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Case Study

The Zebra of Sjogren's Syndrome: A Case Study

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Introduction

Sjogren's Syndrome (SS) is an autoimmune disease that is often either misdiagnosed or diagnosed late. One of the reasons why Sjorgen's syndrome is susceptible to a late diagnosis is because its symptoms can be very subtle. In fact, it may, sometimes take upward of 6 years or more to correctly diagnose [1]. The typical patient would present with complaints of chronic dry eyes, dry mouth, swollen parotid glands and possibly chronic fatigue [2]. This case study discusses the presentation of a patient to the clinic with varying complaints not responding to treatments. By utilizing out-of-the-box thinking, and employing a set of diagnostic tests, the patient was able to receive an expedited diagnosis of primary Sjogren's syndrome.

Presenting Signs and Symptoms

The patient is a 35-year-old mixed race female who presented to the clinic with continued complaints of pruritus, severe itching and a rash that was spreading and not responding to treatments. The patient was first seen in the clinic May 2016 with complaint of pruritus and rash to her bilateral thighs and lower back. The provider documented that on exam the rash was described as scattered annular lesions. It was also noted that the patient's right tympanic membrane had cloudy fluid and she had mild erythema to the pharynx. The patient was diagnosed with Tinea versicolor versus Pityriasis rosea. She was prescribed Augmentin for the right ear, Diflucan, Selenium sulfate lotion and Triamcinolone 1% ointment for the rash. The patient returned to the clinic 11 days later with continued complaint of pruritus and the rash spreading. She stated she followed the previously prescribed regiment without relief. The provider that assessed her on the visit documented that the patient had generalized hives to her torso, arms, legs and annular scaly pruritic lesions to her buttocks. She was diagnosed with generalized rash. She was prescribed Prednisone 10 mg dose pack, Lamisil and Benadryl.

The patient returned 13 days later with the same complaint and stated the rash was spreading. The provider that examined her noted that she had wide spread round dry patches on her trunk and buttocks. She was diagnosed with fungal rash and prescribed Diflucan and Tolfinate. I saw her 10 days later and she had the same complaint of the spreading pruritic rash that was not responding to any of the previous medications and she had developed foul smelling yellowish brown fluid leaking from both her ears. On exam, her left ear canal was swollen with thick yellowish-brown exudate, and the swelling occluded the visualization of the tympanic membrane. The right ear canal was partially swollen with the same characteristic exudate, and the partially visualized tympanic membrane had diminished light reflex and was a dull gray. The pharynx had erythema with no exudate.

The skin was noted to have large raised erythema macular patches with some drying on the bilateral buttocks, thighs to medial lower legs. The rashes were noted to symmetrical on both thighs and calves. The rash noted on arms and torso was erythematous macular with no central clearing; however, inner regions of macular patches had hypopigmentation. The patient was diagnosed with nummular eczema, and otitis externa. She was prescribed Triaminacolen ointment, Solu-Medrol injection with a prednisone dose pack. She was also given Neosporin otic and metrodianzole oral for the ear. Due to the concern of persisting symptoms, which was now accompanied by a possible fungal otitis externa, the case was discussed with the residing physician. It was mutually decided that other disease processes such as Diabetes, HIV and the possibility of an autoimmune disorder needed to be ruled out. Therefore, a battery of lab tests was ordered, which consisted of a culture of the fluid from the ear, Complete Blood Count (CBC), Complete Metabolic Panel (CMP), Systemic Lupus Erythematosus (SLE) Comprehensive Diagnostic Panel, RPR with Reflex Titer, HIV 1/2 Antigen/Antibody 4th Generation and a Hemoglobin A1c. The patient was informed that she would be notified when the

results were back and she was instructed to return to the clinic for the lab results. When the patient returned she reported that the current regiment was starting to ease the pruritus and the rash had stopped spreading.

Laboratory Results

EArobic Bacteria Culture, Ear	Heavy growth of Pseudomonas EAruginosa	
Complete Blood Count (CBC)		
Red Blood Cell Count	3.74 L	3.8 – 5.10 Million/uL
Hemoglobin	10.9 L	11.7 – 15.5 g/dL
MCHC	30.2 32.0 – 36.0 g/dL	
Comprehensive Metabolic Panel (CMP)	Normal	
Systemic Lupus Erythematous (SLE)	Negative	
Sjogren's Antibody (SS-A)	5.0 POS<1.0 NEG AI	
Sjorgen's Antibody (SS-B)	Negative	
RPR w/ Reflex	Non-Reactive	
HIVAG/AB 4th GEN	Non-Reactive	
Hemoglobin A1C	5.1<5.7 % of total Hgb	

