8% more likely to have up-to-date MMR status than were children with low continuity of care. When Medicaid-enrolled children were evaluated separately, high continuity of care was associated with a 22% increase in up-to-date MMR status. The authors point out that these differences in timely immunization status were apparent even within a single health system with relatively uniform access to and quality of care as well as good baseline rates of immunization. While noting that this observational study cannot prove a cause-effect relationship, the authors speculate that increased continuity of care may enhance provider-patient rapport and thus improve the acceptance of immunizations and/or increase the number of well-child visits.

Comments: This study indicates that evaluating access to medical care solely in terms of patients having a usual source of care (i.e., a medical home) may not be specific enough to help us understand utilization of preventive services. Helping children and families establish an ongoing relationship with a primary care provider seems to be important. **RDV**

Address correspondence to: Dimitri A. Christakis, MD, MPH, Department of Pediatrics, Child Health Institute, University of Washington, 146 N Canal St., Suite 300, Seattle, WA 98103-8652

The Association Between Greater Continuity of Care and Timely Measles-Mumps-Rubella Vaccination. Christakis DF, Mell L, Wright JA, Davis R, and Connell FA. *Am J Public Health* 90:962-965, 2000.

30 references