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Progressive Cavus Foot Type Secondary to Sjogren's Syndrome: A Case Report

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Abstract: In this case report, we review a case where a patient was suffering from lateral ankle pain, neuropathic symptoms, and multiple other musculoskeletal/dermatological problems for years. The patient's illnesses were repeatedly misdiagnosed over the years due to normal serological labs. Subsequently, this led to the mismanagement of the patient and her symptoms. It took an ambitious Neurologist to discover the primary diagnosis. Then with collaboration of Rheumatology and Podiatry, an explanation for the patient's secondary manifestations was revealed. The purpose of this report is to review a rare case and to further understand the different causes of lateral ankle pain and cavus foot.

Key words: Sjogren's Syndrome, Cavus, Small Fiber Neuropathy

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Sjogren's syndrome (SS) is a systemic autoimmune disease that is characterized by glandular involvement including xerophthalmia, xerostomia, cutaneous xerosis and possible extraglandular manifestations. Among the extraglandular manifestations occurring in SS, peripheral neuropathy is frequent with a prevalence close to 20% [1]. Small fiber neuropathy (SFN) is a type of peripheral neuropathy that occurs from damage to the small unmyelinated peripheral nerve fibers, usually C-fibers. SFN is particularly common among patients with diabetes, impaired glucose tolerance, and connective tissue disease. Farhad et al reported that "idiopathic" neuropathy was present in patients that were found to have SFN. In the study, a rheumatologic condition was often the underlying cause of neuropathy [2]. Studies examining SFN found vascular and perivascular inflammatory infiltrates with and without necrosis observed in peripheral nerve biopsy specimens. They predicted that neurons could be affected secondary to an inflammatory process involving the vasa nervorum.

Pes cavus foot deformity is a neuromuscular disease that is thought to be related to an imbalance of musculature around the foot and ankle. This condition was first described in the 1880's by Shaffer [3]. Pes cavus can often signal an underlying neurological disorder, including spinal cord and peripheral nerve pathologies, such as spinocerebellar ataxia and hereditary peripheral neuropathies [4]. Two-thirds of patients with pes cavus have an underlying neuromuscular disorder and 50% of these are attributable to Charcot- Marie-Tooth disease (CMT) [5]. CMT is the most common cause of hereditary peripheral neuropathy. Neuropathy is usually associated with a pes cavus foot deformity due to the neurological etiology but this deformity has yet to be linked to SFN or SS. In this case report, a patient with SFN and a progressive cavus foot deformity was found to have an unknown diagnosis of SS.

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Case Report

44-year-old Caucasian female with past medical history of fibromyalgia, cervical pain, and depression presented with left lateral ankle pain for the past 5 years. Patient admitted to ankle weakness, but denied her ankles "giving out". Past treatment included ankle-foot orthosis for 3 years, prednisone, cast boot, injections to her left ankle, and physical therapy with limited success. Patient has a family history of systemic lupus erythematosus and mixed connective tissue disease. Patient does follow with Rheumatology. On review of systems, patient admitted to joint pain and swelling, back pain, muscle pain, and bruising easily.

On physical exam, neurovascular status was within normal limits. No ankle instability was noted. Structurally, the patient had a cavus foot type, with the left being more severe than right. (Fig.1). Pain was elicited with passive and resisted eversion and with palpation to the peroneal tendons of the left foot. Radiographs showed all components of a cavus foot with no acute changes (Fig. 2). Patient was diagnosed with peroneal tendonitis and pes cavus. Treatment plan included weight bearing in a cast boot for 3 weeks then transitioning to an ankle brace. It was recommended that the patient follow up with Rheumatology. Due to the patient's symptoms, past medical history and family history, there was high suspicion for autoimmune disease.

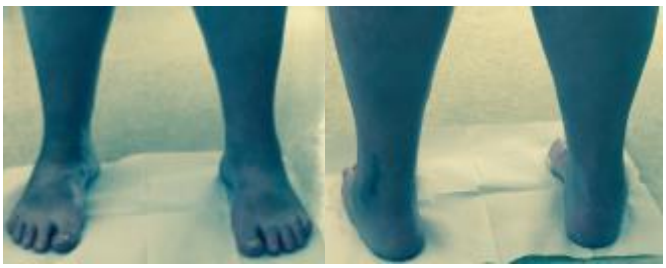

Figure 1. Clinical pictures of patient with cavus foot type

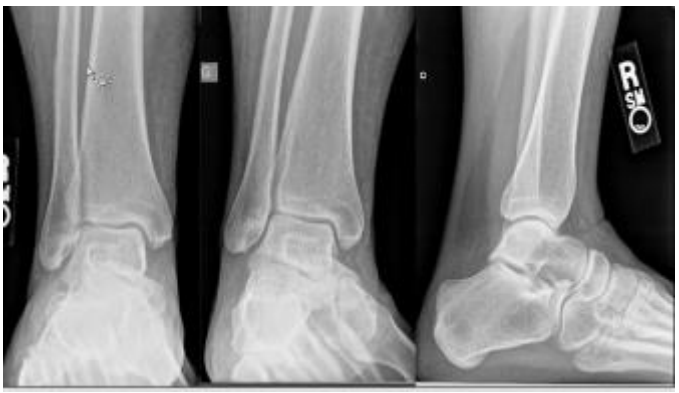

Fig. 2. Radiographs of patient with findings of supination

At her 1.5 month follow up, the patient complained of contralateral ankle pain. She stated that right ankle pain was like her left ankle pain. Patient claimed that she followed the recommended treatment plan for her left ankle with minimal relief. Patient did follow up with Rheumatology who ordered serological labs to rule out autoimmune diseases, however, all labs were negative (Table.1). Per records, patient's labs have been negative for years. Conversely, the patient did acknowledge that her pes cavus foot deformity was progressively getting worse over the years. With this new information, the patient was referred to Neurology to rule out CMT disease.

