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XXXIX Congress
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ABSTRACTS

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was very elevated (3725 ug/L). Doppler revealed bilateral deep vein thrombosis; a CT scan revealed multiple ischemic lesions in lung, liver and spleen, and an ovarian mass. The patient was treated with low-molecular weight heparin and then was submitted to surgery of ovarian lesion removal.

Conclusions: Early detection of a marantic endocarditis may allow the recognition of an occult cancer with consequent timely appropriate anticoagulant therapy and surgery. The last fields have been described as possible sites of cardio-embolic lesions.

SPINAL CORD INFARCTION: MRI AND MEP FINDINGS IN TWO CASES

R. Nardone^{1,2}, J. Bergmann¹, M. Kronbichler¹, S. Klein¹, F. Caleri², H. Ausserer², M. Covi², P. Lochner², K. Unterholzner², F. Tezzon², G. Ladurner¹, S. Golaszewski¹

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Background: Because in the early phases of spinal cord ischemia magnetic resonance imaging (MRI) can be normal and diffusion-weighted MRI of the spinal cord is rarely performed of several technical issues, its clinical diagnosis is often difficult.

Objective: To explore if motor evoked potentials (MEPs) recordings can be a useful test for the early diagnosis of spinal cord stroke.

Methods: The clinical, MRI, and MEP findings in one case each of cervical and lumbar spinal cord infarction were reported.

Results: Spinal MRI at admission was unremarkable in both patients. At that time, MEPs were abnormal in both patients, to the upper and lower limbs in the first patient, exclusively to the lower limbs in the second. Follow-up MRI examinations documented an infarction in the territory of the anterior spinal artery and of the Adamkiewicz artery, respectively.

Conclusions: MEP study can be useful, along with clinical examination, in demonstrating spinal cord involvement and in localizing the site of central motor pathway dysfunction. TMS provides neurophysiological evidence of a corticospinal conduction defect also when radiological evidence for spinal cord damage is absent or equivocal.

MAC CUNE ALBRIGHT SYNDROME. A CASE REPORT

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Fibrous dysplasia is a congenital, non-hereditary skeletal disorder that occurs with equal frequency in males and females. It is a developmental anomaly of bone formation in which the marrow is replaced by fibrous tissue. One percent of biopsied bone lesions are due to fibrous dysplasia. Monostotic disease is more common than polyostotic disease. When polyostotic, all the lesions tend to occur on one side of the body. The bones most frequently involved are the long bones: femur (most common), skull, and the ribs. Polyostotic disease can be associated with abnormal skin pigmentation (ipsilateral to the osseous lesions) and endocrinopathies. The constellation of polyostotic fibrous dysplasia, skin pigmentation and precocious puberty has the eponym McCune Albright Syndrome. Mazabraud Syndrome is fibrous dysplasia associated with soft tissue myxomas. The authors presenting a case of 12 yo girl with palpable mass in right orbital aspect of the skull. The patient is asymptomatic. Guided biopsy and radiological findings was typical of the lesion.

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bone lesion unmasked. *Am J Pathol* 151:1511-1515

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HEMICRANIA CONTINUA: A REMITTING FORM

M. Nicodemo, S. Cevoli, G. Pierangeli, D. Grimaldi, E. Sancisi, S. Zanigni, P. Montagna, P. Cortelli

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Background: Hemicrania continua (HC) is a strictly unilateral, moderate to severe, continuous primary headache associated with ipsilateral cranial autonomic symptoms. Characteristically this headache responds dramatically to therapeutic dose of indomethacin.

Case report: We describe a 42-year-old woman who presented with unilateral headache recurring since the age of 14 years. The pain, throbbing and severe, occurred always on the right temporal area and it was associated with ipsilateral lacrimation and ptosis, photophobia, phonophobia, osmophobia, nausea and vomiting. For the first 10 years the attacks, lasting 24-48 h, occurred each week-end for one or two months consecutively, followed by a remission periods of several months. From 24 to 26 years old, the headache was continuous, characterized by mild, right-sided pain with superimposed throbbing excruciating attacks associated with the usual vegetative symptoms recurring two or three times a day without therapy. After a period of two years without symptoms a severe continuous headache with the previous characteristics reappeared in bouts lasting 20-25 days continuously, two or three times every year usually in the same period. Neurological examination and magnetic resonance imaging were normal. Nonsteroidal anti-inflammatory agents, subcutaneous sumatriptan 6 mg and other oral triptans were completely unsuccessful. The patient took indomethacin 75 mg/day, for one month since the first day of a new attack, with an absolute response.

