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CASE REPORT

SJOGRENS SYNDROME- A BIZARRE CASE REPORT

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ABSTRACT

Sjogren's syndrome commonly known as "Sicca Syndrome" is another representative of long lasting chronic auto-immune disorder primarily affecting exocrine glands such as salivary and lacrimal gland which usually manifest development of lymphocyte infiltration primarily causing destruction of exocrine glands. This is a case of 42 years patient with complaints of difficulty in speaking, development of hoarseness, difficulty in eating, itching of eyes since1 month. Physical Examination revealed tropic ulcers since 2 days, splenomegaly. Ocular signs were confirmed by Schirmer test which revealed <5mm/wetting after 5 minutes. Laboratory investigations revealed detection of anti SS-B antibodies by counter immunoelectrophoresis and Western blotting. Patient was treated with carboxy methyl cellulose-TID, and XyliMelts –BD, lubriderm ointment, amoxicillin and clavulanic acid -625 mg and Prednisolone -5 mg. The main purpose of the author is to make the people well known about the medical field, well acquainted about this disease and its affiliated complications.

INTRODUCTION

Sjogren's syndrome commonly known as "Sicca Syndrome" is another representative of long lasting chronic auto-immune disorder which primarily Affects exocrine glands such as salivary and lacrimal gland which usually manifest development of lymphocyte infiltration primarily causing destruction of exocrine glands[1-2]. Sjogren's syndrome classified into two types primary and secondary, primary form involves dryness of eyes and mouth with or without involvement of connective tissue disease ,on other hand secondary forms includes involvement of connective tissue diseases like SLE, scleroderma, Rheumatoid arthritis[3].

Clnical Manifestations

Sjogren's syndrome manifest as lymphocytic infiltration which results in "Sicca complex"-combination of Dry eyes (keratoconjunctivitis sicca) and Dry mouth (xerostoma)[4]. Initial manifestations will be mild but may progress later which makes difficult to recognise syndrome, its manifestations include-

Glandular manifestations:-Diminished tear production due to destruction of both corneal and bulbular conjuctival epithelium termed as keratoconjuctivitis sicca (KCS), Xerostomia or dry mouth, parotid-major salivary gland enlargement occurs in 60%patients along with dryness of upper respiratory tract and dermal dryness.

Extra glandular manifestations: Include periepithelial organ involvement (arthritis, nephritis) and extra epithelial manifestations (palpable purpura).

Others: Gastrointestinal and hepatobiliary features, Raynaud phenomenon, Renal involvement, vasculitis, cutaneous involvement[5].

Etiopathogenesis

Immune mechanisms have implicated in etiopathogenesis of lesions in SS in which antinuclear antibodies its were about 90% and Rheumatoid factors about 25% cases[6]. Some human leukocyte antigens also increases the risk along with multiple Auto antibodies particularly SS-A and SS-B, Viral infections can also be the leading cause of Sjogrens syndrome [7].

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Diagnosis

SS is more prevalent in women than in men affects 0.5–3% of population [1]. Numerous criteria were proposed for diagnosis of Sjögren syndrome in which , Most widely accepted are American and European group developed international classification criteria for Sjögrens syndrome which include six criteria such as Ocular symptoms for not < 3 months, dry months not<3months, recurrent swelling of salivary glands, persistently need to drink water, Ocular signs by schirmers test. Diagnosis also includes positive histopathology of salivary glands and detection of Antibodies to anti-SS-A, anti SS-B antigens. Also this classification requires four of the six criteria, one of which must be positive-biopsy of minor salivary gland or antibody test [8].

Management

No specific disease modifying treatment has been identified only symptomatic treatment given such as lachrymal substituent in combination with viscous lubricating ointment for dry eyes, Artificial Saliva and oral gels for Xerostoma and stimulation of salivary flow by sugar free chewing gums may be helpful. Extraglandular and musculoskeletal manifestations can be treated by Steroids. If Lymphadenopathy and salivary gland enlargement develops, then Biopsy should be performed to exclude malignancy [9].

