Neurological manifestation of systemic diseases

**PP4253**

**Condition of cognitive functions in patients with systemic lupus erythematosus**

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**Introduction:** Epidemiological studies in recent years have shown that SLE is more common than previously thought, according to J. Klippel and J. Decker, its identified 50-70 new cases annually per 1 million population, approximately 500 patients per 1 million people worldwide. But along with therapeutic symptom-complex in this disease manifests itself a number of neuropsychological syndromes. American College of Rheumatology describes about 20 such syndromes.

**Methods:** We observed 16 patients with clinically justified SLE (study group) and 10 healthy volunteers (control group). Age of patients was 16-49 years (mean age 29.25). SLE at all patients was sub acute or chronic and with pathology of the nervous system: a convulsive syndrome and stroke in 1 (6.25%), intracranial hypertension, vascular encephalopathy and poly neuropathy in 2 (12.5%), hypothalamic syndrome in 3 (18.75%), asthenisation syndrome in 4 (25%) cases. In both groups studied condition of the cognitive functions according MMSE, test of learning 10 words and FAB.

**Results:** According to the results of the MMSE in 37.5% of patients with SLE were defined mild cognitive disorders, in 56.25% moderate cognitive impairment and dementia in 6.25%. For results of memorize 10 words mean point 7.3% and FAB scale mean point was 13.8. In the comparison group cognitive functions were within normal.

**Conclusions:** Cognitive functions should be examined in all patients with SLE as a method of early diagnosis of CNS involvement.

**Disclosure:** Nothing to disclose

**PP4254**

**Neurologic involvement in acquired adult hemophagocytic lymphohistiocytosis: a clinico-pathological case**

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**Introduction:** Hemophagocytic lymphohistiocytosis (HLH) is a syndrome of severe immune activation rarely described in adults. Central nervous system is affected in up to 73% of children with HLH; meningitis, seizures and white matter abnormalities being most frequently reported.

**Case report:** A 47-year-old previously healthy woman presented with a four-month history of fever of unknown origin. Her physical examination was unremarkable. Blood tests showed anemia and high triglycerides, C-reactive protein and ferritin. Her spleen biopsy was relevant for hemophagocytosis. HLH diagnostic criteria of apparently idiopathic cause were met, after diagnostic tests have ruled out every thought hypothesis. Throughout inpatient period she developed a complex partial seizure. Brain MRI showed subcortical fuzzy lesions with hyperintensity in FLAIR, the largest one with contrast enhancement in right temporal lobe. CSF examination showed 6 normal lymphocytic cells, with negative microbiology studies. Metilprednisolone and intravenous immunoglobulin was administered before discharge. She started HLH-94 chemotherapy protocol with symptom and MRI lesions improvement. Two months after HLH-94 induction phase she developed disorientation and visual hallucinations. Brain MRI showed a severe lesion recrudescence with diffuse brain edema causing clinical worsening by coma and death. Post mortem brain pathology was consistent with a hemophagocytosis associated with a perivascular infiltration by monoclonal lymphocytes. Intravascular lymphoma (IVL) was simultaneously identified in the kidneys and lungs.

**Conclusions:** We describe a rare case of proved central nervous system hemophagocytosis in an adult patient with an IVL. HLH should be considered in the differential diagnosis of white matter disease even in adults.

**Disclosure:** Nothing to disclose
PP4255

Frequency of peripheral neuropathy and myogen lesion in antineutrophil cytoplasmic antibody associated (ANCA) small vessel vasculitis

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Introduction: In systemic vasculitis the peripheral nerves are affected in 60-70%, but myogen involvement is less known.

Methods: We tested the frequency of neuropathy and muscle damage in a homogenous ANCA small vessel vasculitis patient group. Eleven patients were recruited. Time since verifying ANCA positivity in average was 4 years. Neurological and neurophysiological examination (electroneurography, electromyography) were performed. Currently we present the preliminary data and results of our long-term prospective study.

Results: The peripheral nerves were affected in the majority of the examined patients. The detailed electroneurography results were the following: 2 patients suffered from mononeuropathy multiplex affecting the lower limbs, 1 of them the sensory and motor nerves and in the other only the sensory part have been affected. 6 patients had neuropathy in the lower limbs, in 2 patients only the sensory nerves were affected and the other 4 patients suffered from sensoro-motor neuropathy. Polyneuropathy was detected in 2 patients. The electroneurography was normal in 1 case, who has been diagnosed two weeks ago. Interestingly high percent of the patients had myogen lesion (54%). But electromyography revealed abnormality in all patients (5 patients neurogen lesion, 1 patient both).

