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A Case of Neuro Besnier-Boeck-Schaumann Disease that Co-existed with Sjogren's syndrome

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Abstract

Sarcoidosis and Sjögren's syndrome (SS) coexistence has long been overlooked because sarcoidosis is considered an exclusion criterion for SS. We described a 55-year-old woman who had been diagnosed for 16 years with coexisting neurosarcoidosis and Sjögren's syndrome. She had erythema nodosum, progressive sensory and motor impairment of the extremities, dry mouth, and dry eyes when she arrived. A symmetrical pulmonary micronodule, interstitial changes, and enlarged mediastinal lymph nodes were seen on high-resolution computed tomography (HRCT) of the chest. Magnetic resonance imaging (MRI) of the spine revealed syringomyelia and thickening of the T3-9 spinal cord. She had positive ANA and anti-SSA antibodies, as well as impaired lacrimal, salivary gland, and renal tubule function. Skin and lung nodule biopsies revealed non-caseous granuloma. A biopsy of the salivary gland revealed focal lymphocyte infiltration. Criteria for classification Based on clinical and laboratory findings, this patient met the criteria for sarcoidosis and Sjogren's syndrome. This case adds to our understanding of Sjogren's syndrome and sarcoidosis overlap and provides a reference value for clinical diagnosis.

Keywords: Neurosarcoidosis • sjogren's syndrome • Rituximab

Introduction

Sarcoidosis and primary Sjögren's syndrome (pSS) are both autoimmune diseases that affect multiple organs and share some clinical characteristics. Sjögren's syndrome is a chronic condition that primarily affects the function of the salivary glands, lacrimal glands, and other vital organs. Sarcoidosis is distinguished by granulomatous inflammation that is not caseating. Neurosarcoidosis (NS) is a rare form of sarcoidosis with neurologic involvement. Nodular and linear enhancement with T1 hypointense/T2 hyperintense is typical imaging performance of spinal cord neurosarcoidosis. Biopsy confirmation in the nervous system, which would show non-caseating granuloma, is the gold standard for NS diagnosis. However, central nervous system specimens are difficult to obtain. Biopsies of the lungs and mediastinal/hilar lymph nodes can also be used as a powerful diagnostic tool. According to both the 2002 and 2016 censuses. Sjögren's syndrome should be diagnosed using the ACR/EULAR classification criteria and sarcoidosis should be ruled out. There have been few reports of pSS and sarcoidosis coexisting. We present the coexistence of sarcoidosis and pSS in a 55-year-old female with central nervous system involvement.

Literature Review

A 55 time old lady had intermittent paresthesia in the right upper branch for 16 times and progressive impassiveness/ chinking in the lower extremities for 4 times. She gradationally developed difficulty in walking, and glamorous resonance imaging (MRI) of the cervical chine showed syringomyelia. She passed 3 operations of "filum terminale resection", "herniated cerebellar tonsil resection" and "suboccipital relaxation". There was a flash enhancement after surgery. still, it didn't stop the complaint from progressing.

As her symptoms worsened, she was admitted to the Department of Rheumatology and Immunology in Peking University People's Hospital. A detailed medical history was collected along with with careful physical examination. Skin lesions of the left inside canthus and nasal alar were noticed. The rash around the root nodes was about 3 × 1 cm in size, with no desquamation, no itching and no pain. The muscle strength of the left lower extremity was grade IV, and the muscle strength of the right lower extremity was grade II. The skin vivisection revealednon-caseating granulomatous. In addition, the case had dry mouth and dry eye over the times, with a history of hypertension and Hashimoto's thyroiditis.

