Inflammatory myopathies: management of steroid resistance

- The inflammatory myopathies include polymyositis, dermatomyositis, necrotizing autoimmune myopathy (NAM), and inclusion body myositis (IBM).
- On the basis of clinical experience, most patients respond to corticosteroids to some degree or for a time period. For patients insufficiently responding or for steroid-sparing, the treatment options vary among practitioners, generating a genuine uncertainty.
- For patients insufficiently responding to corticosteroids, the commonly used immunosuppressants, such as azathioprine, mycophenolate, methotrexate, or cyclosporine, may exert a nonevidence-based 'steroid-sparing' effect but provide minimal benefit on their own.
- The second line therapy is intravenous immunoglobulin (IVlg) based on a controlled study conducted in dermatomyositis; the drug is also effective in many patients with polymyositis and NAM.
- Rituximab and tacrolimus may offer additional benefit. Anti-TNF agents are disappointing. IBM remains difficult to treat; although early on some patients may partially respond to steroids or IVlg, they soon become unresponsive and the disease progresses.
- The main obstacles in the treatment of inflammatory myopathies are the lack of controlled trials and the absence of standardized outcome measures able to capture meaningful changes connected with disability and quality of life.
- The choice of treatment and the sequence by which the various immunotherapeutic drugs are used in inflammatory myopathies remain empirical.
- IVlg is the second line therapy in polymyositis, dermatomyositis, and NAM.