Sudden Sensorineural Hearing Loss and Microangiopathic Antiphospholipid Syndrome (MAPS)

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A 54-years-old man with history of acute pancreatitis 4 years before and secondary DM, he also had history of primary hypothyroidism and hypertension. He was admitted in hospital in 2010 with sudden and bilateral hearing loss, headache and dizziness. Neuro-otological examination revealed a sensorineural hearing loss (SNHL) and he was given Dexamethasone trans-typanic, then he was treated with Ganciclovir (12 mg/kg/24 h) for 10 days + prednisone 50 mg/24 h for 4 weeks. At the end, pure tone audiometry confirmed a SNHL affecting both ears. aCL antibodies were positives and he was send to internal medicine consultation. The diagnosis of microangiopathic an-

Conclusion: MAPS is a subset of Antiphospholipid Syndrome which occlusive thrombosis of small blood vessels and microangiopathy dominate the clinical picture. Several neurological disorders have been described in association with antiphospholipid antibodies (aPL) without thrombosis of large-vessels like Myelitis Transverse, Multiple Sclerosis-like Disease, Dementia, Chorea, Migraine and Epilepsy. SNHL is a rare syn-

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Predictive Significance of CCL21 and CXCL13 Levels in the Minor Salivary Glands of Patients with Sjögren’s Syndrome

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Background: In the current study, we investigated whether the laboratory and clinical manifestations of SS patients were associated with CCL21 and CXCL13 expression levels in the minor salivary gland.

Methods: We obtained sociodemographic data on a total of 106 SS patients, documented glandular and extraglandular manifestations of the disease, performed minor salivary gland biopsies, and analyzed laboratory findings. EULAR index values of SS disease activity (ESSDAI) values at the time of biopsy, and SS disease damage index (SSDDI) values, were also noted. An immunohistochemical approach was used to (semi)quantitatively measure the expression levels of CCL21 and CXCL13 in the minor salivary glands.

Results: The minor salivary glands of SS patients stained positively for CCL21 and CXCL13 in 46.2% (49/106) and 70.7% (75/106) of all cases, respectively. Increased expression of CCL21 was associated with an elevated ESR, an increased IgG level, elevated anti-SS-A and -SS-B titers, a higher focus score, and a greater ESSDAI value at the time of biopsy. Increased expression of CXCL13 was associated with an increased WBC, reduced lymphocyte, low platelet, elevated ESR, an increased IgG level, elevated anti-SS-A and -SS-B titers, a rise in the level of rheumatoid factor, a higher focus score, and a greater ESSDAI value at the time of biopsy. In patients with extraglandu-

Conclusions: The expression levels of CCL21 and CXCL13 within the lymphocytic infiltrates of SS patients were associated with several laboratory features of the disease, lymphadenopathy, and the extent of clinical disease activity. CCL21 and CXCL13 levels should serve as useful markers predicting SS disease activity and prognosis.

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A Case of Sweet’s Syndrome Diagnosed Concomitantly with Sjögren’s Syndrome

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Sweet’s syndrome is an uncommon reactive dermatoses characterized by fever, polymorphonuclear leukocytosis, painful erythematous plaques and dense dermal infiltrate of neutrophils. Sweet’s syndrome can be associated with several diseases, such as infectious diseases, malignant tumors, and autoimmune diseases. However, there is no case report of Sweet’s syndrome associated with Sjögren’s syndrome in Korea. A 44-year-old woman presented with acute pustular rashes and fever. The patient had multiple papulopustular skin rashes, and complained fever, chills, and headache. Our patient had the characteristic clinical and histopathological features of Sweet’s syndrome, in association with Sjögren’s syndrome (SjS), which was diagnosed through a salivary gland scan, positive anti-SSA/SSB antibody, and sicca symptoms simulta-

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