Her routine laboratory tests of complete blood count, renal and hepatic function, erythrocyte sedimentation rate(ESR), C- reactive protein(CRP) immunoglobulin and complement position were normal. Autoantibody diapason showed that Corpus (1160), anti- Ro/ SSA antibodies were all positive. The ANCA, and other seditious autoimmune complaint- associated antibodies, was negative. Serum angiotensin- converting enzyme (SACE) didn't increase (3.4 U/ L, normal 17 – 55 U/ L). HRCT showed symmetric pulmonary interstitial changes closed to pleura, micronodules distributed along with the bronchovascular packets and multiple enlarged lymph bumps in the mediastinum. Bronchoscopy showed that there were numerous small nodes under the bronchial mucosa. The chance of lymphocytes increased in bronchoalveolar lavage fluid (BALF) and the CD 4/ CD 8 lymphocytes rate was11.46, which increased significantly. The histopathology of mediastinal lymph bumps in both group 7 and group 4R necropsies wasnon-caseating granulomatous. MRI showed abnormal signal and thickening of the spinal cord from the position of the medulla oblongata to the position of the spinal cord, with direct improvement in the anterior part of the spinal cord, nodular improvement in the posterior part of the spinal cord at the position of C1, and spinal cord depression at the position of T3- 4. The lumbar perforation showed nonspecific vulnerable-affiliated seditious responses. Testing of serum and CSF for infection (especially tuberculosis), soft bulbar palsy, autoimmune encephalitis, multiple sclerosis, and Aquaporin 4 (AQP4, which is associated with neuromyelitis optica in Sjögren's pattern) were each negative. In addition, she was also examined for the function of the lacrimal gland (eye break- up time (BUT), Schirmer test) and salivary gland (salivary gland nuclear imaging), which showed severe impairment. Ultrasonographic examination of salivary glands revealed bilateral fibrosis changes. The labial glandular vivisection verified a focal lymphocytic sialadenitis.

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Discussion

Sarcoidosis is an idiopathic granulomatous complaint affecting multiple organs, with the pathogenesis that the CD4 T cells interact with antigen-presenting cells to initiate the conformation and conservation of granuloma. In this case, the patient presented with erythema nodosum of skin, multiple lung nodes, blowup of mediastinal lymph knot, and elevation of lymphocytes, especially CD4 T cells, in bronchoalveolar lavage fluid. The vivisection samples showednon-caseous granuloma in both erythema nodosum lesions and mediastinal lymph bumps. thus, the opinion of sarcoidosis was made.

Primary Sjögren's pattern is a multisystem complaint that shares certain clinical features with sarcoidosis. According to the 2002 and 2016 ACR/ EULAR bracket criteria for primary Sjögren's pattern, sarcoidosis should be barred before the opinion of pS. Many studies have paid attention to the coexisting of sarcoidosis and Sjögren's pattern. Casals etal., set up that there's a advanced frequence of articular and optical involvement, antinuclear antibodies (Corpus), RF, and positive anti- Ro/ SSA antibodies in cases with coinciding sarcoidosis and SS. Although Corpus could be detected in sarcoidosis, anti-Ro/ SSA antibodies were uncommon. Due to the symptoms of dry mouth, dry eye, and the laboratory chancing with the positivity of anti- Ro/ SSA antibodies in these cases, Sjögren's pattern was suspected. A labial gland vivisection was made for this case, which showed focal lymphocyte infiltration. Still, the histologic data of exocrine gland vivisection for sarcoidosis isnon-caseating granulomas. The opinion of Sjögren's pattern was made. Our case indicated that sarcoidosis and Sjögren's pattern can be diagnosed contemporaneously. Analysis of immunologic biographies similar as anti- Ro/ SSA antibodies and histopathologic findings may be veritably useful. In sarcoidosis cases with positive anti- SSA/ SSB antibodies and focal lymphocyte infiltration in a labial gland vivisection, samples may suggest a concurrence with Sjögren's pattern [1-5].

Conclusion

As some case reports and reviews showed, rituximab and infliximab were both implicit options to treat NS. Considering that the case combined NS with SS, rituximab was allowed to be a further voluntary medicine to treat both conditions. As the symptoms bettered after the rituximab remedy, we suppose that rituximab could be a implicit option for NS lapped with SS.

Acknowledgement

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Conflict of Interest

None.

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