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A case of Barraquer-Simons syndrome, as a subtype of systemic sclerosis

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Introduction: Barraquer-Simons Syndrome, also called acquired partial lipodystrophy(APL) is rare disease, characterized by the symmetrical loss of fat of face, neck, chest, abdomen and arms. On the other hand, systemic sclerosis is connective tissue disorder of fibrosis and sclerosis of the skin and internal organs. We describe a patient who presented bilaterally symmetrical subcutaneous fat atrophy with systemic sclerosis feature. **Case Report:** A 40-year-old women visited outpatient clinic for evaluation of gradual buccal fat loss. The bilateral symmetrical subcutaneous fat loss developed over her face, abdomen, and upper extremities for two years. Her clinical feature suggested Barraquer-Simons.Abdominal skin biopsy was performed which showed nearly no presence of subcutaneous fat. Only thickened, sclerotic dermis with sclerosis fibrosis extending to the subcutis was observed, raising a possibility of scleroderma. Thorough history taking with physical examination revealed she had Raynaud's phenomenon with puffy fingers, finger tip pitting scar and telangiectasia. Laboratory exam showed no presence of autoimmune antibodies except positive rheumatoid factor (18.2 IU/mL). Nailfold capillary showed hemorrhage. Her clinical manifestation satisfied 2013 classification criteria of systemic sclerosis and anti-rheumatic drugs were started. **Conclusions:** The association between Barraquer-Simons syndrome and autoimmune diseases, particularly systemic lupus erythematosis and dermatomyositis was reported previously. However, the patient presented here had coexistence of Barrquer-Simons syndrome and systemic sclerosis. Fat loss of abdomen began concurrently with development of Raynaud's phenomenon and puffy fingers and subcutaneous fat loss of over face and trunk was observed with skin sclerosis. Thus, there is possibility of Barraquer-Simons syndrome as a subtype of systemic sclerosis and it is strongly recommended that the patient with Barraquer-Simons syndrome be screened for systemic sclerosis.

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다발 신경병증과 혈관염 동반된 쇼그렌 증후군 환자 증례보고

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서론: 쇼그렌 증후군(Sjïgren's syndrome)은 눈물샘, 침샘과 같은 외분비샘에 림프구가 침윤되어 눈 마름증과 입안 마름증이 나타나는 만성 자가 면역 질환으로, 다양한 선외 증상(Extra glandular)을 유발할 수 있으나, 혈관, 신경계를 침범하는 경우는 다른 자가 면역질환에 비해 드물다. 저자들은 쇼그렌 증후군에 의해 유발된 다발 신경병증, 혈관염 환자의 증례를 보고하고자 한다. **증례:** 특이 과거력 없던 59세 여자 환자는, 내원 2개월 전부터 양하지의 열감, 부종 호소하였고 내원 당일, Lt.foot dorsiflexion되지 않으면서, foot drop 증상으로 정형외과로 내원하였다. Lymphangio scan상 Lt.leg lymphedema 소견 보이면서, 검사결과는 ANA 1; 320, C3/C4 60/10, anti R o (+) anti La (-), RF 115, anti CCP (-) 나왔다. Salivary scan상 양측 귀밑샘, 턱밑샘 모두 충만 되지 않으며, 타액선 배출장애 양상 관찰되었어, 쇼그렌 증후군 진단되었다. 이후 양하지 위약감 지속되어 시행한 근전도검사상 양 총비골 신경 손상 및 다발 신경변증 의심양상 관찰되어, 혈관염(vasculitis)에 의한 sensor polyneuropathy 확인 위해 Nerve bx.시행하였고, vasculitic neuropathy 진단되어, Steroid pulse (1 g) IV 3일간 시행후, cyclophosphamide pulse (1 g) IV 시행하였다. 이후 prednisolon 1 mg/kg po유지하면서, 외래 추적 관찰중, 증상 호전되어 점차적으로 감량하면서 현재는 중단한 상태이며, cyclophosphamide는 severe leukopenia로 중단후 Cyclosporine A로 변경하여 복용하며 외래 추적 관찰중이다. 본 증례는 쇼그렌 증후군에서 혈관과, 말초신경 동시에 침범하는 첫번째 사례로 발현 환자 증례 보고하는 바이다.