Discussion

The extraglandular involvement of the cutaneous and xeroderma is a frequent finding for SS. The spectrum of cutaneous presentations is wide and can range from nonvasculitic cutaneous to vasculitis [2]. It has been reported that skin manifestations are described in over half of SS patients; although, they are typically on a lesser scale than the oral, ocular and musculoskeletal manifestations. SS cutaneous most often manifests as xerosis, pruritus, eyelid dermatitis, angular cheilitis, cutaneous vasculitis or erythema annulare (EA) [3]. Retrospectively in consideration of the rash that manifested with the patient in this case study, the original diagnosis was Nummular eczema based on characteristics that were compared to the characteristics cited in Fitzpatrick's Color Atlas and Synopsis of Clinical Dermatology: Sixth edition. When the lab results returned with a positive SS-A Antibody and based on the research that was conducted regarding SS, it is more likely that the cutaneous manifestation was EA. There are 3 types associated with EA, and each type has varying characteristics. Type I characteristics are isolated doughnut ring-like with elevated borders. Type II has marginally scaled polycyclic erythema and Type III has papular insect bite-like erythema [4].

The patient in this case study most likely had Type II. The cutaneous manifestations were described as scaly and erythema on the majority of the examinations. There could possibly be a mix of Type I as well as because the manifestations were also described as raised with induration. Without further histological testing, the exact Type of EA cannot be determined. According to Brito-Zero'n, there are cutaneous manifestations that can mimic EA such as Pityriasis rosea and Tinea Coporis. These two dermatological diagnosis were suspected on the patient's first and third visits. In studies conducted by Brito-Zero'n, EA is most responsive to Prednisone 10-20 mg per day. Although the patient had been prescribed Prednisone after the second visit, it was short duration. The patient stated she began to see a difference in the rash and it had stopped spreading after she received a pulse dose of Solu-Medrol 125mg Intra-muscular and a 10-day Prednisone dose pack. Her response is similar to the responses obtained in the research [2].

EA has been associated with Asian descent since it was first discovered in 1976 and has been known to have a geographical link to that region. It also has been associated as the Asian counter-part in non-Asian patients diagnosed with subacute lupus [2]. SS has a higher rate of being diagnosed in females than males. It also has a higher rate in patients that have a family history of Rheumatoid Arthritis (RA) and SLE [5]. Referring to (Table 1), in the patient's genogram it is noted that her father's ethnicity is Chinese; however, no further information was available in regards to the paternal side of the genogram. On the maternal side it is noted that two of the patient's uncles were diagnosed with RA, as well as her great grandfather. The medical history of her great grandmother is unknown.

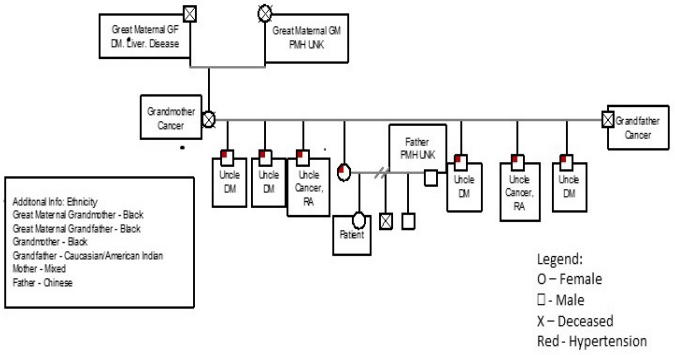

Table 1: Patient's genogram.

According to a study performed at the Department of Dermatology and Itch Center at, Temple University School of Medicine, Valdes-Rodriguez and et.al discovered that pruritus, was one of the presenting symptoms that patient's reported. Fifty-three percent of their participants reported chronic itching and had a higher intensity when compared to itching caused by other

rheumatological disorders. The itching was more prevalent during the summer and winter months and worse in the latter part of the day. It was also noted in their study that xerosis, an abnormal drying of the skin was present in 90% of those that reported itching [6]. In each of the clinic visits, the patient in this case study described itching, which was worse in the evening. The time period covered was May through July in southern Georgia. The temperature range for Georgia during that time period is 70 - 80℉ and the relative humidity is 85-89% in the morning and 50-57% in the afternoon.

SS has also been associated with gynecological problems including: fetal loss, vaginal dryness, and painful intercourse. Patients have also expressed complaints relating to the GI tract and renal function [5]. In this patient's Past Medical History (PMH), she related that as a child she had recurring kidney hydronephrosis that spontaneously resolved at age 24. At the age of nine she was treated for a pelvic issue, in which she does not recall the exact circumstances. She also relayed that during her six pregnancies, she went into early labor at six months and was restricted to bedrest for the duration of each pregnancy [7]. All births occurred with complications. She is also positive for the sickle cell trait. Taking into consideration her PMH, it is very possible that the complications she experienced could be related to SS.

In conclusion although SS typically presents with a complaint of dry eyes, dry mouth and swollen parotid glands, this case study demonstrates that there can be atypical presentations. It is the responsibility of providers to look at each patient with open eyes and open minds. Providers need to continue to take the whole patient into account when making a diagnosis. These patients are slipping through the cracks because the symptomology is vague. It is up to providers to put all the puzzle pieces together, and provide SS patients with a more accurate and timely diagnoses.

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