Case Report

A case of progressive systemic sclerosis/lupus overlap syndrome: Presenting with parotid swelling

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Abstract

An overlap syndrome is a medical condition which shares features of at least two more widely recognized disorders. Autoimmune connective tissue diseases include systemic lupus erythematosus (SLE), scleroderma, polymyositis, dermatomyositis, rheumatoid arthritis and Sjögren's syndrome where overlap syndrome most commonly seen in combination with SLE and systemic sclerosis (SSc). Sjogren's is an autoimmune exocrinopathy, in which systemic diseases such as arthritis, interstitial lung disease and renal disease may develop in addition to the pathognomonic features such as dry eyes and mouth. The other associated disease with Sjogren's includes sialadenitis. Sialadenitis of the parotid gland is one of the major disorders of salivary gland. This article presents a rare case report of a patient diagnosed with sialadenitis of the parotid gland and associated with progressive SSc/lupus overlap syndrome and secondary Sjogren's.

Keywords: Autoimmune connective tissue diseases, lupus overlap syndrome, overlap syndrome, progressive systemic sclerosis, secondary Sjogren's

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INTRODUCTION

Overlap syndrome is an entity that includes the diagnostic criteria of at least two distinct diseases. Twenty-five percent of patients with rheumatic diseases with systemic symptoms cannot be definitely diagnosed, as the patients fail to meet the adequate criteria or have symptoms and signs of several diseases.[1,2] One of the well-characterized overlap syndromes, i.e., mixed connective tissue disease, is defined by *anti-RNP (ribonucleoprotein)* autoimmunity presenting systemic lupus erythematosus (SLE) accompanied by features of systemic sclerosis (SSc).[1] Pulmonary arterial hypertension (PAH) is a severe manifestation of SSc and SLE with a prevalence rate ranging, respectively, from 5% to 60% and from 0.5% to 17.5%.[3,4] Sjögren syndrome (SS) is an autoimmune condition which can be of primary and secondary.[5] SS can be associated with sialadenitis, which is a chronic autoimmune inflammatory exocrinopathy affecting the salivary and lacrimal glands and may occur as a primary or secondary form. The various imaging modalities to investigate the salivary gland disorders include conventional radiography, sialography, ultrasonography, computerized tomography (CT), radionuclide imaging and magnetic resonance imaging (MRI).[6,7] This article showcases a rare case report of chronic sialadenitis of the parotid gland associated with an autoimmune disorder.

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CASE REPORT

A 69-year-old female reported to the department of oral pathology with the chief complaint of pain in the left side of the face for the past 1 month. History of presenting illness revealed a swelling which was initially small and gradually progressed 1 week before to the current state. Pain was sudden in onset, intermittent in nature, which aggravated on mastication and did not subside over medication (the patient was advised Combiflam tablet for 5 days, twice daily and intramuscular injection of cefixime for 3 days by physician whom she visited earlier). The patient revealed a past medical history of regular treatment for progressive SSc/lupus overlap syndrome, secondary Sjogren's with interstitial lung disease and PAH for the past 10 years. The patient was hospitalized for left lower limb cellulitis 8 years back and was discharged after complete observation and medication. Vitamin D insufficiency was suspected and corrected by calcirol granules.

The patient also had a complaint of difficulty in swallowing and spasm-type pain which was diagnosed as esophageal dysmotility and reflux disorder and was advised on prokinetic agents, low-fat diet and to avoid cold exposure to extremities. The patient also revealed a history of whitish skin lesion on both her legs and foot 20 years before these lesions have not progressed. The patient revealed no history of autoimmune disease in the family. The patient had undergone multiple uneventful extractions and complete prosthetic denture treatment. The patient also revealed a history of impaired taste sensation. On general examination, patient's vital signs were normal, and left submandibular lymph node was palpable, roughly oval, firm in consistency, tender and mobile.

On extraoral examination, facial asymmetry was seen in the left side of the face as a solitary diffuse swelling. The swelling extends from the middle third of the body of the mandible anteriorly to 1 cm behind the auricle posteriorly and superiorly at the level of the tragus to inferior border of the angle of the mandible inferiorly. Skin overlying the swelling appeared normal, with no other abnormalities detected [Figure 1]. Swelling was firm to hard in consistency, noncompressible, tender and warm on palpation.

