Paediatric Sjögren’s syndrome with bilateral parotid cysts: A case report.

Jamila Skinner, BSc, James Fowler, MD1, Jonathan Park MD FRCPSC2, Peng You, MD FRCSC1
1Department of Otolaryngology – Head and Neck Surgery, Western University, London, Ontario, Canada; 2Department of Pediatric Rheumatology, Western University

Abstract

Introduction:
Sjögren’s syndrome is an autoimmune disease characterized by the destruction of exocrine glands. Clinically, this results in the loss of tear and saliva production. Although xerophthalmia and xerostomia, also known as sicca, is a common presentation among adults, pediatric patients more often present with recurrent parotitis and glandular enlargement. Overall symptoms can vary, making initial diagnosis challenging. Approximately 80% of patients with Sjögren’s syndrome experience parotid gland enlargement, however, salivary cysts are rare. Herein, we present a case of pediatric Sjögren’s syndrome that presented as bilateral parotid cysts.

Methods:
A comprehensive search was conducted for cases of Sjögren’s syndrome presenting with parotid gland enlargement in the English language.

Results:
A 12-year-old female presented with a 2-month history of bilateral parotid masses. The patient denied any history of xerostomia, xerophthalmia, or constitutional symptoms. Imaging revealed bilateral complex cystic intraparotid masses. A right parotid gland biopsy was performed showing parotid gland parenchyma with dense lymphoplasmacytic infiltrate. Ultimately the presumptive diagnosis of Sjögren’s syndrome was made.

Conclusion:
We present a unique case of Sjögren’s syndrome with bilateral intraparotid cysts. This case illustrates the importance of a thorough workup to aid in diagnostic certainty. Parotid cysts associated with Sjögren’s are rare but should be considered within the differential diagnosis for pediatric patients with parotid swelling/mass.

Introduction

Sjögren’s syndrome is an autoimmune disease whereby the destruction of exocrine glands results in the loss of tear and saliva production.

The diagnostic criteria for Sjögren’s syndrome is both complex and controversial. The 2002 AECG criteria is the most commonly used diagnostic tool.

Diagnosis: 4 of 6 criteria (must include either IV or VI) or 3 of 4 objective criteria (II, IV, V, VI).

AECG Criteria:

I. Oral symptoms (at least 1)
   I. Dry eyes for ≥ 3 months.
   II. Oral foreign body sensation.
   III. Use of artificial tears ≥3 per day.

II. Oral symptoms (at least 1)
   I. Dry mouth ≥ 3 months.
   II. Recurrent or persistently swollen salivary glands.
   III. Need for liquids for swallowing dry foods.

III. Oral signs (at least 1)
   I. Abnormal Schirmer’s Test (<5 mm/5 min).
   II. Positive vital dye staining of eye surface.

IV. Histopathology
   I. Biopsy showing focal lymphocytic sialadenitis.

V. Oral signs (at least 1)
   I. Unstimulated whole saliva flow (<15 ml/15 min).
   II. Abnormal parotid sialography.
   III. Abnormal salivary scintigraphy.

VI. Autoantibodies (at least 1)
   I. Anti-SSA (Ro).
   II. Anti-SSB (La).

Case

A 12-year-old female presented with a 2-month history of bilateral parotid masses. She reported no history of xerostomia, dry eyes, or B symptoms.

Ultrasound imaging revealed bilateral complex cystic intraparotid masses measuring 4.7 x 3.1 x 1.7 cm on the right with a second, more posterior and deep mass measuring 4.3 cm at its greatest diameter. The left mass measured 3.2 cm. [Figure 1]

Magnetic resonance imaging confirmed numerous cystic lesions on the parotid bilaterally. These were hyperintense on T2, hypointense on T1. [Figure 2]

Parotid gland parenchyma with dense lymphoplasmacytic infiltrate was found on parotid gland biopsy.

Given the lack of diagnostic certainty, serology was ordered. Work-up was negative for infectious etiology such as HIV, mumps, CMV, and EBV. Serology was positive for rheumatoid factors, as well as anti-Ro/SSA and anti-La/SSB antibodies consistent with a diagnosis of Sjögren’s syndrome. The patient was subsequently treated with hydroxychloroquine.

Discussion

- Typical differential of parotid cysts include: mumps, HIV, lymphatic malformation
- Occurrence of parotid cyst with Sjögren’s Syndrome has been reported but is very rare especially in the paediatric population
- Parotid swelling, if painful, should warrant imaging whereby in contrast, painful parotitis may first be treated with conservative measures such as massage, analgesics, and sialogues

Conclusion

This is a unique case of pediatric Sjögren’s syndrome with bilateral intraparotid cysts. Our case illustrates the importance of thorough workup to aid in diagnostic certainty. Parotid cysts associated with Sjögren’s are rare, but should be considered within the differential diagnosis for any patient with parotid swelling/mass.

References