Research Article

The Current Underdiagnosis of Sjogren's Disease, Pathophysiology and Comorbidities.

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Abstract

Introduction

Sjögren's syndrome is a chronic autoimmune disease, classically producing dry mouth and dry eyes. Over time it is now known that it goes beyond that. As it progresses, it compromises renal function, it severely affects the joints, the intestines, and both the brain and spinal cord. It's much more common in women, and more frequent between the fourth and fifth decades of life. It is estimated that only 50% of patients with Sjogren´s Syndrome are correctly diagnosed over their lifetime. The reason for this is: fibromyalgia is often mistaken for SS, as are Multiple Sclerosis and many other autoimmune diseases.

This is because SS can mimic many other diseases, and without a certain suspicion of this disorder, it may be difficult to diagnose it due to the required tests. Although its etiology remains uncertain, in my experience I found a common factor in all 22 patients currently under my treatment: an overreaction to the Epstein Barr virus, also thought to cause many cases of MS, cryoglobulinemia, non-Hodgkin´s lymphoma, etc. This would make sense since the Epstein Barr virus can efficiently immortalize primary b cells, and one of the main characteristics of Sjogren´s Syndrome pathophysiology is over-activation of b cells. I also found more cerebrovascular disease than what is currently believed (around 20% is the current belief) affecting 17 of these 22 patients.

Although it is believed that the most common associated autoimmune diseases associated with SS are lupus and rheumatoid arthritis, to my surprise, celiac disease was by far the most associated autoimmune condition in these patients with SS (12 out of 22) (Only one patient also had SLE and none of them Rheumatoid Arthritis). In this article, we will also discuss the importance of Brain SPECT

imaging as a tool for both its diagnosis and as a beginning to understand its pathophysiology.

Purpose

To review the main comorbidities associated with Sjogren's Syndrome, a review of its believed pathophysiology, to review the currently believed percentage of cerebrovascular involvement throughout the disease, and to determine the actual importance of PET imaging as a tool for both diagnosis and treatment follow up.

Materials and methods

I reviewed 22 patients with SS, all of which underwent different treatments. I looked for patterns before each diagnosis was made, trying to add to the still not entirely understood pathophysiology of this disease. I then reviewed how many of them presented cerebrovascular involvement throughout the disease. Looked at which was the most common comorbidity in these cases, and how many of them had benefited from PET scan imaging.

Results

All 22 patients had presented an overreaction to the Epstein Barr virus (11.5UA-ml being considered positive, the lowest one was 92 and the highest one 167), previous to their diagnosis. Sjögren's syndrome (SS) is characterized by B cell infiltrates in the salivary glands and an increased risk of B cell lymphoma. This would make sense since this virus can immortalize b cells. It is known that migration of CD27+ memory B-cells, from peripheral circulation into inflamed salivary glands, results from increased expression of B-cell chemo-attractants CXCL12 and CXCL13 (major B cell attractants mainly produced by follicular dendritic cells) in the inflamed glands.

This is also seen in other autoimmune diseases like cryoglobulinemia, where B-cell expansion is the biological substrate of the disease. It is also known to play a direct role in the pathophysiology of Multiple Sclerosis. It is currently accepted that SLE and RA are the most common comorbidities in this disease. However, celiac disease was by far the most common one (12 out of these 22 cases). A role for B cells in celiac disease pathogenesis is receiving increased recognition. Afterwards, I reviewed how many of them had presented cerebrovascular involvement throughout the disease. To my surprise, 17 out of these 22 patients had experienced some minor or major cerebrovascular involvement throughout the disease (one of them experienced sentinel bleeding accompanied by sentinel headaches) It is believed that only around 20% of patients with Sjogren's Disease develop cerebrovascular involvement.

Finally, I looked at those who had undergone brain SPECT imaging either before or after diagnosis (5 patients), and all of them showed hypoactivity at the brain cortex (at different degrees).

Conclusion

Sjogren's disease is still an underdiagnosed and misunderstood disorder. Some common results are slowly shedding light on its pathophysiology, which will make both the diagnosis and treatment easier over time. Conflicts of interest: none to declare.

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