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SP0074 DIAGNOSIS OF CEREBRAL VASCULITIS

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Among all forms of vasculitis, perhaps that affecting the CNS (CNS-V) poses the greatest challenge in diagnosis given the lack of any non invasive high specificity test, the complex and extensive list of mimicking conditions, its inaccessibility for biopsy and its genuine rarity. The diagnosis still requires the presence of an unexplained neurologic sign or symptom(s) following an exhaustive evaluation, evidence of vascular involvement (by direct or indirect angiography) or biopsy and most importantly the meticulous exclusion of all those conditions capable of producing mimicking clinical, radiographic or histologic findings that would confound accurate diagnosis. Despite these challenges CNS-V has been increasingly reported due to a combination of increased diagnostic awareness and advances in diagnostics. Two of these major advances will be discussed including the use of direct vascular wall imaging to differentiate vascular inflammation from spasm or atherosclerosis and the use of next generations sequencing to identify infectious etiologies and obviate the use of biopsy. Clinical examples will be presented.

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THURSDAY, 13 JUNE 2019

15:30:00 – 17:00:00

How to manage and treat childhood onset lupus? A multidisciplinary point of view

SP0075 CASE PRESENTER: CHILDHOOD ONSET OF NEUROLUPUS

Eve Smith. *Alder Hey Children's NHS Foundation Trust, Paediatric Rheumatology, Liverpool, United Kingdom*

Background: In the series of talks 'How to manage and treat childhood onset lupus? A multidisciplinary point of view' the first talk focuses on achievement of the initial diagnosis of Lupus within the childhood period, including some potential pitfalls and how these can be avoided. Initial and ongoing treatment within the childhood period, alongside paediatric specific therapeutic considerations will be discussed. The role of the multidisciplinary team of doctors, nurses, physiotherapists, occupational therapists, psychologists and sub-specialists will be highlighted. The impact of the diagnosis on the child, family, education and social activities will also be described. This talk will be followed by a subsequent talks following the patient through transition and into adulthood.

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SP0076 CASE PRESENTER: TRANSITION OF NEUROLUPUS PHASE INTO ADULTHOOD

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Childhood-onset SLE (cSLE) accounts for approximately 20% of all SLE cases and is often regarded as one of the most complex rheumatic diseases. With the increase of five-year survival rate of cSLE patients to over 95%, there is a growing number of adolescents and young adults (AYA) transferring from pediatric to adult care. Even in the best-case scenario, there is often considerable challenges regarding this transition. Hence, it is not surprising that morbidity, mortality and disease activity in cSLE, as well as in many other chronic illnesses and conditions, worsen during or just after the transition. The reasons for these deteriorations are numerous, with most prominent arising from the nature of the disease and changes during the tumultuous period of adolescence, but also the differences between pediatric and adult clinical and healthcare settings. Besides, the AYA patients with chronic conditions often feel they are not well prepared for the transition to adult care, while the adult rheumatologists commonly report concerns about assuming the responsibility of patients with the pediatric-onset disease. Some of these concerns are probably motivated by increased vulnerability of AYA patients with cSLE, who are more likely than their adult counterparts to develop lupus nephritis and neuropsychiatric manifestations, as well as other organs involvement and atherosclerosis. Moreover, due to the severity of the disease, patients with cSLE are treated more aggressively, accumulating more drug-related toxicity than patients with adult-onset SLE, which results in significant SLE-related damage and complications such as osteoporosis during the childhood. On the other hand, there is decreased medication adherence in AYA, further complicated by the neurocognitive and memory impact of the disease, as well as high rates of comorbid depression and anxiety. Some additional problems in cSLE patients at transition age are sexuality, fertility, and pregnancy. All this makes patients with cSLE require specialized and multidisciplinary care at the transition, that is capable of addressing medical, psychosocial, educational and vocational needs.

This presentation will be part of the comprehensive session discussing distinct features of the SLE during the different phases of life. It will emphasize specific challenges of the transition from pediatric to adult care, with the use of a compelling clinical case as an example.

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SP0077 CASE DISCUSSANT

Brigitte Bader-Meunier. *Hôpital Necker, Pediatric Immunology and Rheumatology, Paris, France*

Background: Padiatric-onset systemic lupus (pSLE) is a severe multisystem autoimmune condition which mostly manifests after the age of 10 years. It is mainly a polygenic disorder, but a mutation in a single gene may predispose to the development of SLE (« monogenic lupus »). We will describe some unusual presentations which may lead to a delayed diagnosis or misdiagnosis and provide clues for the diagnosis of monogenic lupus.

Objectives: To discuss the difficult diagnosis and pitfalls in pSLE

Results: The most common initial manifestations of pSLE are cutaneous, musculoskeletal, renal, hematologic, and fever. However, atypical symptoms may reveal the diagnosis or occur during the course of SLE. In a French retrospective study¹, thirty-two percent of children had atypical symptoms, mainly including abdominal involvement in 26 patients, which lead to negative laparotomy for presumed appendicitis before the diagnosis of SLE could be established. Abdominal pain was related to lupus pancreatitis or peritonitis. The diagnosis of pSLE should be systematically searched for in a teenager who presents with unexplained mood disorders, psychosis or depression, which may reveal pSLE. In a patient