CASE REPORT

JIACM 2018; 19(3): 207-09

Primary Sjögren's Syndrome without Sicca Symptoms: An Uncommon Entity

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Abstract

Primary Sjögren's Syndrome (PSS) is an autoimmune disorder characterised by chronic inflammation and destruction of lacrimal and salivary glands. Dryness of mouth (xerostomia) and dryness of eye (keratoconjunctivitis sicca) are sine qua non for diagnosis of PSS. Herein, we present a very unusual case of a middle-aged lady who presented with only complaints of unilateral parotid swelling and was subsequently diagnosed as PSS with type 1 renal tubular acidosis (RTA).

Keywords: Renal tubular acidosis, hypokalaemia, xerostomia.

Introduction

Sjögren's syndrome (SS) is a common rheumatological condition characterised by dryness of eyes (keratoconjunctivitis sicca) and dryness of mouth (xerostomia). The mean age at the onset of symptoms is the fourth decade. The disease is nine times more prevalent among females[1]. Xerostomia and keratoconjuctivitis sicca was the presenting symptom in 98% and 93% of SS patients, respectively, in a large prospective cohort[2]. SS is a chronic disease with insidious onset and varied presentation, often making diagnosis difficult or delayed. PSS in childhood often presents without sicca symptoms but in adults absence of sicca symptoms at the time of diagnosis is rare[3]. We present a case of middle-aged lady who was being evaluated for long standing unilateral parotid swelling and was diagnosed to be having PSS.

Case report

A 32-year-old lady presented in Medicine OPD with complaints of gradually progressive swelling on left side of the neck for the last 8 months. There was no history of pain, fever, joint pain, dryness of mouth/eyes or weight loss. A Fine needle aspiration cytology (FNAC) was attempted from the swelling four months back which was inconclusive and resulted in persistent discharge from that site. She gave history of cervical tuberculosis 7 years back, for which she had received nine months of anti-tubercular therapy. On examination, there was a well defined swelling of size 5 x 5 cm in the left parotid region, which was erythematous, non-tender and firm in consistency. There was a pus point just above the swelling with minimal discharge. There was no significant cervical lymphadenopathy and rest of the systemic examination was essentially normal.

A possibility of sarcoidosis/tuberculosis/chronic parotitis was kept and further investigations were planned accordingly. Complete haemogram was normal except for elevated ESR (52 mm/hr). Renal and liver function tests were normal; however, her serum potassium levels were persistently low. Serum ACE levels, serum calcium and 24 hour urinary calcium levels were normal. Mantoux test was strongly positive (27 x 25 mm). But, GeneXpert MTB, AFB and Gram's stain from pus was negative. Hepatitis B surface antigen (HBsAg), antibody to hepatitis C virus (anti HCV) and HIV were negative.

An ultrasound of neck showed multiple hypoechoic areas in both parotid glands. To look for any other evidence of systemic disease, a CECT chest and abdomen was done, which was also normal. A repeat FNAC done from the swelling did not reveal any significant abnormality except

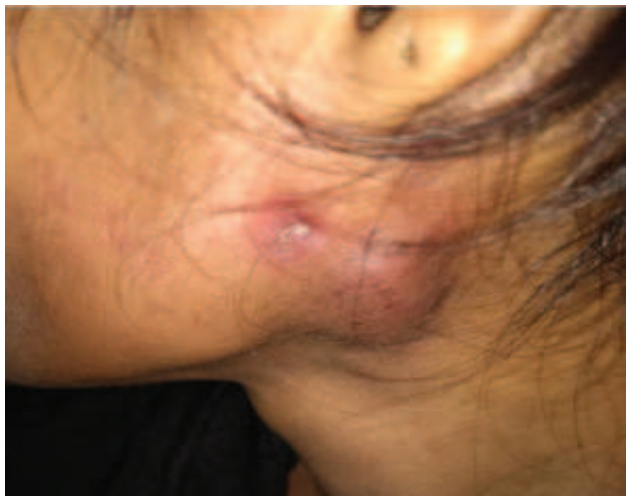

Fig. 1: *Left parotid swelling.*

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some reactive changes with negative AFB stain and Gene Xpert. In the meantime, we also evaluated the cause for persistent hypokalaemia.

Urinary potassium was 54 mmol/day (normal – <15 mmol/day) with high TTKG (10.7), suggestive of renal wasting. An arterial blood gas analysis (ABG) showed normal anion gap metabolic acidosis (pH – 7.34, HCO_3 – 18 mmol/l, pCo_2 – 36 mm Hg). So, a diagnosis of renal tubular acidosis (RTA) was made and since urinary pH was 7, the RTA was labeled as distal (type 1). Subsequently, a MRI scan of parotids was done which revealed bilateral cystic dilatation of the parotid gland with areas of hyperintensities due to fatty infiltration. This honey-comb or salt and pepper appearance of parotids was suggestive of Sjögren's syndrome. Schirmer's test and rose bengal dye test done to look for dry eye was normal. ANA test was strongly positive in a titre of 1:640 with a speckled pattern. ENA profile done showed strongly positive Anti Ro 52 (3+) and Anti La (3+) antibodies. A labial salivary biopsy was done which showed focal lymphocytic sialadenitis with focus score of more than 1. Thus, a final diagnosis of primary Sjögren's syndrome with type 1 RTA and without sicca symptom was made. She was given tablets of sodium bicarbonate with oral potassium supplementation. A short course of antibiotic was given to treat the superadded bacterial infection.

