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Renato Boeri

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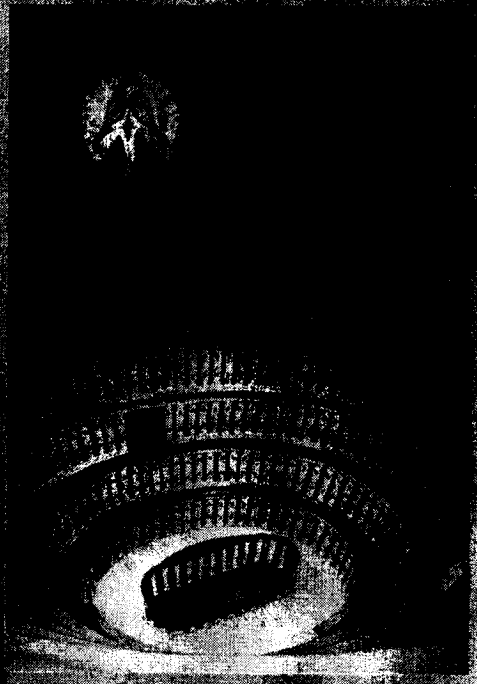

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Neurological Sciences

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noticed and sensory and cognitive functions were normal. Brain MRI, showed diffuse hyperintensity in the pons extending to the right middle cerebellar peduncle, where small foci of post-contrast enhancement were evident. Diffuse hyperintensity of the periventricular white matter and thickening of the pituitary stalk were also evident. Bone radiographs and technetium bone scan showed respectively osteosclerosis and increased uptake in long and facial bones. The patient was treated with corticosteroids but neurological symptoms did not changed. Neurological status worsened within next months and neurological examination revealed severe cerebellar syndrome and nystagmus. MRI was fairly unchanged and no post-contrast enhancement was evident at that time. Despite intravenous steroid pulses and chemotherapy the patient died 4 years after the onset of clinical symptoms. An autopsy was performed which showed typical ECD findings.

Discussion: Although neurological symptoms as the first clinical manifestations of ECD have been reported in less than one third of cases, for most patients the diagnosis of ECD is not made until the onset of neurological signs. MRI plays a major role in the diagnosis of ECD. Characteristic MRI abnormalities include diffuse hyperintense signal in the pons, midbrain and periventricular white matter. Spotted areas of enhancement following iv injection of gadolinium are usually present in the pons and in the white matter. This abnormalities pattern is to be considered highly specific and other possible mimickers such as neoplastic, vascular and infectious lesions as well as pontine and extrapontine and myelinolysis and PRES can be excluded on the basis of clinical data.

Conclusion: ECD is a rare disease and it is difficult to diagnose. When a brain involvement is present, such a diagnosis need to be considerate whenever a diffuse involvement of the brainstem, with spotted areas of post-contrast enhancement, is demonstrated by MRI, in a congruent clinical contest.

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LEUKOENCEPHALOPATHY AND IRREVERSIBLE COMBINED SPINAL CORD DEGENERATION IN A VEGAN SUBJECT

A. Romorini[1], G. Patruno[1], A. Panigati[2], P. Mantegazza[1]

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Objective: We describe a case of leukoencephalopathy and irreversible combined spinal corddegeneration in a young western vegan subject.

Methods: A 38 year old woman, member of a vegan cult for ten year, developed a confusional state with mild cognitive impairment, progressive severe paraparesis with loss of deep sensation in the lower limbs, impairment of superficial sensation of hands and feet and neurogenic bladder. Thyroid, liver and renal function was normal. Screening for the immunologicals disorders, neurological paraneoplastic syndromes, virus diseases, cerebral spinal fluid analysis and oligoclonal bands were negative. Laboratory evaluation revealed a low blood level of vitamin B12 with macrocytic anemia but no copper deficiency. An electrophysiological nerve conduction study showed a sensitive axonal neuropathy. Brain magnetic resonance revealed predominantly symmetrical bilateral periventricular high-intensity changes on T2 and Inversion recovery weighted images that did not enhance with gadolinium contrast. Cervical and dorsal spinal cord magnetic resonance disclosed increased signal on T2 weighted sections in the posterior and lateral columns without gadolinium enhancement.

