Parotitis as the initial sign of juvenile Sjögren’s syndrome

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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. 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 lesions 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.

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.
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.

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.

**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 salivary gland 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.

**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 of minor salivary gland swelling 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 limppness of the scalp hair.

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. 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

CONTINUITY OF MEDICAL CARE AND TIMELY VACCINATION

Abstract of the Scientific Literature

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

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