Conclusion: We found that in most of the ANCA positive patients not only the peripheral nerve lesion is characteristic but the myogenic lesion as well. Follow up of the patients is necessary to detect the progression of the clinical neurophysiological conditions.

Disclosure: Nothing to disclose

PP4256

A case of autoimmune polyendocrine syndrome type 2 presenting with Hashimoto’s encephalopathy and signs of lower motor neuron involvement

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Introduction: Hashimoto’s encephalopathy (HE) is a steroid-responsive encephalopathy associated with elevated antithyroid antibodies, variously presenting with consciousness disturbance, psychosis, cognitive dysfunction, myoclonus, seizures and ataxia. HE can have an acute, relapsing-remitting or chronic course.

Methods: A 27-year-old man presented a progressive myoclonus epilepsy associated with ataxia and right limbs weakness and atrophy, slowly worsening over 5 years. At age 32, worsening asthenia and depression appeared and progressed to a stuporous state. Addison’s disease and Hashimoto’s thyroiditis were diagnosed. L-thyroxine and steroid therapy were started with clinical improvement. Brain MRI showed cerebellar atrophy. Cervical spinal cord MRI was normal. EMG showed widespread active denervation signs. Serum creatine kinase levels were increased. Anti-glutamic acid decarboxylase antibodies and high serum anti-thyroperoxidase and anti-thyroglobulin antibodies were detected. Plasma very long chain fatty acids were normal. Anti-VGCK, anti-VGCC and anti-GluR3 antibodies were absent. CSF oligoclonal bands were detected and confirmed by two different tests, performed six and eleven years after the onset. No mutation was found in the SOD1, SMN, AR, TBP and MT-ATP6 genes. The neuropsychological assessment revealed a cognitive impairment.

Results: HE in a patient with an autoimmune polyendocrine syndrome type 2 was diagnosed. An 11-year follow-up shows reduced myoclonic jerks, better control of seizures with valproate and unchanged limbs strength and atrophy.

Conclusions: This case highlights how HE, an overlooked treatable condition, can present with combined encephalopathy, lower motor neuron and adrenal insufficiency signs, mimicking an inherited metabolic or a neurodegenerative disorder, such as spinocerebellar degeneration or motor neuron disease.

Disclosure: Nothing to disclose
An elderly CTLN2 patient successfully treated with medium-chain-triglyceride-supplemented formula

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Adult-onset type 2 citrullinemia (CTLN2) is a glucose metabolism disorder and shows neuropsychiatric manifestations including aberrant behavior, nocturnal delirium, disorientation, consciousness disturbance and convulsive seizures. A 66-year-old Japanese man presented with recurrent episodes of neuropsychiatric manifestations and hyperammonemia. Liver cirrhosis or portosystemic shunt was not observed. An electroencephalograph showed generalized high voltage slow wave. We considered nonconvulsive status epilepticus and used antiepileptic agent with hyperammonemia treatment, but his symptoms were not improved. Further investigation showed high concentration of citrulline in both blood and urine, and high concentration of arginine in urine. Genetic analysis of SLE25A13 gene (cansative gene of CTLN2) was identified 24TCG (stop codon). Then we diagnosed that this case was having elderly onset CTLN2. He treated a low carbohydrate diet with Medium-Chain-Triglyceride (MCT)-supplemented Formula. His neuropsychiatric manifestations disappeared, and hyperammonemia was improved. MCT-supplemented formula is one of the effective treatments tool for elderly onset CTLN2 patients.

Disclosure: Nothing to disclose
PP4259

Peripheral neuropathy: rare manifestation in Henoch-Schönlein purpura

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Introduction: Henoch Schönlein Purpura (HSP) is a systemic vasculitis characterized by purpura, arthralgia or arthritis, abdominal pain and glomerulonephritis. Nervous system involvement mostly includes headaches, mental changes, seizures and focal neurological deficits. Peripheral nervous system dysfunction is very rare. We present a 35-year-old woman with HSP who suffered from peripheral neuropathy.

