A CASE OF HEMOPHATOCYTIC SYNDROME DEVELOPED IN A KOREAN FEMALE PATIENT WITH DERMATOMYOSITIS

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Background and aims Dermatomyositis (DM) is characterised by chronic inflammation of striated muscle and characteristic cutaneous manifestations. Hemophagocytic syndrome (HPS) is a rare life-threatening condition caused by uncontrolled activation of histiocytes resulting in prominent hemophagocytosis. Particularly, occurrence of HPS in the patients with DM is extremely rare.

Methods We report the first case of HPS in a patient with DM successfully treated in Korea.

Results A 56-year-old female visited our hospital, complaining of general weakness with whole body skin rash for 2 months. She had symmetric proximal muscle weakness and characteristic skin lesions including heliotrope rash, gottron’s papules.
Background and aims Sarcoidosis is known as a Th1-mediated disease which can mimic many primary rheumatologic diseases or sometimes co-exist with them. Clinical characteristics of sarcoid arthropathy are not well described and the studies reported in the literature so far are mostly based on the data from referrals. The aim of this study was to evaluate the incidence and clinical characteristics of sarcoid arthropathy.

Methods All our patients were prospective evaluated in our single Rheumatology outpatient centre from 2011 to 2015. 114 patients with sarcoidosis were included in the study.

Results The mean patient age was 48.1 years and the mean disease duration was 40.5 months. Sarcoid arthritis was observed in 71 (62.3%) and arthralgia in 106 (92.9%) patients. Out of the 71 patients with arthritis, 61 (85.9%) had involvement of ankle, 7 (9.8%) knee, 2 (2.8%) wrist, MCP and PIP joints, and one (1.4%) had shoulder periarthritis. Oligoarthritis (two to four joints) was the most common pattern followed by monoarthritis and polyarthritis. When the correlation between clinical findings was considered, erythema nodosum and arthritis and female gender were found to be correlated (p=0.03, p=0.001, respectively). Again in patients with arthritis, even higher levels of CRP/ESR as well as ANA and RF positivity were observed (p=0.03, p=0.01, p=0.01 and p=0.02, respectively). Eleven patients had another rheumatic pathology concurrent with sarcoidosis.

Conclusions Inflammatory arthritis occurs in a majority of patients with sarcoidosis. Acute arthritis with bilateral ankle involvement is the most common pattern of sarcoid arthropathy. Sarcoidosis can mimic many primary rheumatic diseases and/or may coexist with them.

Background and aims This study aimed to evaluate the hand perfusion scintigraphic features of hand-arm vibration syndrome (HAVS) and to compare these with the features of primary and secondary Raynaud’s phenomenon (RP) associated with rheumatic diseases.

Methods Hand perfusion scintigraphy was performed in 57 patients with primary RP, 71 patients with HAVS-related RP, and 36 patients with rheumatic disease-related RP. Patients’ clinical details were collected by a retrospective review of medical records. We calculated 6 ratios by using the time-activity curve and static blood pool images, the chilled to ambient hand and wrist ratios of the first peak height, initial slope, and blood pool uptake. We analysed 3 morphologic characteristics: slow progress pattern, paradoxically increased uptake pattern in the time-activity curve, and the inhomogeneous radioactivity uptake in the blood pool image.

Results All of the 71 patients with HAVS-related RP were mine workers. The onset of RP after exposure to vibration was at 21.8±7.3 years, with 26.3±7.0 years of vibration exposure time. The chilled to ambient hand ratios of the first peak height and the initial slope were significantly lower in patients with HAVS-related occupational RP than in patients with primary RP. The presence of a paradoxically increased uptake pattern on a scintigraphic feature of HAVS is useful in distinguishing HAVS-related RP from rheumatic disease-related RP.