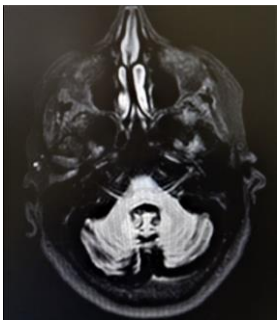
A case report of Cerebellar Cognitive Affective Syndrome diagnosed by accident in the Emergency Department

L. A. Y. Suemitsu[1], F. F. Graça[1], L.G. Piovesana[1], P. C. Azevedo[1]
[1]Dept. of Neurology, FCM, UNICAMP

Introduction: the role of cerebellum in non-motor functions is observed in patients with cerebellar damage from different acquired and non-acquired etiologies [1,4]. The Cerebellar Cognitive Affective Syndrome (CCAS) was first described in 1998 and characterized by executive dysfunction, impaired spatial cognition, personality change and language deficits. [2]. Topography and anatomy studies discovered that different areas of cerebellum act in diverse cognitive and affective domains depending on their connection to supratentorial areas. Posterior vermis is connected to the limbic system and modulates emotion. Posterior lobes of cerebellum are related to cognitive function [3,4,5,6].

Materials and Methods: this article is a case report of a patient in follow-up in the Department of Neurology of University of Campinas.

Results (case report): patient R.V. was hospitalized in April 2017 due to a traumatic brain injury (TBI) after she swerved to avoid being hit by a car and fell to the ground. In the CT scan, no hemorrhage was seen, but a chronic cerebellar atrophy was found. Patient presented a brief confusion state that resolved after one day. On neurologic exam cerebellar signs were present, patient had mild appendicular dysmetria and an incapacity of performing tandem-gait. Furthermore, patient had humor alterations: hyperthymia, with inadequate laughing and commentaries; and an accelerated speech. The patient had also language disturbance with mild agrammatism. Patient´s brother said the humor alteration was chronic. Clinical history included a TBI with 12 years old, after a fall from own height, while patient was running, cigarette smoking and dyslipidemia. Ambulatorial investigation was performed and MRI showed severe cerebellar atrophy predominantly in vermis and posterior lobes bilaterally. MOCA exam resulted in 18 points revealing dysfunction in executive tasks, abstract reasoning and language (patient had 11 years of education). CSF had no significant alterations. Only laboratorial abnormality was B12 vitamin dosage below 150 pg/ml, which was restored, with no change in clinical findings. Affective and neurological signs and symptoms plus CNS image are compatible with CCAS diagnosis.



Discussion and conclusion: this case report supports evidence of non-motor function of cerebellum. Vermis atrophy and posterior lobe atrophy of cerebellum are clearly visible in patient´s MRI image, and affective and cognitive symptoms may be related to these areas, respectively. The accidental diagnosis of the CCAS in this patient may rise concern about structural investigation in psychiatric patients. Besides, medical assistants should pay more attention to identifying emotion symptoms of patients with cerebellar diseases.

References: [1] Schmahmann JD, J Neuropsychiatry Clin Neurosci 16:367–378, 2004; [2] Schmahmann JD et al., Brain 121 (4):561-79, 1998; [3] doi:10.1016/j.cortex.2009.11.008; [4] doi:10.1093/brain/awm201; [5] doi:10.1093/brain/awr266; [6] Middleton FA et. al, J. Neurosci., 21(2):700–712, 2001;