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Possible clinical marker of first attack of sporadic HM and HaNDL (stroke-like syndromes) in emergency room: two case reports

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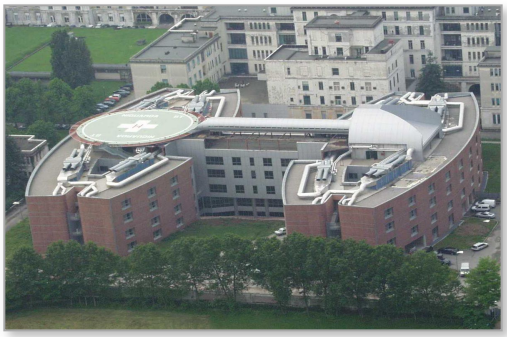
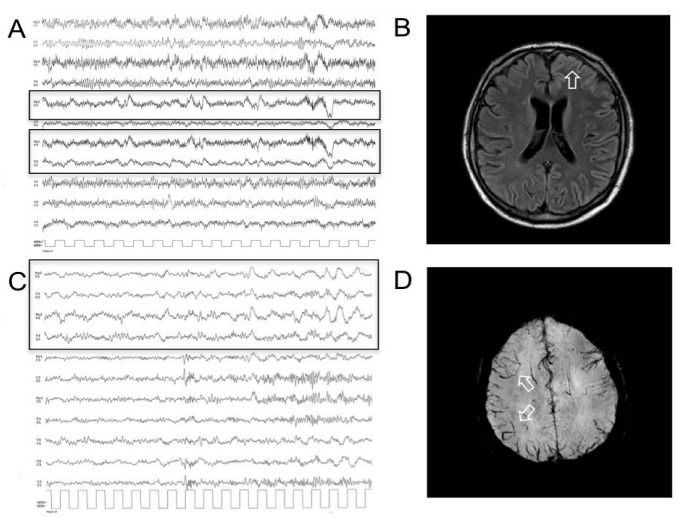


OBJECTIVE Hemiplegic migraine (HM) and headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL) are acute ischaemic stroke differential diagnosis.

CASE REPORT 1 On 7 June 2016 at 8.00 p.m., a 46 years old man, diabetic, with episodic tension type headache since one year, suddenly presented a confusional state, unknowing the road to home. After few minutes dysphasia, paresthesias from the right hand dating back to the elbow and moderate left pulsating migraine with nausea e vomiting appeared. At midnight in emergency room he presented right hemiparesis. Neurological examination showed dysphasia, slight right hemiparesis, right central facial nerve paresis and bilateral plantar flexion of cutaneous plantar reflex (CPR). Brain CT and CT-angiography were normal. On 8 June at 11.00 a.m. neurological examination showed stupor, aphasia, right flaccid hemiplegia and bilateral plantar flexion of CPR. The EEG showed left fronto-temporal cuspidate delta waves with slow pseudo-periodic trend (Fig. 1A); the brain CT control was normal. For a slight fever a lumbar puncture was performed with a protein increase (103.1 mg/dl). On 9 June the gadolinium brain MRI showed a minimal diffuse signal alteration at T2 sequences in the left frontal cortex (Fig. 1B) with negative DWI for acute ischaemic lesions. After 3 days he had a progressive neurological improvement with complete recovery in 5 days. After 2 weeks EEG and brain MRI were normal. Genetic test for familiar HM was negative. The patient was discharged with diagnosis of probably first attack of sporadic HM. After 5 months of follow up the patient was asymptomatic.

CASE REPORT 2 On 20 April 2014 at 11.00 p.m., a 29 years old man, without any pathology in medical history, arrived in emergency room for sudden onset of aphasia and right hemiparesis preceded by left migraine after a slight head injury without loss of consciousness. Neurological exam showed stupor, aphasia and slight right hemiparesis with bilateral plantar flexion of CPR. Brain CT and CT-angiography were normal. The CSF examination showed 50 lymphomonocytes with coltural exam and viral genomes negative. After two hours he had a progressive neurological improvement with complete recovery in two days. Other two episodes preceded by migraine happened after 8 days and 12 days until the first attack with resolution in 24 hours: one characterized by aphasia and right hemiparesis and the other by left hemiparesis and left homonym lateral hemianopsya. During the episodes bilateral CPR was normal and the EEG was asymmetric with slow pseudo-periodic trend in hemisphere affected (Fig. 1C). The MRI performed during the last attack with left hemiparesis showed a greater representation of the veins, hypointense in the Venous Bolt sequences on the right hemisphere. The patient was discharged with diagnosis of HaNDL. After two years the patients did not presented similar episodes and he was in good health (Fig. 1D).

DISCUSSION Patients with HM or HaNDL syndrome can arrive at the emergency room mimicking an acute ischaemic stroke (1, 2). The absence of acute vascular lesions at brain CT and MRI and normal CPR on the hemiparetic side exclude the ischaemic aetiology and the consequent procedure of intravenous thrombolysis. In our cases the encephalitic aetiology was excluded too for the normalization of the EEG. The CSF protein and lymphocytes increase were probably due to the alteration of the encephalic blood barrier secondary to the cortical spreading depression (CSD) associated with the migraine attack. The CSD cause a reversible cerebral cortical dysfunction characterized by a neuronal excitation with an increased excitatory glutamate release followed by a prolonged inhibition of neurological activity. Increased excitatory glutamate release caused a higher oxygen extraction fraction and hypointense veins on Venous Bolt or SWI sequences (3). This metabolic alteration, secondary to ATPase pump dysfunction, causes only a neuronal dysfunction and not a structural neuronal damage, as the vaso-occlusion absence demonstrates (1). For these reason MRI and EEG return normal and CPR is in plantar flexion on the hemiplegic side. The diagnosis of first attack of HM and HaNDL is a diagnosis of exclusion. After a complete diagnostic work up we proposed CPR in plantar flexion on the hemiplegic side like a clinical marker in emergency room to exclude the acute ischaemic stroke.



A) Case 1: during the attack EEG showed left fronto-temporal cuspidate delta waves with slow pseudo-periodic trend.
B) Case 1: after 2 days MRI showed a minimal diffuse signal alteration at T2 sequences in the left frontal cortex.
C) Case 2: during the third attack EEG was asymmetric with right slow pseudo-periodic trend.
D) Case 2: during the third attack MRI showed a greater representation of the veins, hypointense in the Venous Bolt sequences on the right side.

CONCLUSIONS Considering that in emergency room neuroradiological exams (brain CT and CT-angiography) are generally normal, the possibility to have a clinical marker like normal CPR in plantar flexion on the emiplegic side can be helpful in differential diagnosis between ischemic stroke and stroke mimics (HM and HaNDL) and to avoid intravenous thrombolysis with consequent bleeding risk. Other cases are necessary for support our hypotesis.

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