Distinct clinicopathologic and radiological manifestations of the skin, lung, and muscle diseases in patients with dermatomyositis positive for anti-aminoacyl tRNA synthetase antibodies (AB-aminoacyl tRNA synthetase antibodies score (AB-ARS Score))

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A group of patients with anti-synthetase syndrome (ASS) may constitute a unique subtype of dermatomyositis. We compared the cutaneous, muscular and pulmonary manifestations between the ASS and non-ASS groups of dermatomyositis patients. In our cohort, 59 patients with dermatomyositis were analyzed. Each patient referred to our university hospital from 2008 to 2016. The patients were examined for skin lesions, routine blood tests, autoantibody profiles using ELISA kits, skin biopsy, and radiological imaging of the lung and muscle diseases. The ELISA kit for measurement of anti-ARS antibodies is prepared to detect antibodies to Jo-1, PL-7, PL-12, EJ, and KS. We also examined antibodies to TIF1γ, MDAS and Mi-2. Of 59 patients with dermatomyositis, 20 patients were classified into the ASS group, and the remaining 39 patients were of the non-ASS. Patients with ASS more frequently presented with cutaneous lesions (p = 0.0019), arthritis (p = 0.0037), associated with elevated serum levels of C-reactive protein (p = 0.0029). Nineteen of 20 (95%) patients with ASS had interstitial lung disease (ILD) with fibrotic non-specific interstitial pneumonia (NSIP) or organizing pneumonia (OSIP), whereas patients with ASS showed the presence of myositis as well as myositis more frequently (p = 0.0041). Patients with ASS are characterized by the higher incidence of mechanic’s hands, systemic inflammation, ILD, and inflammatory myopathy associated with fasciitis, and elevated serum levels of ALD.

Clinical and genetic characteristics of cutaneous melanoma with clinical history of early childhood onset

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In the diagnosis of melanocytic lesions, it has not been fully established whether a clinical history of early childhood onset is sufficient to differentiate melanoma from congenital melanocytic nevus in adults. To clarify this point, 253 histopathologically proven adult cases of melanoma primarily developing on the skin and examined at Shinshu University Hospital between 2006 and 2015 were retrospectively reviewed. Melanoma arising from giant congenital melanocytic nevus was excluded. We selected patients who were identified as having a lesion at birth or in early childhood and evaluated their clinical, histopathological, and genetic findings. Oncogenic BRAF mutations were analyzed by the Sanger method. Ten (3.95%) of 253 melanoma cases satisfied the above clinical history, including 3 lesions found at birth. Among the 10 cases (median age: 39.5 years; 2 male and 8 female), the lesion was located on the extremities in 6 patients, on the trunk in 2 patients, and on the head and face in 1 patient each. Histopathological examination revealed that 9 lesions had developed on non-congenital sun-damaged (CSD) skin. The oncogenic BRAFV600E mutation was positive in 6 patients, negative in 2 patients, and undetermined in 2 patients. Our findings indicated that cutaneous melanoma with a clinical history of early childhood onset most frequently developed on non-CSD skin. Other characteristics, such as prevalence on extremities, young female predisposition, and positivity for the oncogenic BRAFV600E mutation, were common with melanoma occurring on non-CSD skin. Although the clinical history provided by patients is not always reliable, melanoma cannot necessarily be ruled out by a history of early childhood onset.

Eosinophil Cationic Protein (ECP), a predictive marker of bullous pemphigoid severity and outcome

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In the evaluation of bullous pemphigoid (BP), the target lesion sub-scoring of each patient and the best representatives of each severity level will be established. Previous studies determining severity cut-offs have yielded inconsistent results. Our objectives were to ascertain the responsiveness of the PDAI and ABSIS, establish MCID and re-affirm cut-offs. Prospective and retrospective cohort data collected from 2014-2016 (n = 20) were used to define a final routine visits. Patients were evaluated: baseline, day 60 and day 210. For the PDAI, ABSIS, Physician Global Assessment Visual Analog Scale (P-GA-VAS), Physicians Subjective Assessment of Disease Severity (PSADS), and Likert scale of the magnitude of change. The Likert scale and P-GA-VAS were used for responsiveness by using Pearson’s chi-squared test. MCID was obtained under order effect to the P-GA-VAS, and ≥3 on the Likert scale. Cut-scores were reaffirmed using PSADS. For responsiveness, using the Likert scale r = 0.4752 (p < 0.001), r = 0.0869 (p = 0.458), r = -0.5475 (p < 0.001) for PDAI activity, damage, and ABSIS, respectively. MCID for the PDAI was r = 1.3 and r = 1.585 for the ABSIS. Disease severity although cut-offs of 0.3/0.5/0.2 for the PDAI and 0.3/1.3/0.2 for the ABSIS were suggested to distinguish none, mild, moderate and severe disease A limited number of patients, patients with limited disease extent. We concluded that the PDAI is more responsive than the ABSIS. PDAI damage is not responsive. Standardised and practical use of the PDAI is now possible.