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Abstracts

Articles appearing in the May 2018 issue

Architectural B-cell organization in skeletal muscle identifies subtypes of dermatomyositis

Objective To study the B-cell content, organization, and existence of distinct B-cell subpopulations in relation to the expression of type 1 interferon signature–related genes in dermatomyositis.

Methods Evaluation of skeletal muscle biopsies from patients with adult dermatomyositis (aDM) and juvenile dermatomyositis by histology, immunohistochemistry, electron microscopy, and quantitative reverse-transcription PCR.

Results We defined 3 aDM subgroups—classic (containing occasional B cells without clusters), B-cell–rich, and follicle-like aDM—further elucidating IM B-lymphocyte maturation and immunity. The quantity of B cells and formation of ectopic lymphoid structures in a subset of patients with aDM were associated with a specific profile of cytokines and chemokines involved in lymphoid neogenesis. Levels of type 1 interferon signature–related gene expression paralleled B-cell content and architectural organization and link B-cell immunity to the interferon type I signature.

Conclusion These data corroborate the important role of B cells in dermatomyositis, highlighting the direct link between humoral mechanisms as key players in B-cell immunity and the role of type I interferon–related immunity.

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Symptomatic muscular sarcoidosis: Lessons from a nationwide multicenter study

Objective To describe clinicopathologic features of muscular sarcoidosis and the associated sarcoidosis phenotype through a nationwide multicenter study.

Methods Patients were included if they had histologically proven sarcoidosis and symptomatic muscular involvement confirmed by biological, imaging, or histologic examinations.

Results Forty-eight patients (20 male) were studied, with a median age at muscular symptoms onset of 45 years (range 18–71). Four patterns were identified: a nodular pattern (27%); smoldering phenotype (29%); acute, subacute, or progressive myopathic type (35%); and combined myopathic and neurogenic pattern (10%). In all patterns, sarcoidosis was multivisceral, with a median of 3 extramuscular organs involved (mostly lungs, lymph nodes, eyes, and skin) and a prolonged course with long-term use of corticosteroids and immunosuppressive drugs. Muscular patterns differed according to clinical presentation (myalgia, nodules, or weakness), EMG findings, muscular MRI, and response to sarcoidosis treatment. The myopathic and neuromuscular patterns were more severe.

Conclusion This nationwide study of muscular sarcoidosis allowed the identification of 4 patterns of granulomatous myositis, which differed by phenotypes and the clinical course.

NPub.org/N2/9112b

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M. Varrin-Doyer, K.L. Pekarek, C.M. Spencer, et al. 2016;3:e272. doi.org/10.1212/NXI.0000000000000272

Normal volumes and microstructural integrity of deep gray matter structures in AQP4+ NMOSD

C. Finke, J. Heine, F. Pache, et al. 2016;3:e229. doi.org/10.1212/NXI.0000000000000229

CSF isoprostane levels are a biomarker of oxidative stress in multiple sclerosis

F. Mir, D. Lee, H. Ray, and S.A. Sadiq. 2014;1:e21. doi.org/10.1212/NXI.0000000000000021

NMDA receptor antibodies associated with distinct white matter syndromes

Y. Hacohen, M. Absoud, C. Hemingway, et al. 2014;1:e2. doi.org/10.1212/NXI.0000000000000002

Aquaporin-4 autoimmunity

V. Lennon and A. Zekeridou. 2015;2:e110. doi.org/10.1212/NXI.0000000000000110