Plenary III: autoimmune diseases beyond SLE (hybrid)

33 AUTOIMMUNE ENCEPHALITIS

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Patients with autoimmune encephalitis can present with a variety of neurological symptoms such as short-term memory loss, behavioral changes, abnormal movements, seizures or focal deficits.[1 2] These complex and severe disorders have become increasingly recognized in the past 15 years, thanks to the discovery and characterization of neural autoantibodies that cause autoimmune encephalitis.[3] They can affect people of all ages and the clinical progression is acute or subacute, from days to weeks, in contrast to neurodegenerative disorders. Evaluation for infectious or other alternative conditions is of high clinical relevance as a first approach. Cerebrospinal fluid studies, magnetic resonance imaging and appropriate detection of neural antibodies have a key role in the diagnosis and management of patients with autoimmune encephalitis.[4 5] A rapid identification is crucial, as these diseases are treatable. Neural antibodies associate with specific clinical syndromes, different comorbidities (such as tumours), and show different responses to immunotherapy and prognosis. These novel antibody-mediated syndromes have changed dramatically the field of neurology, with impact in other disciplines such as pediatrics, psychiatry, or infectious diseases.

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Learning Objectives

- Describe the general clinical features of autoimmune encephalitis and the diagnostic and therapeutic approach
- Explain the difference between cell-surface and intracellular neural antibodies and their implications
- Discuss the role of potential triggers of paraneoplastic and post-infectious syndromes

34 INTERSTITIAL LUNG DISEASE

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Interstitial lung diseases (ILD) encompass a broad group of more than 200 parenchymal pulmonary disorders in which the lung tissue is comprised. Connective tissue disease related ILD comprise almost 20% of all ILD, rheumatoid arthritis and systemic sclerosis (SSc) making up the largest proportion of CTD-ILD.[1] Some CTD-ILD may have an acute course, such as anti-MDA5 positive rapidly progressive amyopathic dermatomyositis related ILD, others may have a chronic course. CTD-ILD are frequently progressive bringing along an as dismal disease course as idiopathic pulmonary fibrosis. Several challenges are related to CTD-ILD. One of the challenges concerning is firstly to think of them. The second challenge is to screen for them. The third challenge is the optimal management. In this way only for SSc related ILD we have generally adopted screening/monitoring strategies as well as several randomised controlled trials to evaluate the efficacy of immune suppressive and antifibrotic drugs.[2] Multidisciplinary approach in diagnosis and management of CTD-ILD is paramount.

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35 SYSTEMIC CAPILLARY LEAK SYNDROME

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Capillary leak syndrome (CLS) is a rare, life-threatening disorder characterized by recurrent episodes of fluid leaking from the blood vessels into the surrounding tissues, hypotension, oedema, haemoconcentration and hypoalbuminemia.[1] CLS can be idiopathic (Clarkson's disease) or secondary to various conditions and treatments.[1] Secondary CLS typically results from, systemic autoimmune disorders, viral infections, malignant haematological diseases and treatments such as chemotherapies and therapeutic growth factors.[1] Diagnosis of idiopathic CLS is made by exclusion of secondary diseases, especially as a serum monoclonal immunoglobulin is present, or when there is a relapsing disease, no initial lung involvement or preserved consciousness despite low blood pressure.[1] Prophylactic treatment of Clarkson's disease with intravenous immunoglobulin may reduce the frequency and severity of attacks and may improve survival.

REFERENCE

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Learning Objectives

- Describe the clinical presentation of systemic CLS, including symptoms, signs, and complications
- Discuss the diagnosis of systemic CLS, including diagnostic tests and criteria.
- Discuss treatment options for systemic CLS, including both acute and chronic management