ISSN: 2581-5989
PubMed - National Library of Medicine - ID: 101738774

International Journal of Dental Science and Innovative Research (IJDSIR)

IJDSIR : Dental Publication Service

Available Online at: www.ijdsir.com

Volume – 3, Issue – 2, April - 2020, Page No. : 36 - 39

Mononeuritis Multiplex: A Rare Case Report of the Extra-glandular Manifestation of Sjogren syndrome

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Citation of this Article: Dr. Nimisha Divakar, Dr. Manoj Vengal, Dr. Auswaf Ahsan, Dr. Ashir K R, "Mononeuritis Multiplex: A Rare Case Report of the Extra-glandular Manifestation of Sjogren syndrome", IJDSIR- April - 2020, Vol. – 3, Issue -2, P. No. 36 – 39.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

The neurologic symptoms of Sjogren Syndrome are often ignored. Mononeuritis multiplex is a peripheral motor neuropathy that is not rare with primary Sjogren Syndrome, but is less reported in literature. Early diagnosis of neurological symptoms can help prevent the progression of disease and thereby improve the patient's quality of life. Here, we present a case report where the patient was under neurological consultation for mononeuritis multiplex with no reduction in symptoms for the past one year. She visited us with a complaint of recurrent parotid abscess, after clinical and laboratory examination, diagnosis of Sjogren syndrome was made. Identification of the relationship between Sjogren Syndrome and mononeuritis multiplex was the key to delivering proper treatment to this patient

Keywords: Mononeuritis multiplex, Sogren Syndrome, Parotid abcess,

Introduction

Sjogren's syndrome (SS) is an autoimmune inflammatory disorder characterized by lymphocytic infiltration of exocrine glands, mainly the lacrimal and salivary glands leading to a chronic sicca syndrome. Extra glandular organ systems may frequently be involved, including both the central and peripheral nervous systems. However there is limited mention in the literature, it is commonly acknowledged that about 20% of pSS (primary Sjogren's Syndrome) patients will present neurological manifestations.[1] Treating the neurological condition alone

without considering Sjogren syndrome as the underlying cause will not bring reduction in the manifestation of the syndrome. Here the patient came with the complaint of recurrent parotid abscess and further examination helped to arrive at the diagnosis of Sjogren syndrome which is the main underlying cause of both the neurologic complication and parotid abscess.

Case report

A 52-year-old Indian female patient came to the dental clinic complaining of pain in the left ear region since 2 days where she noticed a pus filled swelling (Fig.1). She reported that she has experienced the same problem around 3 times for the last four years, bilaterally. She visited a dentist where incision and drainage of the swelling was done each time. She was also prescribed antibiotics but she never had complete remission of her symptoms. One year back, the patient experienced tingling in her hands and feet along with foot and hand drop and numbness on one side. On neurological examination, she was diagnosed to have mononeuritis multiplex and was under treatment for the same.

On clinical examination, the patient had dry eyes which caused her much difficulty in opening them. Rheum was present in the medial canthus of both eyes. (Fig. 2) When touched with cotton, she experienced a gravel-filled feeling in her eyes. Her lips were cracked, tongue was fissured, (Fig.3, Fig.4) and tongue blade sign was positive. While milking the gland, there was pus discharge from the parotid duct which after bacterial culture was diagnosed to be bacterial sialadenitis. The culture was positive for E.Coli and methicillin resistant Staph. aureus. Taking into consideration the recurrent bacterial sialadenitis, dry eye, dry mouth and neurologic complications, a provisional diagnosis of Primary Sjogren's syndrome was made. Schirmer's eye test and minor salivary gland biopsy were done and the patient was asked to do blood investigation for anti- Ro, anti-La autoantibodies and cryoglobulin. All the investigations were positive which helped arrive at a diagnosis of Sjogren's syndrome, which was in turn the root cause for mononeuritis multiplex.

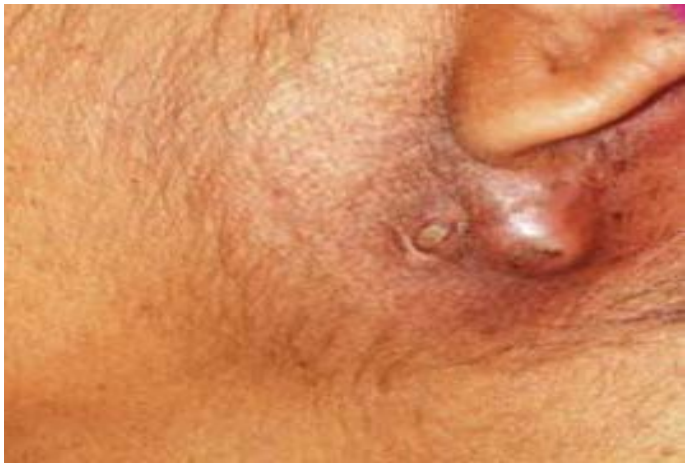

Fig 1: Parotid abscess below the left ear lobe (thick arrow) and scar of old abscess (thin arrow)