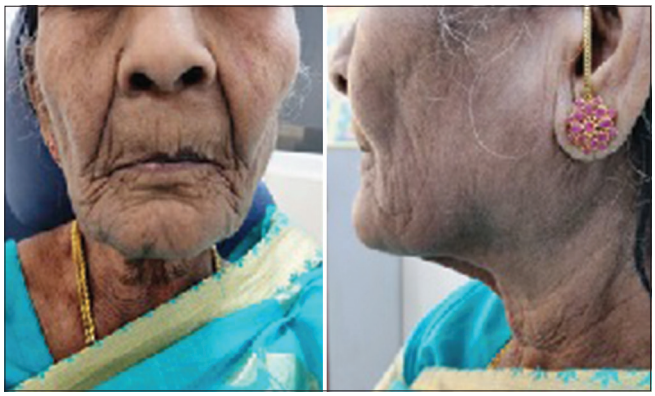

Figure 1: Profile and facial asymmetry of the patient

On intraoral examination, inspection revealed inflamed parotid duct orifice on the left side and was tender on palpation. The presence of erythematous ulceration was seen at the right and left commissures of the lip suggestive of angular cheilitis [Figure 2]. A solitary ulcer was present in the lower labial vestibular region having erythematous margin with sloping border which was freely movable and tender on palpation suggestive of denture-induced traumatic ulcer. Hard-tissue examination revealed upper and lower completely edentulous arch.

Salivary flow rate and pH were estimated to be 1 ml/min and 6.7, respectively. The quality of life of the patient was assessed using a questionnaire [Figure 3]. This self-evaluation was made to educate the patient about the impact of the oral dryness.

Patient informed consent was obtained. Blood investigations were performed which were within normal limits. The patient was subjected to ultrasound examination which revealed minimal enlargement of left parotid gland with mild-to-moderate intraglandular ductal dilatation [Figure 4a]. CT and MRI investigation revealed asymmetrical diffuse enlargement of left parotid gland with mild hyperintense signals, few specks of calcification and multiple branching and nonbranching cystic spaces. Mild irregular dilatation of the left parotid duct was elicited. Multiple enlarged submandibular level neck nodes were seen (largest – 2.5 cm × 1.1 cm, III level). Multiple calcific specks with few cystic spaces were seen in the superficial

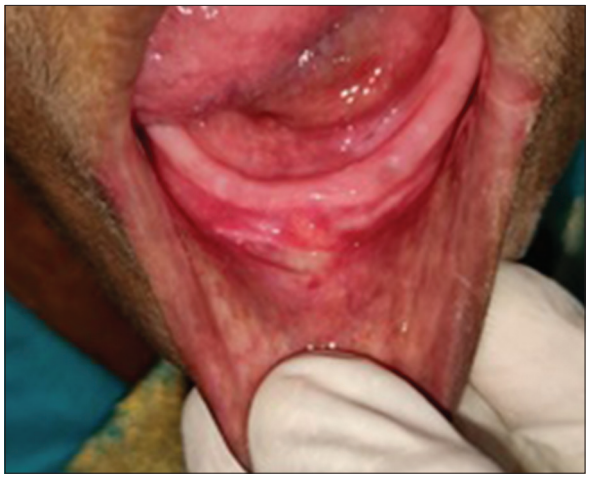

Figure 2: Traumatic ulcer in the lower labial vestibule and angular cheilitis

lobe of the right parotid gland with no significant duct dilatation [Figure 4b and c].

Based on the thorough history, clinical examination and investigations, the case was diagnosed as right chronic sialadenitis and left acute sialadenitis of parotid gland. The differential diagnosis considered was viral sialadenitis, pleomorphic adenoma and myositis ossificans. Ketorol DT 10 mg (analgesic) was prescribed for 1 week, and warm compression was advised to the patient following a review after 1 week the swelling reduced considerably (70%).

Xerostomia - Quality of Life Questionnaire

1. My mouth dryness limits the amount of food I eat

Not at all	A little	Somewhat	Quite a bit ✓	Very much

2. My mouth dryness causes discomfort.

Not at all	A little	Somewhat	Quite a bit ✓	Very much

3. My mouth dryness causes lot of worry /concern

Not at all	A little	Somewhat	Quite a bit	Very much ✓

4. My mouth dryness makes me uncomfortable eating/speaking to others

Not at all	A little	Somewhat	Quite a bit	Very much ✓

5. Mouth dryness has bad effect on tasting food

Not at all	A little	Somewhat	Quite a bit	Very much ✓

6. My mouth dryness reduces my general happiness with life

Not at all	A little	Somewhat	Quite a bit ✓	Very much

7. My mouth dryness interferes with my daily activities

Not at all	A little	Somewhat	Quite a bit ✓	Very much

8. If you were to spend rest of your life with your mouth dryness just the way it is now how would you feel about this

Delighted	Mostly satisfied	Mixed : equally satisfied/dissatisfied	Mostly dissatisfied	Terrible ✓

Figure 3: Questionnaire on Quality of Life

Complete resolution was seen on the 2nd week. The patient was followed up for 2 months at an interval of 15 days. No recurrence was reported to date.

DISCUSSION

"Overlap syndrome" is the terminology used to report patients with two or more widely recognized disorders. Usually, they present subacutely with clinical manifestations that include different organ systems. Overlap syndrome can be found in many medical specialties; one such is connective tissue disorder of rheumatology and overlapping genetic disorder in cardiac system. It has been reported that the pattern of organ involvement reflects the characteristic features of the well-defined rheumatic diseases occurring together. Few other commonly associated conditions include polymyositis with either SLE or SSc.[8] Very rarely cases have been reported with the combination of both SLE and SSc; our case was one such.