Results: The patient was treated with rehabilitative and vitamin B12 therapy. After about two months neuropsychological examination was normal. Despite a normal blood level of vitamin B12, after about six months, the patient developed a spastic hypertonia with severe paraparesis and mild improvement of deep sensation of lower limbs and unable to walk.

Conclusion: In a strict vegan diet combined irreversible spinal cord degeneration is a very rare, but possible, neurological complication.

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POSTERIOR REVERSIBLE ENCEPHALOPATY SYNDROME IN A PATIENT WITH ARTHRITIS RHEUMATOID PRESENTING WITH TETRAPARESIS

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Introduction: Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey and colleagues in 1996. This is a clinical and neuroradiological syndrome characterized by headache, vomiting, altered mental status, blurred vision and seizures. Neuroimaging studies demonstrated white-grey matter edema involving predominantly the posterior region of the brain. We describe an atypical case presenting acute tetraparesis. We have taken in consideration the differential diagnosis, a review of the existing literature and the clinical and radiological features of PRES.

Case presentation: We report a 78-year-old woman affected by arthritis rheumatoid who developed posterior reversible encephalopathy syndrome presenting with an acute tetraparesis. She had been taking steroids per os (prednisone) for 15 years. The typical MRI features of PRES were recognized in the white matter of the bilateral posterior regions of the cerebral hemispheres and in the pons. In order to exclude a multifocal progressive leucoencephalopathy (PML), we performed real time PCR in cerebro-spinal fluid to detect DNA JC virus sequences, that was negative. We immediately stopped steroid treatment and observed a significant improvement in the MRI after two weeks, with a partial improvement of the neurological symptoms.

Discussion: We believe this is the first posterior reversible encephalopathy syndrome presenting with acute tetraparesis. We didn't find in literature other cases of PRES in arthritis rheumatoid. Due to the well-known reversibility of this pathology, we suggest to evaluate carefully this diagnosis in patients with hypertension, eclampsia, metabolic diseases or connective tissue diseases, taking steroids and/or immunosuppressive drugs.

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TICLOPIDINE-INDUCED AGRANULOCYTOSIS AND CHOLESTATIC HEPATITIS: A CASE REPORT

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Introduction: Ticlopidine is a platelet-inhibitor used to prevent thrombosis in patients with cerebrovascular or coronary artery disease. Because of its adverse effects, the use is reserved for patients in whom aspirin is contraindicated, not tolerated or where aspirin treatment fails. The most common side effects are mild and transitory: diarrhoea, dyspepsia, nausea and rashes. More serious, but less frequent, adverse effects are hematologic dyscrasia and cholestatic hepatitis. We report a case of agranulocytosis associated of cholestatic hepatitis probably related to the use of ticlopidine.

Case report: A 70-year-old woman was admitted to the Department of Rehabilitation because of gate ataxia after right bulbar stroke, which was happened ten days before. She had no previous history of hema-tologic or liver disease, alcol abuse, blood trasfusion. Her regular medications were aspirin 100 mg/d, atorvastatine 20 mg/d, amlodipine 5 mg/d. Immediately after stroke she discontinued aspirin and was started on ticlopidine 250 mg twince daily. On admission her blood test were normal. About four weeks later she developed agranulocitosis (neutrophil count was 100/μL) and marked liver disorder with elevated levels of alanine and aspartate aminotransferase, -glutamiltranspeptidase and alkaline phosphatase. Total and direct bilirubine and coagulation tests were normal. Serology tests for hepatitis A, B, C, Epstein-Barr virus, Cytomegalovirus, antinuclear and antimitocondrial antibodies and anti-smooth muscle autoantibodies were all negative. Cobala-mine and folate dosage were normal. Abdominal ultrasound showed steatosic liver without any evi-dence of common bile duct stone or biliary dilatation. Bone marrow aspirate showed myeloid matu-ration arrest, with decreased myeloid precursors and immature forms, ascribing to iatrogenic attack. Ticlopidine was immediately discontinued and was started aspirin 25 mg and dipiridamole 200 mg twince daily. She was treated with hematopoietic growth factors-granulocyte colony stimulating factor with a progressive growth of white blood count and a progressive normalization of liver test.