Case Report

A 42 years old female patient came to SVS Medical College with chief complaints of difficulty speaking for longer periods and development of hoarseness and also patient was feeling difficulty in eating, and itching of eyes also observed since1 month. Along with this patient is also experiencing generalized weakness of upper and lower limbs. Physical Examination revealed tropic ulcers since 2 days, splenomegaly also present. No other risk factors such as diabetes, smoking, contraceptive drug use, or family or personal history of thromboses were present.

Parameters	DAY 1	DAY 2	DAY 3	DAY4	DAY 5	DAY 6	DAY 7
BP(mmHg)	120/90	120/90	100/80	120/80	120/70	100/80	120/70
Pulse bpm	120	96	88	98	91	88	92

Past Medical History

Patient past history revealed Chronic Liver Disease (on Medication with Spiranolactone-100mg TID), Portal HTN with Oesophageal Varices, (on Medication with propanolol-5mg) Patient was known case of Iletis (Biopsy proven). Patient has no signs and symptoms of salivary gland enlargement.

Lab Investigations

Apart from above mentioned physical Examination, patient underwent Routine Screening tests as well as Definitive Tests

Routine Test

Complete Blood Picture was done which revealed decrease Hb-8.5gm/dl and RBC count also decreased to- 2.7 ml/cumm,Also total circulating WBC count decreased to 3500/ mcL, platelet count also reduced to- 55,000/mcL indicating anaemia, leukopenia and thrombocytopenia which increases the chance of diagnosis. Apart From CBP other test such as Liver function test was also performed to rule out HEPATTIS which revealed-increased Alkaline Phosphate- 592 U/L and Serum creatinine - 3.6 mg/dl. Electrolytes revealed decrease Potassium and calcium levels of 2.2 mEq/L and 0.57 mmol/L respectively.

Definitive Tests

Ocular signs were confirmed by Schirmer test which revealed <5mm/wetting after 5 minutes. Interpretation of sjogrens syndrome was confirmed by laboratory investigations which revealed detection of anti SS-B antibodies by counter immunoelectrophoresis and Western blotting.

Treatment

On final confirmation of diagnosis patient was treated with carboxy methyl cellulose tear drops-TID, and XyliMelts –oral adhering discs-BD for dry eyes and mouth given for symptomatic relief, along with lubriderm ointment for skin. Antbiotics such as amoxicillin and clavulanic acid -625 mg given to treat infections, Prednisolone -5 mg to treat systemic manifestations given for 5 days

Discharge medication

On discharge patient was asked to continue above mentioned medications such as Pilocarpine -5mg TID, XyliMelts, and Lubriderm ointment for dry eyes, mouth and skin respectively.

DISCUSSION

Sjogrens Syndrome is the most common systemic chronic autoimmune disease [10]. Characterized by traid of xerostomia, xerophthalmia, and lymphocytic infiltration of the exocrine glands[11]. Clinical features of SS given in Table 1 [9].Few patients have been described with PNS, CNS symptoms, in which PNS complications are common than CNS (25%)[12]. SS diagnosis is not straight forward as it is a multifactorial disease which requires good command and collaboration between multiple specialties [13]. The treatment of Sjögren's syndrome is to give symptomatic relief of the effects of chronic xerostomia and keratoconjunctivitis, which is achieved by keeping the mucosal surfaces moist. Dry eyes need artificial tears. Hydromellose (Hydroxyethylcellulose) helps to replace aqueous layer. Drugs like anti-hypertensives, diuretics, anti-depressants. decongestants can cause decrease in lacrymation and salivation and should be avoided. Bromhexine (48 mg/day) Pilocarpine hydrochloride (5 mg three times daily) may help sicca symptoms. Lubricant jellies are used to treat vaginal dryness. Dry skin is treated with moisturizing creams. Parotid gland infection is treated with tetracycline. (500 mg four times a day), Corticosteroids (0.5 - 1.0 mg/kg/day) given forpatients with severe extra glandular diseases. [14].

