中文題目：一位年輕女性罹患腦梗塞與乾燥症：病例報告與文獻回顧
英文題目：A young woman with Sjogren's syndrome and ischemic stroke: A Case Report and Review of the Literature
作者：張書明、李麗青、羅健賢、徐北辰
服務單位： 苗栗大千綜合醫院內科部

Introduction: Sjogren's syndrome (SS) is a systemic autoimmune disorder of unknown pathophysiology, characterized by destructive lymphocytic infiltration of the exocrine glands. The neurologic manifestations of SS involve both the peripheral and central nervous systems (CNS). CNS involvement occurs in 5.8~68% of primary SS patients. However, SS-associated infarction seldom occurs. Here, we report a young woman with Sjogren syndrome and ischemic stroke

Case report: A 39-year-old woman was admitted to the Neurologic Ward for acute onset left sided motor weakness and facial palsy for 2 days.
Tracing back her history, she suffered from dry eye, dry mouth and severe decayed teeth for years, and intermittent skin purpura on both the low legs. She had no history of hypertension, hypercholesterolemia, or diabetes mellitus, and no other medical history. She was a non-smoker and non-drinker. On physical examination the patient was found to have hypertension, skin hyperpigmentation in legs, motor weakness, hyperreflexia and hypoesthesia of the left limbs and left sided face. Laboratory examinations showed a microcytic anemia (Hb: 9.4g/dl; MCV: 73.4 fl) with normal white blood cell and platelet counts; hypergammaglobulinemia (IgG 1994 [normal range 870-1700]); positive rheumatoid activity (108 , [0-20]), ANA (>600, [<0.7]), Anti Ro (>600, [<7]), Anti La (13.7, [<7]). Ant-ds DNA antibodies and other anti-extractable nuclear antigens (anti Smith, anti RNP), anticardiolipin antibodies were negative. Prothrombin time (INR: 0.98) and activated partial thromboplastin time were all in the normal range. Schirmer's test was positive (OD 2mm, OS 2mm in 5 minutes). Transthoracic and transesophageal echocardiography showed adequate LV and RV contractility with normal wall motion and no intra-cardiac thrombus. MRI scans of the brain revealed evidences of new infarct lesions at right internal capsule upward extended to right corona radiata.
The presence of four of the revised international classification criteria for SS in this patient confirmed the diagnosis. In view of the underlying autoimmune disorder (SS and possible cutaneous vasculitis) and low risk for stroke, it seemed likely that the etiology of the cerebral vascular event is autoimmune-related vasculitis. The patient was therefore treated by aspirin 100mg/day, hydroxychloroquin 400mg/day, amlodipine 10mg/day, trandate 200mg bid. We continued to follow up her for more

than 4 years, and she did not present with any other cerebral ischemic symptoms or cutaneous vasculitis.

Discussion: Because of young age and low vascular risk for cerebrovascular disease, autoimmune diseases were considered in the causes of the stroke. Other clinical clues that alerted us to the presence of an underlying immunologic disorder were intermittent purpura and hyperpigmentation on legs. After thoroughly history taking and examination, our patient fulfilled the diagnostic criteria for SS, but not for systemic lupus erythematous or antiphospholipid antibody syndrome. The etiopathogenesis of the neurological damage in SS seems to be immunologically mediated. The most common mechanism is due to vasculitis of small vessels.
In conclusion, we suggest that young stroke female patients should be evaluated carefully and thoroughly for SS and other autoimmune diseases.