Discussion: The onset of haematologic dyscrasia is temporally related tn the initiation of ticlopidine therapy, generally occurring within the first 3 months, and the dyscrasia resolves within 3 weeks after discontinuation of therapy. The latent period between ticlopidine introduction and the appearance of hepatotoxicity is in the range of 2–12 week in most patients; upon discontinuing ticlopidine, symptoms and liver abnormalities usually resolved within 1–3 months, without any evidence of chronic hepatic injury. Besides complete blood cell count, periodic checks of liver function tests in the first one to three months following initiation of ticlopidine therapy may be recommended.

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NEUROSONOLOGICAL TECHNIQUES IN THE DIAGNOSIS OF BASILAR ARTERY (BA) STENOSIS. A SINGLE CENTRE CASE SERIES

G. Malferrari[1], M. Zedde[1], G. De Berti[2], M. Maggi[2], A. Dallari[1], A. Nucera[1], F. Nicoli[2], N. Marcello[1]

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Intracranial stenosis are a recognized cause of ischemic stroke in a significant rate of patients, both in asian-black people and in westem population. This condition is widely underdiagnosed and it is associated to a high recurrence risk and a poor response to treatment. Poster circulation lesions are traditionally related to a poorest outcome and a higher recurrence and fatality rate than anterior circulation ones. Among patients evaluated in our neurosonological laboratory by ultrasound examination of cerebroafferent vessels and intracranial vessel (by TCCS), we selected ten patients with BA stenosis of atheromasic origin. 6/10 patients had a symptomatic BA stenosis and 4/10 patients had an asymptomatic one. All patients underwent a neuroradiological examination too, all by brain MRA (coupled with MRI) and three patients also by a catheter angiography. Mean age of patients was 71.3 + 12.2 years (7 males and 3 females). A follow-up was performed within 12 months and one patient died by a sudden death of supposed cardiac origin, but two patients in the symptomatic group had a recurrence. Patients with BA stenosis often had a widespread intracranial involvement by atheromasic lesions, therefore multiple intracranial stenosis could be present and should be searched. Neurosonological examination with TCCS is a reliable and useful tool to diagnose BA stenosis and predict prognosis.

EPILEPSIA PARTIALIS CONTINUA AS FIRST MANIFESTATION OF DIABETES

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A 50-years-old man, right-handed was admitted because of recurrent focal seizures which had started 10 days before. His medical history was unremarkable except for a mild hypertension treated with low dose of ACE-inhibitors. There was familiarity for diabetes. At the neurological examination, the patient was awake and well oriented. No rigor neither intracranial hypertension signs were detectable. Seizures were characterized by emifacial left jerks with head and eyes turning to the left side sometimes followed by elevation and extemal rotation or clonic jerks of left arm and tonic extension of omolateral leg. Once we also observed secondary generalization with impairment of consciousness and urine loss. On admission laboratory exams showed high glucose level (299 mg/dL), increased CPK (517 U/L) and low K level (3 mEq/L). Serum osmolarity was normal (278 mOsm/kg) and blood-gas analysis didn't show acidosis. Autoantibodies were absent, so anti-TPO, anti-GAD and anti-ICA. EEG recording showed low-voltage alpha activity intermitted by slow activity with sharp-waves in right hemisphere; synchronously with jerks we observed discharges of spikes, polispike-waves in right temporal region spreading to the right emisphere and lasting 30 to 80 seconds. Brain CT scan, performed at the admission, didn't show abnormalities. Interictal brain MRI showed a slight right paratrigonal hyperintensity at DWI, while brain SPECT (Tc 99) was normal. On admission patient was treated with Lorazepam infusion (4 mg e.v. administered four times in 48 hours) and oral Levetiracetam (500 mg bid) without resolution of seizures. Only correction of hyperglycaemia with insulin and an appropriate fluid and electrolyte replacement resulted in a resolution of seizures within 60 hours. As previously published, seizures associated with hyperglycaemia are resistant to anticonvulsant treatment and respond best to insulin and rehydration.

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