

Recurrent Parotitis as a First Manifestation in a Child with Primary Sjogren's Syndrome

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ABSTRACT

Recurrent parotitis is an acute, severe inflammation of one or both parotid glands, the major salivary glands in young children. We report the case of a seven-year old boy with Primary Sjogren's syndrome (PSS) who presented with 15 episodes of painful recurrent bilateral swellings of the parotid glands over a four-year period.

Keywords: Child, recurrent parotitis, Sjogren's syndrome

Parotitis Recurrente como Primera Manifestación en un Niño con el Síndrome de Sjögren Primario

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RESUMEN

La parotitis recurrente es una inflamación aguda, severa y reiterada de una o ambas glándulas parótidas – la mayor de las glándulas salivares – en los niños pequeños. Se reporta el caso de un niño de siete años de edad con el síndrome de Sjögren primario (SSP). El niño presentó 15 episodios de dolorosas inflamaciones bilaterales recurrentes de las glándulas parótidas por un periodo de cuatro años.

Palabras claves: Niño, parotitis recurrente, síndrome de Sjögren

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INTRODUCTION

Recurrent parotitis is an acute, severe inflammation of one or both parotid glands, the major salivary glands in young children. It has a multifactorial aetiology. Investigations for immunodeficiency *eg* HIV infection, Sjogren's syndrome (SS), and a family history is essential in cases with recurrent parotitis (1–7). Primary Sjogren's syndrome (PSS) is an uncommon cause of recurrent parotid inflammation in childhood (1, 2, 5, 6, 8, 9).

We report the case of a seven-year old boy with PSS who presented with 15 episodes of painful recurrent swellings of the parotid glands bilaterally over four years.

CASE REPORT

A seven-year old boy was referred to our clinic with recurrent episodes of parotitis every three to four months. He had 15

episodes of painful swellings of both parotid glands, over the last four years; it lasted 1–2 weeks each time.

The last episode began four to five days previously. There was no history of dryness of the eyes and mouth, joint swellings, pain and skin rashes as a feature of autoimmune disorders and syndromes. He was treated with antibiotics in each previous episode, but the symptoms recurred after stopping the antibiotics. He was well and asymptomatic between the episodes of parotitis.

On physical examination, his weight and height centiles were in normal ranges. The parotid swellings on examination were smooth, firm, painful and tender. There was no erythema around the parotid duct openings and there was no discharge on pressing the glands. The other systems were normal. His haemoglobin level and erythrocyte sedimentation rate were normal. Total leukocyte count was 18700/mm³; the distribution of cells (number and type) was unremarkable. HIV serology, biochemical analysis and immunoglobulin levels were normal. He had a positive antinuclear antibody. Serum rheumatoid factor, anti-double-stranded DNA and anti-smooth muscle antibody (anti-SMA)

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were negative and C3, C4, CH50 were within the normal ranges. Antibodies including anti-Ro and anti-La were “borderline positive” for Sjogren’s syndrome [ss-A:14 U/ml (0–15) ss-B:15] U/ml (0–15). Ophthalmological assessment revealed no abnormalities, his schirmer’s test was negative, staining with a 1% Rose-Bengal solution detected no corneal abnormality.

Ultrasonographic evaluation showed bilateral enlargement of the parotid glands with diffuse hypoechoic areas within the parenchyma. Computed tomography scan revealed a left parotid gland with increased contrast uptake and asymmetric size compared with the right gland. Hypodense areas were present in the left parotid gland. This view was assessed as parotitis without sialolithiasis. Salivary gland scintigraphy showed bilateral functional failure of the gland with symmetrical insufficient saliva excretion after secretory stimulus. A labial biopsy of the minor salivary glands showed several foci of periductal lymphocytic infiltration (Figure).