Conclusion: Diagnostic criteria of the International Headache Society (IHS) for hemicrania continua include a chronic, continuous, strictly unilateral headache of moderate intensity with intermittent exacerbations of severe pain associated with ipsilateral autonomic symptoms. A necessary feature is a prompt response to indomethacin. Our patient presented an headache fulfilling IHS criteria for HC only in a relatively short period of her headache history. Before and after this period the headache showed a remitting pattern, resembling migraine and cluster headache respectively. Remitting forms of HC were just described in the past, mainly as an initial stage of chronic forms. Our case report contributes to highlight the clinical variability of HC and suggest the necessity of include a chronic and a remitting form of HC in the next revision of the IHS classification.

THE "CEREBELLUM COGNITIVE AFFECTIVE SYNDROME": A CASE REPORT

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The "cerebellar cognitive affective syndrome" is a well defined clinical entity that could be diagnosed after a cerebellar lesion at the bedside and further quantified by neuropsychological tests. It is characterized by impairments of executive functions, impaired spatial cognition, personality changes and linguistic difficulties. The net effect of these disturbances in cognitive abilities appears to be a general lowering of intellectual function. In this report we describe PV, a 69 years old right-handed woman, who was admitted to our department about 15 days after a left cerebellar stroke for rehabilitative treatment. On admission, clinical examination revealed ataxia and impaired postural

righting reactions with left fall. Brain MRI demonstrated a subacute cerebellar ischemic lesion in the territory of the left posterior inferior cerebellar artery (PICA) and of the left anterior inferior cerebellar artery (AICA). Before stroke, PV was described as a tidy and a quiet person, while during her stay in hospital she developed inappropriate behaviours (indifference, perseverations, useless re-quests, disinhibited behaviours, blunting of affect) that limited rehabilitative treatment. Neuropsychological evaluation conducted about 60 days after stroke, demonstrated severely impairment in sustained and divided attention, visuo-spatial disorganisation, impaired visual memory, mild anomia and reduced verbal fluency. This case report confirms that the cerebellum is not only a motor control device but is an essential component of the brain mechanisms for personality, mood and intellect. Cognitive and emotional disorders are as important as motor deficits in cerebellar patients and they need to be precociously recognized in order to give a correct diagnosis and to program adequate rehabilitative treatment. During rehabilitation of a cerebellar patient, careful clinical observation could be supported by a detailed neuropsychological evaluation that allows recognizing, quantifying and monitoring any cognitive deficits with time.

NEUROPSYCHOLOGICAL INVOLVEMENT IN PRE-SYMPTOMATIC SCA2 PATIENT

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Background: Spinocerebellar ataxia type 2 is an autosomal dominant neurodegenerative disorder characterized by an expanded CAG trinucleotide repeats in SCA 2 gene resulting in abnormal polyglutamine sequence. It is a slowly progressive neurological disease which affect the cerebellum and its related pathways. The typical neuroradiological pattern is olivopontocerebellar atrophy (OPCA). Several studies reported on neuropsychological involvement during the course of the disease.

Methods: We report a 21-year-old woman with familial history of SCA 2 (father and sister), that asked to be submitted to the genetic test for SCA 2 in order to plan a pregnancy. DNA analysis, neuropsychological examination and brain MRI were performed.

Result: Neurological examination was normal. DNA analysis showed 39 CAG repeats in 12q 23-24.1 chromosome. Neuropsychological examination showed mild deficits of executive functions. MRI showed a mild increasing of the angle between pons and brainstem; morphometry evidenced reduction of the pons area (373 mm², n.v. >400); the linear measure of the middle cerebellar peduncle was reduced (6.2 mm n.v. - 8).