During the patient's visit with Neurology, several key discoveries were revealed. The patient admitted to developing neuropathic symptoms, having a dry mouth and constant fatigue. Based off the patient's symptoms, family history of autoimmune diseases, no family history of neuropathy, and tendon issues/injury for 5 years, the neurologist believed that a lower lip and sural nerve biopsy would confirm the proper diagnosis.

The sural nerve biopsy (Fig. 3) highlighted minimal loss of myelinated axons and decreased intra-epidermal nerve fiber density consistent with SFN. The lower lip biopsy (Fig. 4) showed minor salivary gland tissue with mutlifocal chronic inflammation, periductular lymphocytic infiltrate, acinar atrophy, fibrosis, hyalinization, and fatty replacement of the gland, which was consistent with SS. For further confirmation, the patient was refereed to ophthalmology and a Schirmer's test was performed and confirmed eye dryness.

Lab Component	Value	Range & Units
Rheumatoid Factor	<7	<20IU/mL
Sm Antibody	<0.2	<1.0AI
RNP Antibody	<0.2	<1.0AI
SSA Antibody	<0.2	<1.0AI
SSB Antibody	<0.2	<1.0AI
Centromere Ab	<0.2	<1.0AI
Scleroderma Ab, IgG	<0.2	<1.0AI
Jo 1 Antibody	<0.2	<1.0AI
Chromatin Antibody	<0.2	<1.0AI
ANA FIA	1.4	<1.5 OD Ratio
CRP	0.5	0.0-1.0mg/dL
WSR	6	0-15mm/hr

Table 1. Serologic labs to rule out autoimmune diseases

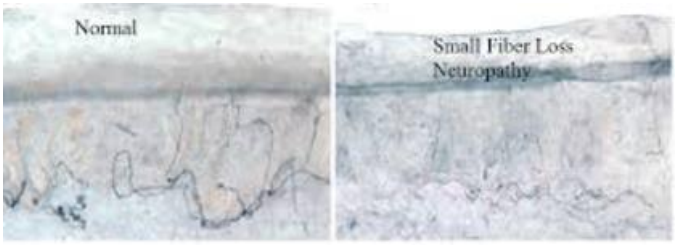

Fig. 3. Sural nerve biopsy comparing a normal nerve to small fiber neuropathy (These pictures are representative of our case)

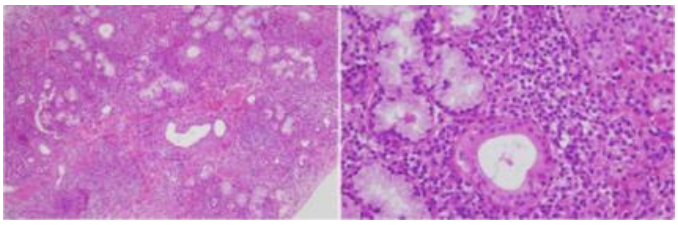

Fig. 4. Lower lip biopsy comparing normal salivary gland tissue to abnormal tissue seen in Sjogren's syndrome (These pictures are representative of our case)

For the past 5 years, the patient had dry eyes that she described as feeling like sand in her eyes and a dry mouth that had her "constantly sipping on water." She was also suffering from pain in her shoulders, hands, hips, knees, and ankles. Rheumatology confirmed that the patient met the criteria for Sjogren's syndrome based on oral/ocular sicca, positive Schirmer's and positive minor salivary gland biopsy. New blood work was ordered to compare to previous serological labs and they came back negative again.

After Podiatry communicated with Neurology and Rheumatology, it was discovered that SS caused the SFN resulting in muscle imbalance, specifically weakening of the peroneal muscles. Impairment of the peroneals, resulted in increased supination causing the pes cavus foot deformity.

Discussion

SS is a chronic autoimmune inflammatory disorder characterized by diminished lacrimal and salivary gland function which results in dryness of the eyes and mouth (sicca syndrome). In addition, a variety of other disease manifestations affecting organ systems may occur. SS is associated with rheumatoid arthritis and systemic lupus erythematosus. SS is most common in women in their fifties and sixties but can affect adolescents and young adults, as well as men. Overall incidence of SS is 7 per 100,000 people with most reported studies in Europe and Asia [6]. Other symptoms include fatigue, myalgia, and mild cognitive dysfunction. It is clinically difficult to distinguish from fibromyalgia and/or depression.

The diagnosis of SS should be suspected in individuals with persistent symptoms of dry eyes, dry mouth, parotid gland enlargement, unexplained increase in dental caries, and abnormal results of specific serologic tests. There is no single diagnostic test for SS. Thus, the clinical diagnosis of SS is made in the presence of compatible clinical and laboratory features and after the exclusion of other diseases. SS should not solely be diagnosed upon the presence of antibodies because they can occur in other rheumatic/connective diseases, as well as in normal individuals.

SFN is found in 9-30% of SS pts [1,7]. The neurological examination and the electroneuromyogram in these patients are usually normal. The diagnosis of SFN can be confirmed by the evidence of decreased intra-epidermal nerve fiber density after a skin punch biopsy or the presence of abnormal nonconventional neurophysiological tests exploring the A-delta and C small nerve fibers. However, there is little to no evidence linking pes cavus foot type to SFN or SS. Therefore, this is a rare case that should be taken into consideration when treating patient with similar symptoms.

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