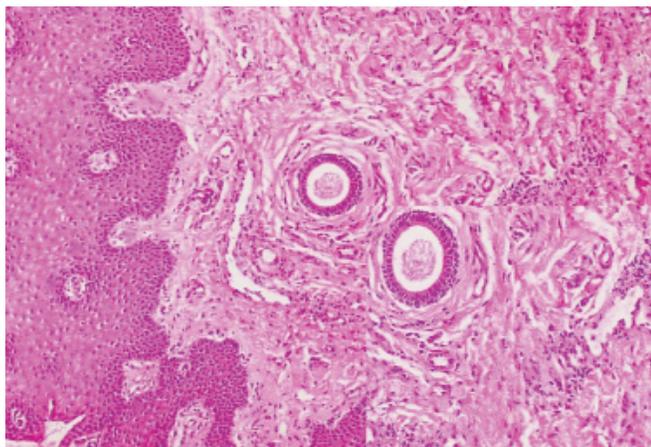


Figure: Microscopic section of buccal mucosa biopsy, stained with haematoxylen eosin. The dermis becomes increasingly collagenized, the epidermis thins and loses its adnexa (“atrophy”), and lympho-cytic infiltration in dermis (HE x 100).

Based on these findings, he was diagnosed with PSS. Because of severe pain and frequently repeated attacks of parotitis, he was given oral prednisolone (10 mg/day) and methotrexate (10 mg/m²/week) for 18 weeks. One year later he had no episodes of recurrent parotitis. He has remained clinically well and after 16 months of follow-up he has had only one episode of parotid swelling (resolved spontaneously).

DISCUSSION

Sjogren’s syndrome is a chronic inflammatory systemic autoimmune disease mainly affecting the exocrine and particularly, the salivary and lacrimal glands (1, 2, 5, 8). The condition usually occurs in adults. Primary Sjogren’s syndrome is rare in the paediatric population. The classic symptoms are keratoconjunctivitis sicca (dry eyes) and

xerostomia [dry mouth] (1–6, 8–11). Systemic manifestations of SS include vasculitis, autoimmune hepatitis, alveolitis, neuropathy, arthritis, renal tubular acidosis and nervous system involvement (1, 5, 8–11). Review of the literature identified case reports and small case series with PSS (1, 5, 8, 10, 12). The index patient was diagnosed with PSS. He presented with bilateral recurrent parotitis. His investigations revealed “borderline positive” anti-SSA and anti-SSB antibodies for PSS. However, the diagnosis was confirmed by minor labial salivary gland biopsy which showed lymphocytic infiltration.

Recurrent parotid swelling is the most common clinical manifestation of SS in children in the literature (1, 4, 5, 10–13). Recurrent parotitis is thought to be the first manifestation in childhood. This condition can present many years before the development of SS (1, 3, 14). McGuirt *et al* suggested that parotid inflammation is likely to occur earlier than minor salivary or lacrimal inflammation (13). Other important causes in the differential diagnosis of parotid swelling were juvenile recurrent parotitis, diffuse infiltrative lymphocytosis syndrome, sialoadenosis, sarcoidosis, lymphoma and infections with HIV, paramyxovirus, Epstein-Barr virus, hepatitis C virus, cytomegalovirus and parvovirus (1, 5, 12).

Also, recurrent parotitis has been associated with immune deficiency in the literature (5, 10, 12). Our patient had no primary or secondary immune deficiency. The presence of ANA (antinuclear antibody) has been reported in more than 80% of patients in most series of childhood SS with the majority of published reports since 1988. Anti-Ro/SSA antibodies are found in more than 70% of patients in most reports and anti-La/SSB antibodies are detected ranging from 30% to 70% (1, 5, 8, 10, 12, 13, 15, 16). Rheumatoid factor presence and hypergammaglobulinemia in SS are seen rarely in childhood than in adults (1, 6, 15).

The architecture of salivary ducts in childhood SS could be demonstrated by sialography. But it is an invasive procedure. Magnetic resonance imaging sialography is less invasive and less sensitive than traditional sialography. The diagnosis of SS requires a characteristic appearance of a biopsy sample from a minor salivary gland (13). A minor salivary gland biopsy with at least one foci of inflammation of greater than 50 cells is consistent with an SS diagnosis. Biopsy of the parotid gland may also have an important role, especially when SS is suspected and when a minor salivary gland biopsy is nondiagnostic (13). Our patient was diagnosed as primary SS with a positive minor salivary gland biopsy after other causes of recurrent parotitis were excluded.

