Primary Sjögren syndrome with pulmonary alveolar hemorrhage and other extra glandular manifestations- A case report

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ABSTRACT

Sjögren syndrome (SS) is an under-diagnosed complex autoimmune disease with a multi factorial pathogenesis. It is classified as primary or secondary depending on the co-existence of other rheumatic disease. It predominantly affects the exocrine glands with the non-exocrine involvement termed as extra-glandular manifestations. The most common clinical presentation of primary SS is of fatigue and depression. The respiratory involvement in this disease is varied. However, pulmonary alveolar haemorrhage in a case of primary SS is rare with only one reported case in literature. We present a case of a pulmonary alveolar haemorrhage in a patient of Sjögren syndrome with extensive multi system involvement. In view of the varied clinical and laboratory manifestations, it is important for physicians to be aware of this disease and the significance of extra-glandular involvement.

Key words: Primary Sjögren, alveolar haemorrhage, extra- glandular

INTRODUCTION

An estimated 0.4 to 3.1 million adults are affected by primary Sjögren syndrome (SS) in USA alone.[1] In India, the incidence of reported cases is about 0.0045 percent.[2] This may be due to this disease being misdiagnosed, unreported, or due to the lack of awareness of this disease. This complex autoimmune disease has a multifactorial pathogenesis. The criteria for the diagnosis of Sjögren syndrome and further classification into primary and secondary Sjögren syndrome is based on the revised international classification criteria by the American-European consensus group in 2002.[3]

To best of my knowledge the combination of primary Sjögren syndrome with pulmonary alveolar haemorrhage, cutaneous small vessel vasculitis, polyclonal hypergammaglobulinemia, renal dysfunction, hypothyroidism, vitiligo, arthralgia, depression and peripheral sensory motor neuropathy has not been previously reported in literature.

CASE REPORT

A 64 year old female woman presented with a history of low grade fever, dry mouth, arthralgia, tingling and numbness over the lower limbs since past 2 months. She was on treatment for hypothyroidism since last 4 years. Clinical examination revealed pallor, dry mouth, vitiligo and absent ankle reflexes. She also had features of depression. There was no proximal muscle weakness. Other systemic examinations were within normal limits. The Shirmers test showed moderate dry eyes bilaterally.

Investigations revealed elevated ESR of 140mm with an elevated anti SSA, anti SSB and ANA 2+ positive. She also had hypergammaglobulinemia with the serum electrophoresis showing a polyclonal pattern. The biopsy of the lower lip was suggestive of Sjögren syndrome. The bone marrow biopsy revealed a cellular marrow. The EMG/NCV showed asymmetric sensory motor polyneuropathy and skin biopsy was suggestive of small vessel vasculitis. She also had an elevated serum creatinine of 2.3mg/dl; and urine microscopy showed numerous RBCs. The work up for other autoimmune disease was negative.

She was diagnosed to have primary Sjögren syndrome with extra-glandular manifestations of cutaneous small vessel vasculitis, polyclonal hypergammaglobulinemia, renal dysfunction, hypothyroidism, vitiligo, arthralgia, depression and peripheral sensory motor neuropathy. She was started prednisolone and mycophenolate mofetil.
She presented two weeks later with sudden onset

breathlessness, hemoptysis, a drop in hemoglobin and a positive direct Coombs test. CT scan of chest revealed pulmonary haemorrhage. She was pulsed with methylprednisolone for three days, and discharged with oral steroids.

DISCUSSION

Sjögren syndrome is often underdiagnosed due to lack of awareness. To improve and assist in accurate diagnosis of this condition, the American–European consensus group in 2002 developed the revised international classification criteria.[3] It also helps to differentiate primary from secondary SS on basis of the presence of another connective tissue disorder.

In patients with SS, serology of ANA, anti-SSA and anti-SSB are found to be positive in 74, 40 and 26 percent of the patients respectively.[4] Anti SSA and SSB positivity has also been shown to be a risk factor for development of lymphoma.[4,5] The skin changes of vitiligo and cutaneous small vessel vasculitis has been described in literature.[6] Cutaneous vasculitis is seen in 10% of patients with SS and this subgroup has an increased risk of lymphoma and death due to disease related complications.[7] Hypergammaglobulinemia has been seen in 22% of the patients with SS.[5] This contributes to increase in ESR in the absence of infection, vasculitis or other causes.[5] Studies showed that arthralgia is present in 50% of patients of SS with or without arthritis.[8] In a study which evaluated 506 patients of primary SS; hypothyroidism was seen in 17 % and hyperthyroidism in 6%.[9] The incidence of thyroid dysfunction was higher in patients with a positive Anti-SSA and SSB.[9] Sensory neuropathy is seen in 20% of the patients with SS and the presentation is similar to that of our patient.[7] Renal involvement in the form of glomerular disease is also seen in patients with SS; however the etiology for renal dysfunction is proposed to be multifactorial. Studies show that primary SS with cutaneous vasculitis, peripheral neuropathy, rheumatoid factor positivity, type 2 cryoglobulinemia is at an increased risk of development of lymphoma.[5] The respiratory system involvement in SS can vary from upper respiratory symptoms to cough. There has been only one previously published case report of pulmonary alveolar haemorrhage in primary SS.[10]

CONCLUSION

This case is of importance as it shows the varied manifestations of this disease. It is important for clinicians to consider Sjögren Syndrome as a possibility in cases of multisystem involvement.

AUTHOR NOTE

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