The key diagnostic factors include some of the following factors such as arthritis/arthralgia, swollen hands, sclerodactyly, skin rashes and lymphadenopathy. The occurrence of Sjogren's is more frequent in female, especially in the middle age.[9]

The treatment followed is mainly the use of immunosuppressants and corticosteroids. Recently, replacement intervention followed for refractory cases of overlap syndrome is the use of biologic drugs, i.e., anti-tumor necrosis factor α or anti-CD20 monoclonal antibodies. However , the ill effect of using such drugs are high chance of disease exacerbation in these patients.[10]

Our patient was also diagnosed with secondary Sjogren's with interstitial lung disease and PAH for the past 10 years. Although overlap syndrome could lead to a diagnostic

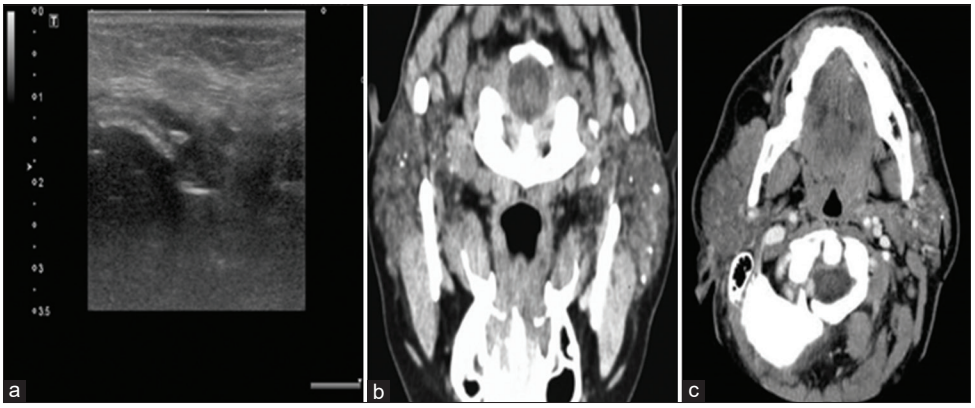

Figure 4: (a) Ultrasound examination reveals minimal enlargement of the left parotid gland with mild-to-moderate intraglandular ductal dilatation. (b) Computerized tomography and (c) magnetic resonance imaging showing multiple calcific specks with few cystic spaces seen in the superficial lobe of the right parotid gland and multiple branching and nonbranching cystic spaces with specks of calcification seen in the left parotid gland

challenge, it is of great importance as it provides queue for mechanisms of various other diseases related to it.

Sialadenitis is an inflammation of the salivary glands; subsequently, it can be classified as acute, chronic or recurrent forms. Chronic sialadenitis of the parotid is an inflammatory disorder which is often associated with pain.[11] In most of the cases, when left untreated often leads to fibrous mass formation in the gland. The most common age where sialadenitis can be encountered is middle age.

Painless swellings (unless secondarily infected) classically occur in *Autoimmune sialadenitis* (i.e., SS) and may be unilateral or bilateral. These conditions include calculi, salivary stasis and a change in the fluid and electrolyte composition of the gland. The pathogenesis of the disease is not fully understood but is likely to involve the interplay of several etiological factors, of which reduced salivary flow is the most significant. Once reduced, either by decrease in the acinar tissue or duct obstruction, ascending infection more easily occurs. There are two theories which have been put forward regarding the initiation of chronic sialadenitis. One postulates that retrograde infection by low-grade opportunistic oral flora can result directly in chronic recurrent sialadenitis. The other proposes that repeated episodes of acute infection may lead to mucus metaplasia of ductal epithelium, resulting in increased mucus content of secretions, stasis and further episodes of inflammation.[6,7]

In our case, the patient only had unilateral swelling; however, on investigation, it was diagnosed as right chronic sialadenitis and left acute sialadenitis of parotid gland. In this case, the parotid swelling well responded to our treatment protocol. Owing to the medical history of our patient, such conditions could be misdiagnosed or misinterpreted. This report adds to the list of differential diagnosis for unilateral parotid swellings, especially in the context of patients presenting with overlap syndrome. Although salivary gland involvement and enlargement of parotid gland swelling is very common in cases with a prior history of autoimmune diseases, it has been very sparsely reported with overlap syndrome.

CONCLUSION

This case report adds to the sparse literature that has been reported with a case of progressive SSc/lupus overlap syndrome – presenting with parotid swelling. Our patient was managed very effectively and no recurrence of swelling to date. Knowledge of such rare diseases is very important for a clinician as overlap syndrome is associated with a high risk of morbidity and mortality than each of the diseases alone.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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