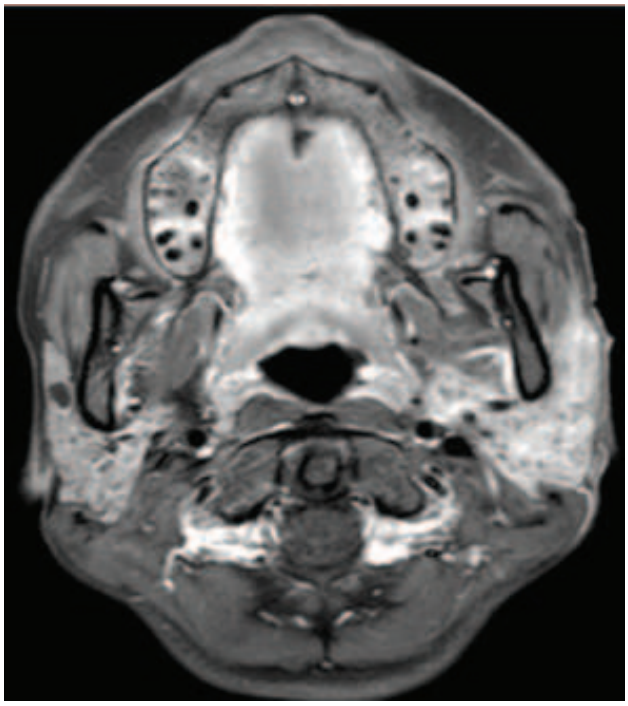

Fig. 2: *MRI parotid T1W post-contrast image showing cystic dilatation (left > right).*

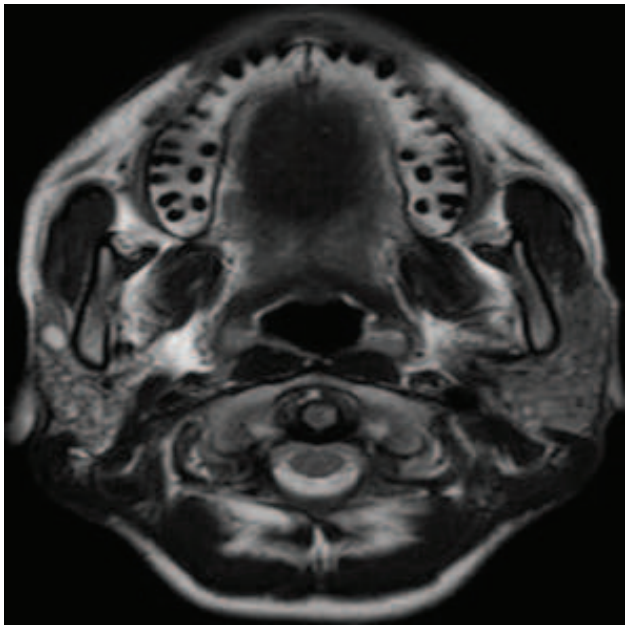

Fig. 3: *MRI parotid T2W image showing honeycomb appearance of parotids.*

Discussion

Sjögren's syndrome is a common chronic autoimmune condition causing destruction of exocrine glands. Sjögren's syndrome is further classified into primary (PSS) and secondary (SSS) [4]. Secondary Sjögren's syndrome is characterised by sicca symptoms in presence of other connective tissue disease such as rheumatoid arthritis, systemic lupus erythematosus or scleroderma. The pathogenesis of SS includes chronic inflammation of lacrimal and salivary glands due to dysregulation of B and T cells[5].

Extraglandular manifestations are common in PSS and the risk increases in presence of hypocomplementaemia and hypergammaglobulinaemia. The commonly involved organ systems in PSS include lung, musculoskeletal system, kidney and nervous system. Renal disease in PSS includes interstitial nephritis, type 1 RTA, glomerulonephritis and nephrogenic diabetic insipidus. The diagnosis of type 1 (Distal) RTA is generally made while evaluating asymptomatic hypokalaemia, as in present case. However, rarely type 1 RTA can lead to severe potassium wasting and subsequently muscle paralysis[6]. Distal RTA is characterised by normal anion gap metabolic acidosis, urinary potassium wasting and high urinary pH.

There are several classification criteria for diagnosis of PSS which includes both subjective and objective involvement of exocrine glands along with presence of autoantibodies (Anti Ro/Anti La) and focal lymphocytic sialadenitis[7]. Salivary gland involvement can be assessed by scintigraphy, parotid gland sialography and by ultrasound or MRI of parotids. MRI

is now a commonly used modality in diagnosis of PSS and has high sensitivity[8]. The MRI finding in advanced cases of PSS consists of multiple areas of high-signal intensity with areas of low-signal intensity, also known as "honeycomb" or "salt and pepper" appearance[9]. The other differentials that should be considered in presence of bilateral parotid enlargement includes sarcoidosis, tuberculosis, chronic hepatitis C infection, diabetes, alcoholism, IgG4 related disease, chronic recurrent parotitis and lymphomas[10]. Non Hodgkin's lymphomas (NHL) are also common in PSS and should be suspected if there is rapid increase in size of one of the parotid glands. Diagnosis of NHL can often be made by imaging of parotids followed by biopsy. Treatment of PSS includes management of sicca symptoms by artificial tear substitutes and cholinergic drug such as pilocarpine or cevimeline for xerostomia. Immunosuppressants are reserved for severe extra glandular involvement.

To conclude, PSS can present without sicca symptoms and there should be high index of suspicion for the same while evaluating any chronic parotid swelling or type 1 RTA.

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