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Case report

Brainstem arteriovenous malformation presenting with dyspraxic handwriting in a young girl

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Abstract

We report the case of a 11-year-old girl who developed an isolated hand-writing disorder with dysgraphia at the beginning of the school year in the sixth grade. A brain magnetic resonance angiography showed a round arteriovenous malformation sited in the left side of the midbrain extending to the ipsilateral medio-basal thalamus. Child neurologists should never neglect a thorough neurological evaluation in case of isolated worsening of handwriting, to rule out possible underlying organic causes. © 2013 The Japanese Society of Child Neurology. Published by Elsevier B.V. All rights reserved.

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1. Introduction

Two types of dysgraphia are so far recognized: the core ones, which reflect damage to the linguistic orthographic routes, and the peripheral ones produced by alterations in the selection or execution of graphic patterns [1].

Among the peripheral forms, one of the main functional networks responsible for the peripheral control of writing abilities may include the cerebellum, which not only maintains previously learnt writing processes but is also involved in the evolutionary acquisition of this ability, by modulating between supratentorial (premotor cortex) and peripheral proprioceptive afference during the ongoing handwriting movement.

Among the main peripheral causes of poor hand writing skills there is the tremor quite related to cerebellar dysfunctioning. Other causes of tremors such as dys-

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tonic, postural, psychogenic and essential need to be considered.

In this report we describe the case of a 11-year-old girl who came to our child neuropsychiatric facility for a consultation, urged by her school teacher because of dysgraphia manifested from the beginning of the school year in the sixth grade.

2. Case history

A 11-year-old, right-handed girl, was born to healthy, non consanguineous parents. She was born at term (38th gestational age), by cesarean section after an uneventful pregnancy. Her birth weight was 3860 g; she was the second of three healthy siblings. Psychomotor development, language skills and scholastic achievement were reported as normal. She was never affected by neurological or psychiatric conditions and was attending the sixth grade of secondary school.

About 6 months before, she had been hospitalized in a pediatric department for an episode of vomiting, drowsiness and nuchal rigidity associated with fine tre-

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mor in her right hand. Assuming a meningoencephalitis, a lumbar puncture showed a normal CSF, and antibiotics combined with steroids were given for about 3 days, when symptoms disappeared and the girl was discharged.

Approximately five months later, with the start of school attendance, her teacher reported an unusual writing, not appropriate to the age and level of education, so she urged pupil's mother to consult specialists. At admission, the neurological examination was essentially normal with not apparent neurosensory deficits, language disorders or other difficulties caused by hand dyspraxia. While reading ability was age-appropriate, handwriting displayed an increased amplitude of writing with an uncertain and irregular line in absence of misspellings (Fig. 1). Drawing of geometric figures and crenellated lines confirmed the dyspraxic handwriting. Fig. 1(d) shows also regular and normal amplitude writing at the age of 7 and 8 years. On the occasion of the same visit, a brain neuroradiologic assessment was programmed.

Five days after the specialist consult, a brain Magnetic Resonance Imaging (MRI) supplemented with MR Angiography (MRA) of the Willis circle, without gadolinium injection, was executed using a 1.5 T machine. It showed an arteriovenous malformation (AVM) with a diameter over 20 mm of the nidus (Fig. 2a). It was located in the left cerebral peduncle, involving the red nucleus and posteriorly the Werneking area. At the top the ipsilateral mesio-basal and antero-

ventral parts of the thalamus were also involved (Fig. 2b). The AVM had arterial feeders from the left posterior cerebral artery (Fig. 3a) and perhaps from the left anterior choroidal artery. Some ectatic peripheral veins drained toward the left basal vein of Rosenthal.

A marked reduction of the normal hyperintense signal of the left cortico-spinal tract (Fig. 3b), could correlate with a local damage of the left piramidal tract.

Four days after the neuroimaging study, the girl sustained an acute hemorrhagic event with severe worsening of neurologic condition including right hemiplegia and aphasia.

3. Discussion

The thalamic involvement in this girl brings us back to the few reported cases of adult patients with isolated thalamic agraphia with impaired grapheme formation and micrographia due to a lesion in ventral and ventro-posterolateral nuclei in the left thalamus or with pure agraphia or with alexia and agraphia due to left thalamic bleeding [2–4]. All these authors suggest a potential role of the thalamus in writing disorders with secondary negative effect of the lesion on the function of left cerebral cortex.

Some features of the handwriting in this girl, such as the increased letter height and the trembling writing, suggest the contribution of cerebellar dysfunction, as part of a larger cerebellar-thalamic-cortical (premotor

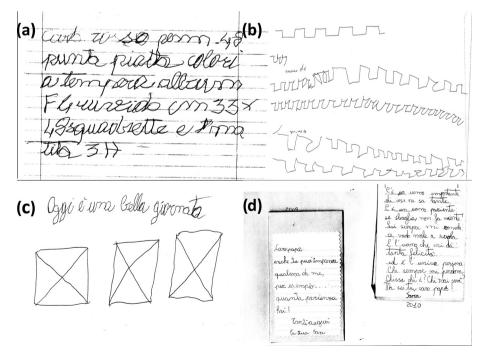


Fig. 1. (a) Increased amplitude of writing with an uncertain and irregular line in absence of misspellings. Geometric figures (b) and crenellated lines (c) confirming dyspraxic handwriting. (d) Regular and normal amplitude writing at the age of 7 (left) and 8 years (right).

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