Case: A 35-year-old woman was admitted with complaints of numbness in the hands and feet. She had a history of arthritis, palpable purpura concentrated on arms and legs, and gastrointestinal bleeding 10 years ago. Skin biopsy had been performed and with clinical features she was diagnosed as HSP. Since then, oral prednisolone had been given when she had symptoms and signs. Physical examination was normal. Neurological examination revealed hypoesthesia with distribution of symmetrical glove and stocking type. Deep tendon reflexes of lower extremities were absent. Laboratory investigations were unremarkable. Electrophysiological findings on both upper and lower extremities showed moderately reduced sensory nerve action potential amplitudes with normal latencies, suggesting mild axonal neuropathy mostly in peripheral sensory nerves. After initiation of the oral prednisolone therapy, she improved rapidly.

Conclusions: We present a case with HSP with peripheral nervous system involvement. Neurological manifestations are very rare in patients with HSP. Peripheral nervous system dysfunction presents as polyneuropathy, mononeuropathy or mononeuropathy multiplex. Aggressive dosage corticosteroids and cyclophosphamide are not advised for HSP patients with a peripheral or cranial neuropathy, since they tend to full spontaneous recovery. In conclusion, although it is rare, neurologic involvement should be thought in HSP.

Disclosure: Nothing to disclose

PP4260

The value of repeat CT imaging of the thorax in the diagnosis of neurosarcoidosis: report of a case

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Case report: A 36 year old woman presented with a three day history of diplopia. On examination there was a left oculomotor nerve palsy. Cerebral MRI demonstrated enhancement of the affected nerve. CSF showed a lymphocytosis with raised protein but normal glucose. Microbiological investigations were negative as was cytology. Serum and CSF angiotension converting enzyme (ACE) levels and CT thorax were normal. A diagnosis of possible neurosarcoidosis was made, and treatment started with prednisolone. Although the oculomotor palsy resolved within three weeks, MRI showed deterioration with new parenchymal lesions in both cerebellar hemispheres. The CSF was re-examined and found unremarkable. A repeat CT thorax identified an increase in size of several lymph nodes, biopsy of which confirmed a histopathological diagnosis of sarcoidosis. High dose steroids and methotrexate resulted in an ongoing good clinical response and progressive improvement in MRI appearances.

Discussion: Neurosarcoidosis can be difficult to confirm: there may be no disease outside the nervous system, and the neurological lesions may not be amenable to biopsy. Normal serum and CSF ACE levels do not exclude the diagnosis. High resolution CT imaging of the thorax is commonly performed in this condition, although rarely repeated. In this case, a repeat of this investigation demonstrated progression of appearances which allowed a definitive diagnosis to be made and avoided the risks of brain biopsy. Although the risks of radiation need to be borne in mind, repeat CT thorax after an interval should be considered in patients with an unconfirmed diagnosis of neurosarcoïdosis.

Disclosure: Nothing to disclose
PP4261
Celiac disease and leukoencephalopathy
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PP4262
Pain syndrome intensity, psychoemotional condition and quality of life in patients with multiple myeloma
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PP4263
Systemic diseases revealed by neuropsychiatric features
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PP4264
Rendu-Osler-Weber disease and prothrombin mutation in heterozygosity - imminent danger?
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PP4265
Hearing loss: Behçet's disease or Susac's syndrome
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PP4266
Progressive dementia in a young woman revealing mixed connective tissue disease
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PP4267
POEMS syndrome: a diagnostic and therapeutic challenge
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PP4268
Chronic paraneoplastic leukoencephalopathy associated with multiple myeloma: a rare association
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PP4269
Delayed onset of dementia and parkinsonism in a postoperative hypoparathyroidism case
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PP4270
An autopsy case of multiple myeloma which revealed cerebellar ataxia and progressive dementia
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PP4271
A Creutzfeldt-Jakob disease patient treated with Korean medical treatment: a case report
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PP4272
Parkinson syndrome revealing a systemic lupus erythematosus disease: a case report and review of literature
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PP4273
Ischemic stroke revealing Wegener’s disease: one case study and review of literature
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PP4274
Adrenal tumor presenting as tetraplegia
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PP4275
Systemic lupus erythematosus manifested as drug-resistant mesial temporal lobe epilepsy
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PP4276
Loss of taste
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PP4277
Central nervous system (CNS) involvement in multiple myeloma (MM)
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