Therapy includes topical agents to improve moisture and decrease inflammation. Systemic therapy includes steroidal and nonsteroidal anti-inflammatory agents, disease-modifying agents and cytotoxic agents to address extraglandular manifestations involving lung, heart, kidneys, skin, haematological, nervous and lymphoproliferative disorders (4). Although appropriate corticosteroids and

immunosuppressants may provide the suppressive effects on the progressive inflammatory destruction of secretory glands, further evaluation with more patients is needed to determine the inclusion criteria of these treatments for sicca (Sjogren's) syndrome, especially in cases with no other organ involvement (17).

There is no consensus about the diagnosis and management of primary SS with recurrent parotitis. Further evaluation is needed.

In conclusion, although PSS is a rare diagnosis in childhood, it should be considered in the differential diagnosis of recurrent parotitis, especially in a child with "borderline positive autoantibody" profile and a positive minor salivary gland biopsy, even in the absence of sicca symptoms.

REFERENCES

1. Singer NG, Tomanova-Soltys I, Lowe R. Sjogren's syndrome in childhood. *Curr Rheumatol Rep* 2008; **10**: 147–55.
2. Fox RI. Sjogren's syndrome. *Lancet* 2005; **366**: 321–31.
3. Sugimoto T, Uzu T, Kashiwagi A. Recurrent parotitis as a first manifestation of adult primary Sjogren's syndrome. *Intern Med* 2006; **45**: 831–2.
4. Houghton K, Malleon P, Cabral D, Petty R, Tucker L. Primary Sjogren's syndrome in children and adolescents: are proposed diagnostic criteria applicable? *J Rheumatol* 2005; **32**: 2225–32.
5. Civilibal M, Canpolat N, Yurt A, Kurugoglu S, Erdamar S, Bagci O et al. A child with primary Sjogren syndrome and a review of the literature. *Clin Pediatr (Phila)* 2007; **46**: 738–42.
6. Munro J, Allen R. Recurrent parotitis and Sjogren's syndrome. *J Paediatr Child Health* 2003; **39**: 158–9; author reply 159.
7. Leerdam CM, Martin HC, Isaacs D. Recurrent parotitis of childhood. *J Paediatr Child Health* 2005; **41**: 631–4.
8. Stiller M, Golder W, Döring E, Biedermann T. Primary and secondary Sjogren's syndrome in children – a comparative study. *Clin Oral Invest* 2000; **4**: 176–82.
9. Ohlsson V, Strike H, James-Ellison M, Tizard EJ, Ramanan AV. Renal tubular acidosis, arthritis and autoantibodies: primary Sjogren's syndrome in childhood. *Rheumatology* 2006; **45**: 238–40.
10. Cimaz R, Casadei A, Rose C, Bartunkova J, Sediva A, Falcini F et al. Primary Sjogren syndrome in paediatric age: a multicentre survey. *Eur J Pediatr* 2003; **162**: 661–5.
11. Chudwin DS, Daniels TE, Wara DW, Ammann AJ, Barrett DJ, Whitcher JP et al. Spectrum of Sjogren syndrome in children. *J Pediatr* 1981; **98**: 213–17.
12. Nikitakis NG, Rivera H, Lariccia C, Papadimitriou JC, Sauk JJ. Primary Sjogren syndrome in childhood: Report of a case and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2003; **96**: 42–7.
13. McGuirt WF Jr, Whang C, Moreland W. The role of parotid biopsy in the diagnosis of pediatric Sjogren syndrome. *Arch Otolaryngol Head Neck Surg* 2002; **128**: 1279–81.
14. Stein M, Miller G, Green L. Prophylactic antibiotics in recurrent parotitis in a patient with Sjogren's syndrome. *Clin Rheumatol* 1999; **18**: 163–4.
15. Tomiita M, Saito K, Kohno Y, Shimojo N, Fujikawa S, Niimi H et al. The clinical features of Sjogren's syndrome in Japanese children. *Acta Paediatr Jpn* 1997; **39**: 268–72.
16. Pessler F, Emery H, Dai L, Wu YM, Monash B, Cron RQ et al. The spectrum of renal tubular acidosis in paediatric Sjogren syndrome. *Rheumatology (Oxford)* 2006; **45**: 85–91.
17. Iwata N, Miyamae T, Kikuchi M, Kishi T, Hara R, Kaneko U et al. Clinical and histological experience of labial lip biopsy in juvenile Sjogren syndrome. *Nihon Rinsho Meneki Gakkai Kaishi* 2009; **32**: 195–200.