Fig 2: Rheum in the medial canthus of left eye

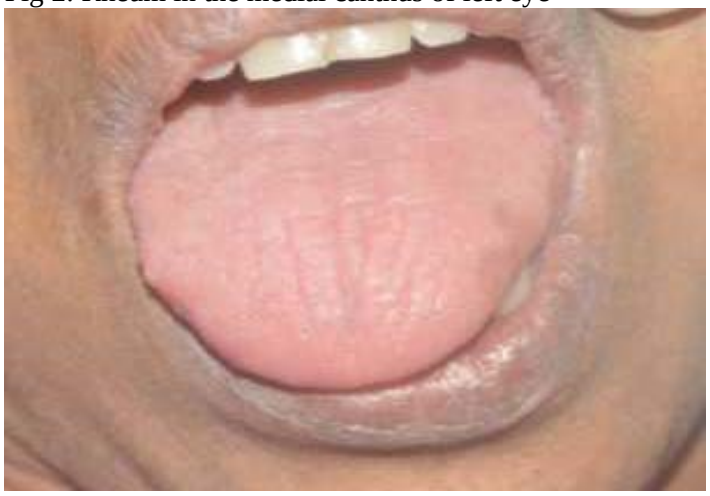

Fig 3: Fissured tongue

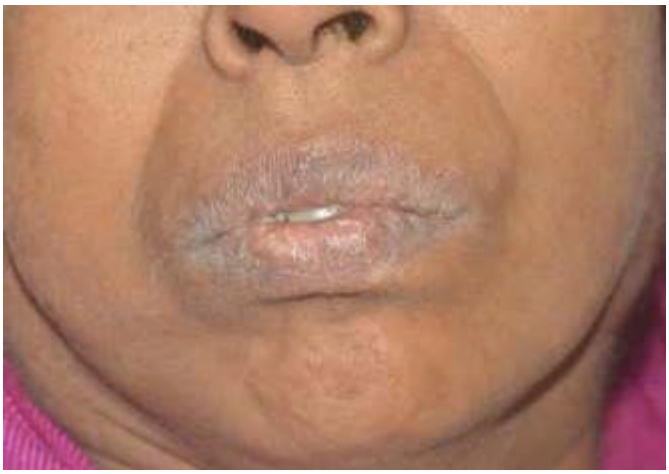
Fig 4: Cracked lips

Discussion

Sjogren's syndrome (SS) is an autoimmune disease characterised by xerophthalmia, xerostomia, arthralgia, myalgia and severe fatigue. Lymphocytic infiltration of the lachrymal and salivary glands is the hallmark of the disease and results in partial destruction of the gland parenchyma. Disease classification is based on the new preliminary American College of Rheumatology (ACR) criteria as given [2]

- Positive Serum anti SSA(Ro) and or anti SSB (La) or positive rheumatoid factor & ANA ≥ 1.320)
- Labial salivary gland biopsy exhibiting focal lymphocytic sialadenitis with a focus score ≥ 1 focus/4mm^2
- Keratoconjuctivitis sicca with ocular staining score ≥ 3

Prior diagnosis of any of the condition would exclude participation in Sjogren's syndrome studies or therapeutic trials because of overlapping clinical features or interference with criteria tests:

- History of head and neck treatment
- Sarcoidosis
- Amyloidosis
- Graft versus host disease
- Hepatitis C infection
- Acquired immunodeficiency syndrome
- IgE related disease

The autoantibodies and salivary gland biopsy was positive in this particular patient along with keratoconjuctivitis sicca substantiating for the diagnosis of Sjogren syndrome. Along with these features, patient also had neurologic disorder, mononeuritis multiplex.

Mononeuritis multiplex is a painful, asymmetrical, asynchronous sensory and motor peripheral neuropathy involving isolated damage to at least 2 separate nerve areas. Multiple nerves in random areas of the body can be affected. As the condition worsens, it becomes less multifocal and more symmetrical. Mononeuropathy multiplex syndromes can be distributed bilaterally, distally, and proximally throughout the body.[3] Conditions were mononeuritis multiplex can be seen are described here.

Conditions with Mononeuritis Multiplex

- Leprosy
- Diabetes mellitus
- Connective tissue disorders
- Sarcoidosis
- Primary systemic vasculitis
- Paraneoplastic syndromes

Causes of Recurrent Parotid Abscess

- Xerostomia (dehydration, xerostomic drugs)
- Ductal obstruction (sialolith, salivary strictures, foreign bodies)
- Duct compression by tumour
- Injury to duct and papilla
- Sjogren syndrome
- Diabtes mellitus
- Immunosuppression
- Lipoid proteinosis
- Post irradiation patients
- Trauma

Xerostomia is pathognomic sign in Sjogrens syndrome. This results in a permanently reduced salivary flow. Antimicrobial activity of saliva will be reduced because of that. Anatomic location of parotid gland increases the susceptibility of microorganisms to cause ascending infection. Recurrent parotid abscess. A study by Wang et al[10] revealed that in 12 of the 22 patients initially diagnosed as having Chronic recurrent parotitis, symptoms of Sjogren Syndrome developed later. Ten of these 12 patients had primary Sjogren Syndrome and two had secondary. According to that study, 10% of Sjogren syndrome patients have recurrent bacterial parotitis.

Conclusion

CNS involvement is an uncommon complication of primary Sjogren syndrome. Pathogenesis of CNS involvement in primary Sjogren syndrome remains unclear. Early diagnosis of neurological manifestations in Sjogren Syndrome improve the quality of life of the patient considerably.

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