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Sjogren's Syndrome in a Patient with Maculopapular Cutaneous Mastocytosis

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Abstract

We report a case of a 42-year-old female with past medical history significant for cutaneous mastocytosis who presented with complains of progressive dry eyes, and dry mouth for the past few weeks. Her vital signs were remarkable for blood pressure of 106/71 mmHg, pulse of 86 beats per minute, temperature of 36.7°C (98.1°F) (oral), and respiratory rate of 16 breaths per minute. Physical exam was significant for bilateral dryness in eyes, cracks at the corners of the mouth, dry and smooth tongue. Lab work revealed a benign blood count and chemistry. Sedimentation rate was elevated at 25 mm/hr, CRP was also elevated at 11.1 mg/L, and ANA with speckled pattern was elevated at 1250 1/dil. Speckled pattern was elevated at 1250 1/dil. Hence patient was managed with muscarinic agonists such as cevimeline and pilocarpine, which improved her symptoms. We report a rare case of a patient with maculopapular cutaneous mastocytosis who develops Sjogren's syndrome. Literature review revealed that in the past there have been case reports which linked systemic mastocytosis to Sjogren's syndrome, however there are no case reports of maculopapular cutaneous mastocytosis linking to Sjogren's syndrome. Clinicians need to be aware that even the milder form of mastocytosis, cutaneous type, can also be associated with Sjogren's syndrome. Absence of well defined histopathological features and lack of clinical awareness can delay its diagnosis and treatment.

Keywords: Sjogren's syndrome; Mastocytosis; Mast cells

Case Report

Mastocytosis is the accumulation of mast cells in one or more tissues, and it can be broken down into two varieties, cutaneous and systemic. Cutaneous mastocytosis is limited to the skin while systemic mastocytosis involves at least one other organ system. The incidence of mastocytosis is 5-10 cases per million. There are many variants of cutaneous mastocytosis, including the nodular type, maculopapular type, diffuse type and telangiectasia macularis eruptiva perstans. Symptoms vary, but

approximately 80% of patients present with urticaria pigmentosa, described as flat or slightly elevated brownish-red papules that urticate when exposed to mechanical trauma [1,2]. Other symptoms of disease may include pruritus, flushing, syncope, nausea, vomiting, diarrhea, angioedema, depression, and diffuse bone pain. These symptoms can be attributed to the release of mediators such as histamine, and leukotrienes from mast cells. An increased number of mast cells could also cause elevated IL-6, which is commonly seen in patients with Sjogren's syndrome, an autoimmune inflammatory disorder characterized by lack of lacrimal and salivary gland function resulting in eye and mouth dryness [3,4]. We present the first reported case of a patient with maculopapular cutaneous mastocytosis who develops Sjogren's syndrome.

Our patient is a 42-year-old female with past medical history significant for cutaneous mastocytosis who presented with complaints of progressive dry eyes and dry mouth for the past few weeks. Her associated symptoms included progressive fatigue and diffuse joint pain. Her vital signs were unremarkable with a blood pressure of 106/71 mmHg, pulse of 86 beats per minute, temperature of 36.7°C (98.1°F) (oral), and respiratory rate of 16 breaths per minute. Physical exam was significant for bilateral dryness in eyes, cracks at the corners of the mouth, and dry, smooth tongue. Lab work revealed a benign blood count and chemistry. Her sedimentation rate and CRP were elevated at 25 mm/hr and 11.1 mg/L respectively, and ANA with speckled pattern was elevated at 1250 1/dil. She had a negative work up for CCP antibody, anti-centromere antibody, rheumatoid factor, SCL-70 autoantibody, and anti-smooth muscle antibody. The patient's SSA autoantibody was elevated at 229 [AU]/mL but the SSB autoantibody levels were within normal range. After receiving the diagnosis of Sjogren's syndrome, the patient was managed with muscarinic agonists, such as cevimeline and pilocarpine, which improved her symptoms.

This is a rare case of a patient with maculopapular cutaneous mastocytosis who subsequently developed Sjogren's syndrome. Our patient presented to us with cutaneous mastocytosis of the maculopapular type, and after initial presentation a bone biopsy was done to rule out systemic mastocytosis. Due to the oral and

ocular symptoms in setting of elevated SSA antibody our patient qualified for a diagnosis of Sjogren's syndrome. Worldwide incidence and prevalence of primary Sjogren's syndrome are estimated at 7 and 43 per 100,000 respectively. Diagnosis once centered upon salivary gland biopsy showing focal lymphocytic sialoadenitis, but this has changed over the years to a more clinical approach. Now, American-European Consensus Group Criteria centers diagnosis of Sjogren's syndrome around the presence of ocular and oral dryness, with no other secondary cause, in conjunction with high SS-A/SS-B antibody titers. The new criteria have allowed earlier diagnosis and treatment for affected individuals [5]. Literature review revealed that in the past there have been case reports which linked systemic mastocytosis to Sjogren's syndrome, however there are no case reports linking maculopapular cutaneous mastocytosis to Sjogren's syndrome [3,6]. An absence of well-defined histopathological features and a lack of clinical awareness can delay its diagnosis and treatment, clinicians need to be aware that even the milder form of mastocytosis, cutaneous type, can also be associated with Sjogren's syndrome.

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