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IRCCS Istituto in tecnologie avanzate e modelli assistenziali in oncologia

Paroxysmal NonKinesigenic Dyskinesia (PNKD)

in a 27-year-old man patient with Multiple Sclerosis (MS) at clinical onset

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SUMMARY

PURPOSE: Paroxysmal dyskinesias (PxD) are rare involuntary and episodic movement disorders that can be precipitated by sudden voluntary movements (paroxysmal or acquired metabolic abnormalities. We describe a 27-year-old man patient with multiple sclerosis (MS) at clinical onset, presented with PNKDs
kinesigenic dyskinesias PKDs) or may occur spontaneously at rest (paroxysmal nonkinesigenic dyskinesias (PNKDs). Most cases of PxD are primary, categorized as either familial (usually autosomal dominant) or idiopathic. However in some cases a specific cause for the PxD has been identified, such as multiple sclerosis (MS), vascular lesions, trauma
METHODS: a 27-year-old man, admitted to the hospital because of weakness in the left arm, developed in the course of hospitalization short, unilateral dystonic attacks with preserved awareness, characterized by right inclination of the head with right face contraction, right wrist and elbow flexion with fingers extension and thumb adduction, right leg extension with fingers flexed, lasting about 50 seconds but frequent up to 100 per day. The patient underwent investigations including: video-EEG with polygraphic study; brain and spinal cord MRI; cerebrospinal fluid (CSF) examination; neuropsychological evaluation; somatosensory (SSEP), motor (MEP), visual (VEP) and acustic (BAEPs) evoked potentialsa 27-year-old man, admitted to the hospital because of weakness in the left arm, developed in the course of hospitalization short, unilateral dystonic attacks with preserved awareness, characterized by right inclination of the head with right face contraction, right wrist and elbow flexion with fingers extension and thumb adduction, right leg extension with fingers flexed, lasting about 50 seconds but frequent up to 100 per day. The patient underwent investigations including: video-EEG with polygraphic study; brain and spinal cord MRI; cerebrospinal fluid (CSF) examination; neuropsychological evaluation; somatosensory (SSEP), motor (MEP), visual (VEP) and acustic (BAEPs) evoked potentials
RESULTS: video-EEG recording (Fig. 2,3) 2 dystonic attacks confirmed the absence of epileptiform discharges during the episodes. Brain MRI (Fig. 1) showed: typical demyelinating lesions in the periventricular white matter and corpus callosum; lesions with enhancement after gadolinium were detected in multiple iuxta-cortical and subcortical regions (right frontal, biparietal, left precuneus and paratrigonal, left paracentral lobule, retrorolandic, mesial frontal and cingulate cortex) and in the brain stem (right cerebellar and middle cerebellar peduncle). Spinal cord MRI detected not active demyelinated cervical (C2-C3) lesions. IEF revealed oligoclonal IgG in the CSF. Neuropsychological evaluation was normal. SSEP, VEP and BAEPs were normal. MEP showed: central motor conduction time slowed at left and amplitude reduced at right arm. Intravenous steroid treatment improved left arm strength and PNKDs completely responded to carbamazepine 200 mg daily
DISCUSSION: difficult to define a clear anatomo-clinical correlation between PNKDs and the multiple iuxta-cortical and subcortical lesions in our patient
CONCLUSIONS: we suppose a neural network involving the iuxta-cortical left mesial frontal lesion

MRI

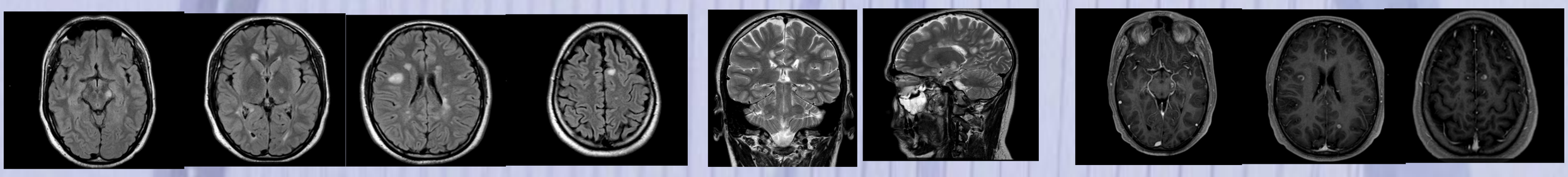


Fig. 1 C.L. 27 yrs MRI 2 May 2017

EEG AND VIDEO-EEG

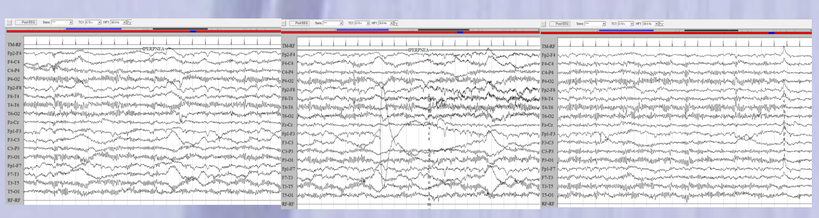


Fig. 2 C. L. 27 yrs EEG 8 May 2017

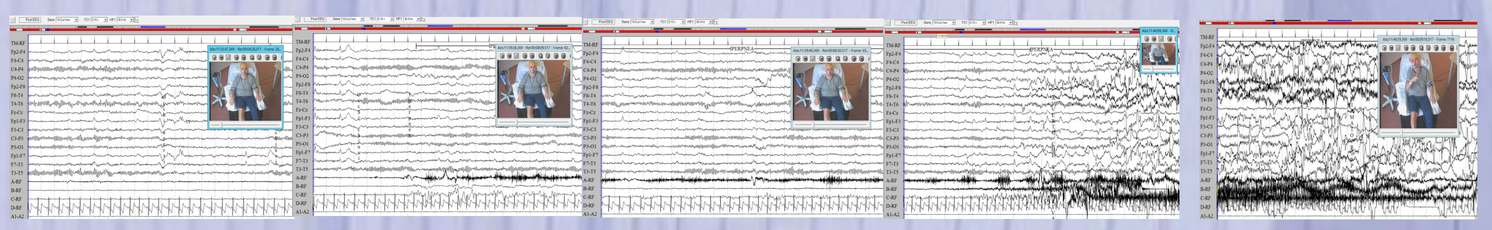
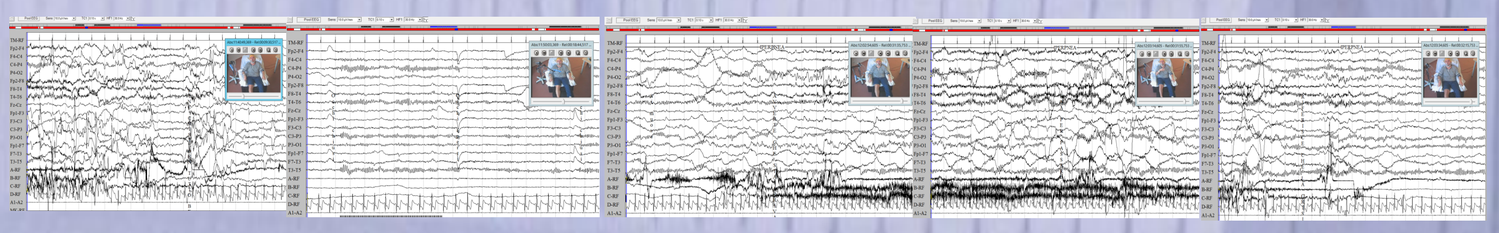


Fig. 3 C. L. 27 yrs Video-EEG 18 May 2017



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