Promising Steps Towards a Better Understanding of OMS

K. Rostásy

Opsoclonus-myoclonus syndrome (OMS) is a rare disease characterized by opsoclonus, myoclonus, cerebellar ataxia, and behavioural disturbances associated in part of the affected children with a neuroblastoma. In the last years it has been increasingly recognized that a substantial number of children has a poor neurological and developmental outcome [1–3].

This has brought specialists in the field together to form a multinational European OMS working group with the principal aim to build up a network of experts involved in the care of and research on children with OMS.

The pathogenesis of this autoimmune-mediated syndrome is only incompletely understood. The current understanding is that a T-cell-dependent response to tumor-associated antigens leads to subsequent B-cell activation. This hypothesis is supported, for example, by the presence of T-lymphocytes in the neuroblastoma, expanded populations of CD5 B-cells in the cerebrospinal fluid and serum antibodies with cytotoxic properties in children with OMS [4–6].

Blaes and colleagues in their rapid communication add another layer to the different aspects involved in the manifestation of OMS. Similar to other immune-mediated diseases they show that parents of children with OMS have an increased frequency of autoimmune diseases. They further found that parents of children with OMS have a broader spectrum of autoantibodies including antibodies against structures of the central nervous system than the control group (43 % vs. 8 %, $p < 0.001$) [7]. Together with the observation that children with OMS have an increased frequency of the HLA class II locus DRB1*01, this indicates that genetic factors which, in particular, influence T-cell functions such as self tolerance and antigen presentation predispose a child to develop OMS and may explain at the same time why only a small number of children with neuroblastoma develop OMS [8].

Both observations should spark further research in the pathogenesis of this rare disorder.

The above-mentioned European OMS working group, which presented its objectives at the European Pediatric Neurology Society meeting 2007 in Izmir, Turkey, will serve as an ideal platform for recruiting further patients, coordinating research efforts between scientists and at the same time recommending treatment strategies. This combined effort will hopefully lead to new insights in the pathomechanisms and, more importantly, should improve the neurological and developmental long-term outcome of children affected by OMS.

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Correspondence
K. Rostásy
Department of Pediatrics,
Division of Pediatric Neurology
Medical University of Innsbruck
Anichstr. 35
A-6062 Innsbruck
Austria
Tel.: +43/512/5042 35 17
Fax: +43/512/5042 35 20
Kevin.Rostasy@uki.at