Conclusion: Our data showed that neuroradiological changes anticipate the onset of motor symptoms in SCA 2 and neuropsychological tests showed an early appearance of deficit of the executive functions.

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APHASIA AND RIGHT HEMIPARESIS AS PRESENTING SYMPTOMS OF A DEMYELINATING DISEASE WITH PSEUDOTUMORAL LESION

E. Peila, S. Masera, A. Romagnolo, A. Tribolo, A. Cicolin, P. Mortara, R. Mutani

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Introduction: Multiple sclerosis (MS) is one of the most common diseases of the central nervous system with a variety of clinical and radiological presentations. Aphasia is very rare as the initial symptom. Several cases have been reported of demyelinating processes mimicking a tumour of central nervous system; just one case has been described of aphasia and parietal syndrome as presenting symptoms of a demyelinating disease with pseudotumoral lesions.

Case report: A 38-year-old man was admitted with a 72-hour history of right hemiparesis and motor aphasia; the right hemiparesis receded spontaneously in 4-5 days, but aphasia kept on several weeks. Magnetic resonance imaging showed a lesion 3 cm in diameter in the subcortical left white matter, that was hyperintense in T2 and FLAIR with mild non homogeneous gadolinium enhancement and without perilesional edema. Evoked potentials have been performed: normal values in VEP and BAEP and bilateral prolonged central motor conduction time in median- and tibial-SEP has been found. The cerebrospinal fluid exam showed mild increased proteins (63 mg/dL), normal glucose value, 12 cell/mm³ (all mature T-lymphocytes) at the cell count and oligoclonal band of IgG at isoelectrofocusing. A systemic corticosteroid therapy has been performed with methylprednisolone 1000 mg for five days, with mild progressive improvement of language functions.

Conclusions: Giant pseudotumoral plaques are a rare MRI aspect of MS lesions; the suspect of a demyelinating tumor-like lesion must be valued in differential diagnosis with a CNS tumor, in particular in young adults. A bioptic study of the lesion is often necessary to perform a definitive diagnosis. Moreover Primary central nervous system lymphoma (PCNSL) can be associated with preceding 'sentinel' demyelinating pseudotumoral brain lesions.

A CASE OF TOXIC LEUKOENCEPHALOPATHY

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Toxic leukoencephalopathy requires 3 criteria for diagnosis: documented exposure to a toxin; neurobehavioral deficits and neuroradiologic abnormalities. Toxic leukoencephalopathy secondary to "chasing the dragon" was first described by Wolters et al. in 1982 and has been reported in multiple cases during the past 3 decades. The most distinctive manifestations are an alteration in mental status and, in contrast to disorders of the cortical grey matter, there is typically not a primary effect on language, praxis, or perception. Clinically, patients with toxic leukoencephalopathy secondary to drugs abuse (heroin or cocaine) use progress through 3 stages. Initial symptoms include motor restlessness, apathy, bradyphrenia, and cerebellar ataxia, which are followed by worsening cerebellar symptoms, hyperactive deep tendon reflexes, myoclonic jerks, and hypertonia with hemiplegia or tetraplegia in the second stage. In the final stage, the manifestations include stretching spasms, profuse diaphoresis, central fever, hypotonia and areflexia, and akinetic mutism. Not all patients progress through all 3 stages. We report a case of a male (24 years old) with subacute apathy, ataxia and bradyphrenia. MRI showed diffuse supratentorial lesions of white matter suggestive for toxic leukoencephalopathy. We found drug abuse by examination of the hair because the patient first refused and all blood and urine examination were negative. Other diagnosis were excluded. The outcome in despite of the severity of MRI alterations was good. An MRI examination performed one year later was normal. The aetiology of toxic leukoencephalopathy subsequent to drug use is poorly understood at this time. It has been postulated that mitochondrial dysfunction may play a role, in addition to direct toxicity to the myelin and/or axons, and that the lipid-rich myelin may "trap" the toxins, leading to further tissue dam-

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