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	Case 1	Case 2	Case 3	Case 4
Age at diagnosis of morphea (years)	11	11	8	11
Age at diagnosis of thrombocytopenia (years)	9	11	8	11
Classification of morphea	En coup de sabre	En coup de sabre	En coup de sabre (Figure 1)	En coup de sabre
Haemoglobin (gm/L) (Normal range 110-130)	128	117	122	127
White blood cell counts ($\times 10^9$cell/L) (Normal range 40-110)	108	91	80	70
Lowest platelet counts ($\times 10^9$/L) (Normal range 150-400)	8	44	82	120
ANA (antinuclear antibody)	Positive (2+ speckled)	Positive (3+ homogenous)	Negative	Negative
Anti double stranded DNA	Negative	Negative	Not done	Not done
Antibodies to extractable nuclear antigens	Negative	Negative	Not done	Not done
Antiphospholipid antibodies	Positive	Negative	Negative	Negative
Treatment for morphea	Topical calcipotriol ointment and Injection anti D	Oral methotrexate	Subcutaneous methotrexate	Subcutaneous methotrexate
Time to recovery of thrombocytopenia	1 month	1 year	Still persisting	4 months
Follow up (months)	78	54	10	8

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

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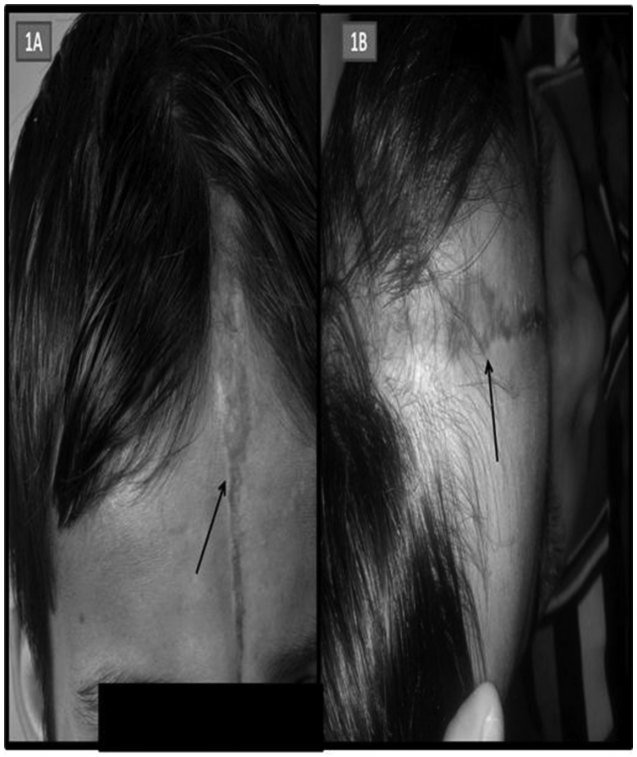
Background and aims Dermatomyositis (DM) is characterised by chronic inflammation of striated muscle and characteristic cutaneous manifestations. Hemophagocytic syndrome (HPS) is a rare life-threatening condition caused by uncontrolled activation of histiocytes resulting in prominent hemophagocytosis.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



Abstract 368 Figure 1

and V sign. She also had swallowing difficulty proven by abnormal videofluoroscopic swallowing test. Laboratory findings showed anaemia, thrombocytopenia, elevated muscle enzymes, hyperferritinemia and hypertriglyceridemia. The autoimmune profile revealed positive antinuclear antibody (1:160, homogenous pattern) and negative anti-Jo-1 antibody. In addition to electromyography and skeletal muscle biopsy, bone marrow biopsy was performed to find the cause of microangiopathic hemolytic anaemia and thrombocytopenia. Numerous CD68-positive macrophages engulfing erythrocytes and platelets were revealed in bone marrow study. She was finally diagnosed as DM with secondary HPS. After steroid pulse therapy for 3 days, we continued high dose steroid therapy for 1 month. Thereafter, we gradually tapered the steroid and started methotrexate. After 1 year of treatment, she was completely recovered from muscle weakness, swallowing difficulty, skin lesions and cytopenia.

Conclusions With this unique case, we would like to assert that HPS should be considered when cytopenia is observed in the patients with DM and that early aggressive therapy is needed.

370 THE CLINICAL CHARACTERISTICS OF SARCOID ARTHROPATHY BASED ON A PROSPECTIVE COHORT STUDY

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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.

371 THE USE OF HAND PERFUSION SCINTIGRAPHY TO ASSESS RAYNAUD'S PHENOMENON ASSOCIATED WITH HAND-ARM VIBRATION SYNDROME

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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