Neurological Sciences (2019) 40:2653–2655
https://doi.org/10.1007/s10072-019-03983-4

LETTER TO THE EDITOR

Agenesis of internal carotid artery and deformation of caudate and nucleus accumbens

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Received: 19 March 2019 / Accepted: 13 June 2019 / Published online: 25 June 2019

Dear Editor,

Agenesis of the internal carotid artery (ICA) is a rare neuroimaging finding. Certain insults during the embryologic period or genetic predisposition may result in agenesis of the ICA (AgICA) [1]. Although accompanied deformations in the brain have been reported in a few cases, subtle structural abnormalities in the brain were not detected on routine radiologic evaluations in some cases [2]. Herein, we report the case of a patient with unilateral AgICA and deformation in the subcortical structures.

A 50-year-old female visited the out-patient department with complaints of weakness and recurrent cramping of left arm and leg. She experienced persistent mild clumsiness in her left arm and leg, and since the past few years, the clumsiness transiently worsened at instances when she gets physically exhausted. More frequently, brief jerky movements of the left arm and leg were observed particularly during sleep. Eighteen months ago, she underwent total thyroidectomy and parathyroidectomy due to hyperparathyroidism. She reported that her father had a congenital anomaly of the cervical region, including the thyroid gland. Her father suffered from right hemiplegia owing to unknown brain disease 20 years before he died.

Neurologic examination revealed normal results. No definite motor weakness and sensory loss were found. Mild rigidity was noted in her left arm and leg. Moreover, mild bradykinesia was observed based on left finger and foot tapping examinations.

Magnetic resonance cerebral angiography revealed agenesis in the right ICA. The right middle cerebral artery (MCA) was connected to the left ICA via cavernous anastomosis. The right MCA was also connected with the right posterior communicating artery (Fig. 1A). Brain magnetic resonance imaging (MRI) revealed that the volume of the bilateral caudate bodies and tails was slightly increased (Fig. 1B). Gross abnormalities in the pituitary gland were not observed. Electroencephalogram findings during the inter-ictal period demonstrated no abnormal findings. Laboratory test results were normal, including normal serum calcium level, except for the low thyroid-stimulating hormone level (0.02 uIU/ml) compatible with the use of synthetic thyroid hormone.

To quantitatively assess the morphology of her brain, brain MRI results were statistically compared with those of 20 age-matched women (mean age = 51.1, standard deviation = 2.99) using voxel-based morphometry (VBM) and vertex analysis. The controls were recruited from the MRI database of the author's institution. The institutional review board (IRB) approved the retrospective use of MRI data (IRB approval number: GCH2018003). VBM was performed using SPM12 (Welcome Department of Imaging Neuroscience, London, UK, www.fil.ion.ucl.ac.uk/spm/) implemented in MATLAB R2011a (Mathworks, Inc., Sherborn, MA). Gray matter was segmented from the T1-weighted images of the recruited individuals. Segmented gray matter images were normalized into a standardized space. Thereafter, two-sample *t* test was performed to compare the controls with the patient. The shape of the subcortical structures of the patient's brain was further assessed using FMRIB's Integrated Registration and Segmentation Tool (FIRST), which is part of the FMRIB's Software Library (FSL 5.0.8, http://www.fmrib.ox.ac.uk/fsl). The subcortical structures, including the putamen, caudate, thalamus, and globus pallidus (GP), were segmented on the brain MRI scans of the patient and controls. Vertex analysis using the FIRST tool was performed to assess the differences