Risk factors :Age of onset 40-60, Female>male
Common clinical features:-Keratoconjuctivits sicca, xerostomia, salivary gland enlargement, Fatigue, Raynaud phenomenon Less common features: Low grade fever, Anaemia, Leucopenia, Thrombocytopenia, vasculitis, lung disease, gloumerulonephritis, cryoglobunemia, lymphadenopathy Antibodies frequently detected: RF,ANA, SS-A, SS-B, Thyroid

In present case patient was treated with carboxy methyl cellulose tear drops-TID, and XyliMelts-oral adhering discs-

BD for dry eyes and mouth given for symptomatic relief, along with lubriderm ointment for skin. Antbiotics such as amoxicillin and clavulanic acid -625 mg given to treat infections, Prednisolone -5 mg to treat systemic manifestations given for 5 days. Same drugs were prescribed after discharge also.

CONCLUSION

Sjogren's syndrome is slowly developing in its course. The risk of lymphomas and mortality of this disease is very high. Diagnosis of this disease in early stages is imperative so that all oral complications can be minimized. Proper management requires cooperation among all health professionals and all the professionals should be fully aware of all symptoms and management of these patients. The main purpose of the author is to make the people well known about the medical field, well acquainted about this disease and its affiliated complications.

References

1. Marcos de Mendonça Invernici *et.al*, Management of Sjogren's Syndrome Patient: Case Reports in Dentistry, Volume 2014, Article ID 761251, 5 pages.
2. Kaushalendra Tripathi *et.al*, Diplopia and Sjogren's disease: A rare case report *Journal of Neuroimmunology* 302 (2017) 7-9.
3. Dellafiore *et.al*, Sjo¨gren syndrome: A case report, Journal of Ultrasound (2012) 15, 108-110.
4. Kunduru Buchi Reddy *et.al*, Sjögren's Syndrome-Vasculitis: Azathioprine Induced Myelosuppression-A Case Report, *Indian Journal of Pharmacy Practice*, Vol 9, Issue 3, Jul-Sep, 2016.
5. Pasaramelli karteek *et.al*, Sjogrens syndrome: A review of clinical features, diagnosis and treatment available, *International Journal of Pharmaceutical Sciences Review and Research*, volume 5, issue1,November-December 2010; Article-014,pg-93-98.
6. Harsh mohan, textbook of pathophysiology, sixth edition, jaypee brothers medical publishers, page no: 81.
7. M Öğütcen-Tolle *et.al*. Sjögren's Syndrome: A Case Report and Review of the Literature, West Indian Med J 2011; 60 (3): 305.
8. Santosh Jadhav *et.al,* Sjögren's Syndrome: A Case Study, *Journal of International Oral Health 2015; 7(3):72-74.*
9. Brain R.walke, Nicki R.colledge, Staurt H.ralston, Lan D.Penman Davidson's principles and practice of medicine,22nd edition, UK, CHURCHIL LIVINGSTONE,1114.
10. Paul ruszka *et.al,* Dagnosis and management of sjogrens syndrome, American Famly Physcian, volume 79,number 6,March 15,2009 pg 465-470.
11. Sapna Raut Dessai *et.al*, Primary Sjogren's Syndrome: A Case Repor**t,**Journal of Oral Health Research, Volume 7, Issue 1, 2016.pg 22-26.
12. M. Bragoni et.al Sjogren's Syndrome Presenting as Ischemic Stroke, Stroke Vol 25, No 11 November 1994 pg 2276-2278.
13. Baceam AE Ottoman and Rania A.Ali, A Rare Case Report of Sjogrens Syndrome in a Young Female with Clnicosonographical Characterization and Literature review, *American Journal of Ultrasound* (2015) vol 2 No1 pp, 1-8.
14. KM Mahendranath, sjogren's syndrome - diagnosis and management, suppliment to japijune 2006 vol. 54, pg 54-57.

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