

Systemic lupus erythematosus presenting as parotitis and secondary Sjogren's syndrome

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ABSTRACT

A 38-year-old woman presented with right-sided parotid swelling, dry mouth and dry eyes of one year duration. Her Schirmer's test and sialometry were positive and histopathology showed lymphoplasmacytic infiltration. She also had concomitant normochromic, normocytic anaemia and mild haematuria. Her anti-nuclear antibody titre was also positive, 1:640, with a speckled pattern on immunofluorescence. We discuss the atypical presentation of systemic lupus erythematosus, particularly parotitis and secondary Sjogren's syndrome.

Keywords: parotitis, Sjogren's syndrome, systemic lupus erythematosus

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INTRODUCTION

Systemic lupus erythematosus (SLE) is a multiorgan autoimmune disease which is characterised by the presence of auto-antibodies and immune complexes.⁽¹⁾ SLE usually presents as a multi-systemic disease and frequently manifests as musculoskeletal and cutaneous presentations, such as arthritis, myalgia and malar rash.^(2,3) SLE, however, can present atypically and manifest as parotitis, pericarditis, hepatomegaly or peripheral lymphadenopathy.^(2,3) In these instances, factors that may allude to an underlying diagnosis of SLE include abnormal full blood count (FBC), abnormal urinalysis and positive anti-nuclear antibody (ANA) titre and positive anti-double stranded deoxyribonucleic acid antibody (anti-dsDNA) assay.^(2,4) We discuss the atypical presentation of parotitis and secondary Sjogren's syndrome in a patient with SLE. We also discuss the myriad of differential diagnoses that need to be considered and systematically ruled out in order to establish the diagnosis.

CASE REPORT

A 38-year-old woman presented to the otorhinolaryngology department with right-sided cheek swelling that was progressive over the preceding one year. She also

complained of intermittent dryness of her mouth and eyes. There was no fever, night sweats or weight loss, and her appetite was good. There was no mouth ulcer, alopecia, joint pain, photosensitivity or skin rash. She denied any history of "rheumatism". She was a teetotaler, non-smoker and was not on any regular medication. She had three children, and had a history of two spontaneous abortions in the past. None of her children developed congenital heart block, or had symptoms and signs suggestive of neonatal lupus.

On clinical examination, she was of normal stature, and was pale but not jaundiced. There were no arthritic changes in her hands, nor was there any nail dystrophy. There was no butterfly rash, petechiae or ecchymoses. She had no mouth ulcer and her dentition was good. There was no alopecia or scar apparent. The swelling of her right cheek was diffusely enlarged, firm, non-tender, and extended from just below the right ear lobe to the angle of the jaw. There were no cervical or peripheral lymphadenopathy, no hepatosplenomegaly and no erythema nodosum. Her cranial nerve examination was normal and the rest of her systemic examination was unremarkable.

A Schirmer's test was positive with ≤ 5 mm of tear production in 5 minutes. Her sialometry was also positive with ≤ 1.5 ml of saliva in 15 minutes. A fine needle aspiration biopsy revealed lymphoplasmacytic infiltrates adjacent to the salivary glands and ducts. There was no granuloma, no abnormal lymphocytes or plasma cells, and no sinus histiocytosis or lymphocytophagocytosis. An initial diagnosis of primary Sjogren's syndrome was made and she was started on artificial tears and saliva. Concurrent serological investigation then showed a positive assay for rheumatoid factor, an ANA titre of 1:640 with a speckled pattern on immunofluorescence, and a positive qualitative assay for anti-dsDNA. She was then referred to the rheumatology clinic for further assessment.

A peripheral blood film showed normochromic, normocytic anaemia (haemoglobin 10 g/dL) with no evidence of haemolysis, erythrocyte sedimentation rate (ESR) of 80 mm/hour, 1+ haematuria on urinalysis which was dysmorphic on phase contrast microscopy,

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and normal C-reactive protein (CRP) of ≤ 0.5 mg/dL. Laboratory evaluation showed reduced C3 and C4, positive anti-Ro/SSA and anti-La/SSB assays and negative anticardiolipin, anti-Sm and anti-RNP antibody assays. A chest radiograph showed no apical fibrosis or hilar lymphadenopathy. Her serum calcium and lactate dehydrogenase (LDH) were normal, and her human immunodeficiency virus (HIV), hepatitis-B virus (HBV) and hepatitis-C virus (HCV) screens were negative. She was subsequently diagnosed to have SLE with secondary Sjogren's syndrome, and started on low-dose prednisolone. At follow-up three months later, she noted that the parotid swelling was less and her symptoms had improved.

DISCUSSION

SLE is a multiorgan systemic autoimmune disease characterised by the presence of autoantibodies and immune complexes.⁽¹⁾ SLE usually manifests as musculoskeletal and cutaneous presentation, such as arthritis, malar rash and photosensitivity. SLE may also present atypically as parotitis, pericarditis, peripheral lymphadenopathy or hepatosplenomegaly.^(2,3) SLE manifests atypically in up to one-third of cases. Parotitis per se occurs in 2.5% of cases of SLE presenting atypically.⁽²⁻⁴⁾ In cases where SLE manifests atypically, it is usually accompanied by elevated ESR, abnormal FBC, abnormal urinalysis, positive ANA titre and anti-dsDNA assay and low complement, as was found in this patient.⁽²⁻⁴⁾

However, before a diagnosis of SLE with secondary Sjogren's syndrome can be adopted, several differential diagnoses need to be ruled out. In this case, lymphoma was ruled out by the negative histopathology, normal serum LDH and calcium levels, absence of constitutional symptoms and the absence of any peripheral lymphadenopathy or hepatosplenomegaly. These findings also ruled out another less common but important differential diagnosis, namely, Rosai-Dorfman disease. Furthermore, pulmonary tuberculosis and sarcoidosis were ruled out by a normal chest radiograph and the absence of any clinical indicators such as prolonged cough with expectoration, weight loss or the presence of erythema nodosum. Serology for HIV and the hepatitis viruses HBV and HCV were also negative.

These negative findings effectively ruled out the differential diagnoses associated with parotitis.

In this case, SLE was diagnosed based on the findings of normochromic, normocytic anaemia, haematuria, elevated ESR despite a normal CRP, reduced C3/C4 and strongly positive titre for ANA and a positive anti-dsDNA assay. These, coupled with a positive Schirmer's and sialometry, positive anti-Ro/SSA and anti-La/SSB and lymphoplasmacytic infiltrates on histopathological examination of the salivary gland, satisfied the American-European Revised Classification of Sjogren's Syndrome criteria for secondary Sjogren's syndrome.⁽⁵⁾ It has been reported that up to 30% of patients with SLE go on to develop an additional autoimmune disease and of these, the most common was secondary Sjogren's syndrome, which occurred in 13% of cases.⁽⁶⁾ The probability of secondary Sjogren's syndrome in this patient was further enhanced by the presence of positive anti-Ro/SSA and anti-La/SSB assays.⁽⁶⁾

In conclusion, SLE is a great imitator and may present atypically in up to 30% of cases. When it does present atypically, it behoves clinicians to systematically rule out other potential differential diagnoses, and corroborative clinical indicators to support the diagnosis of SLE need to be elucidated. Only then can the diagnosis be safely adopted and the appropriate management instituted.

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