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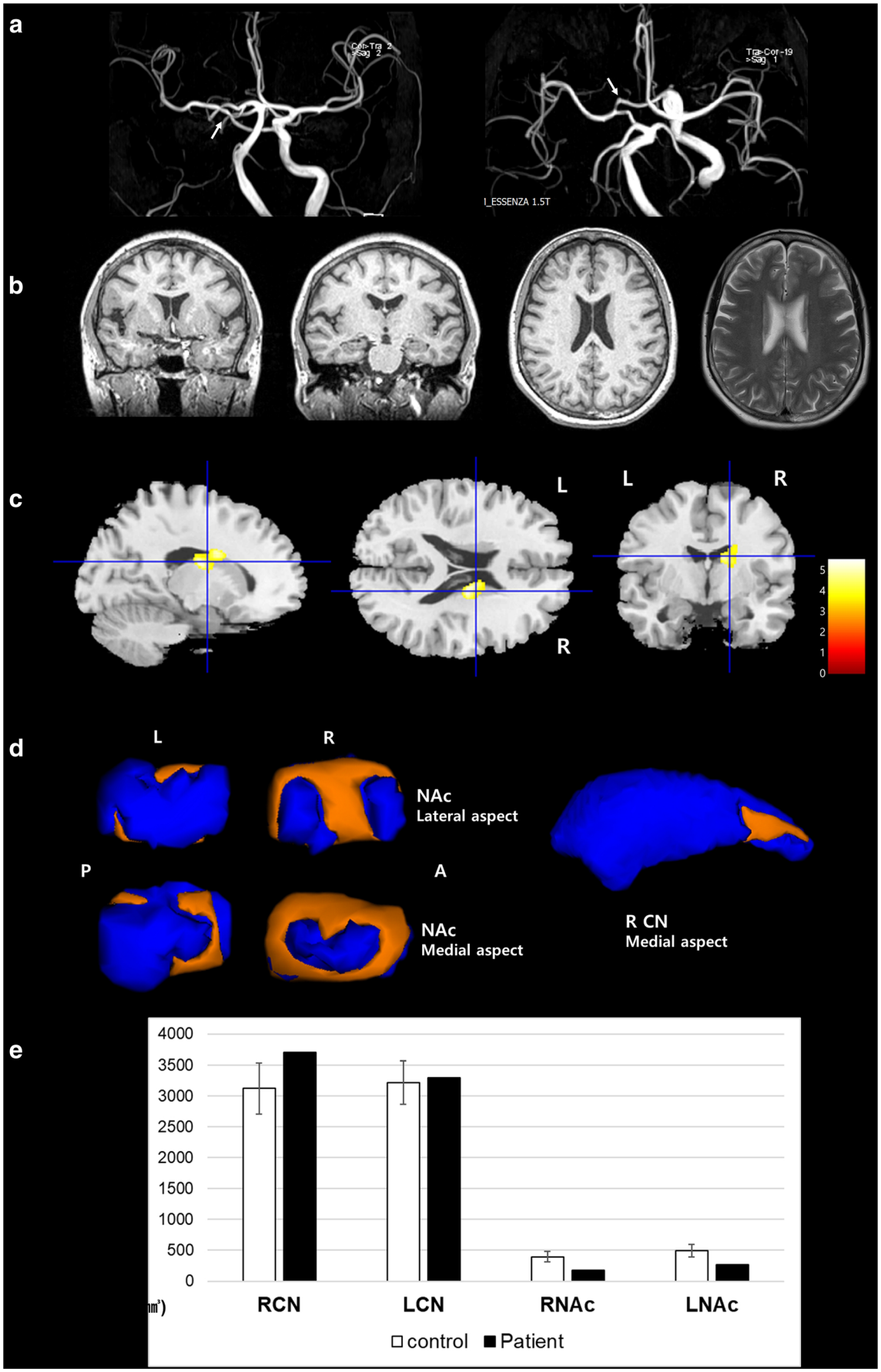
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◀ **Fig. 1** (A) The right internal carotid artery (ICA) is not visible on magnetic resonance angiography. The right middle cerebral artery is supplied by the left ICA via cavernous anastomosis (white arrow). (B) Brain magnetic resonance imaging showed that the volume of the bilateral caudate bodies and tails slightly increased. (C) Significant results of voxel-based morphometry are displayed on a normalized space. A cluster with a significantly larger gray matter volume in the body and tail of the right caudate nucleus (CN) was observed in the patient's brain. The height threshold was set at $p < 0.001$ (uncorrected). Multiple comparison problems were corrected at the cluster level using false discovery rate with a corrected p value of < 0.05, and the figure was displayed based on neurologic convention. (D) The comparison results (controls versus the patient) of the vertex analysis are displayed (orange: significant shape deformation of the patient's brain). A significant shape deformation in the tail of the right CN and bilateral NAc was identified. A p value of < 0.05 was considered statistically significant (corrected for multiple comparisons [family wise error, FWE] using the cluster-wise approach). (E) When the total volumes of the right CN and bilateral NAc were quantitatively measured, the volume of her right CN was higher than the standard deviation of the controls' right CN, whereas the volume of her bilateral NAc was lower than the standard deviation of the controls' bilateral NAc

in the shape of the segmented subcortical gray matter structures between the controls and patient.

The volume of the body and tail of the right caudate nucleus (CN) significantly increased based on VBM (Fig. 1C). A significant shape deformation in the tail of the right CN and bilateral NAc was also found via vertex analysis (Fig. 1D). There were no significant abnormalities in the shape of the bilateral amygdala, hippocampus, putamen, thalamus, GP, and left CN based on vertex analysis. When the total volumes of the right CN and bilateral NAc were quantitatively measured, the volume of the patient's right CN was higher than that of the controls, which exceeded the standard deviation, whereas the volume of the patient's bilateral NAc was less than the standard deviation of the controls (Fig. 1E). The patient's total gray matter volume (TGV: 593300.00 mm^3) and total intracranial volume (TIV: 1274900.00 mm^3) were within the standard deviation of the controls (TGV: 612810 ± 82,097.50 mm^3, TIV: 1307120 ± 46,948.94 mm^3).

Unilateral AgICA is commonly an accidental finding [1, 2]. In few cases, patients present with cerebral ischemic symptoms, seizure, or ill-defined headache and dizziness [1, 2]. In this case, the patient presented with recurrent left arm/leg motor disturbances and extrapyramidal signs correlated with right-sided AgICA.

In few cases, AgICA is associated with the gross deformation of the central nervous system, such as midbrain atrophy [3], optic nerve atrophy [4], and Chiari I malformation [5]. Subtle abnormalities might not have been identified in previous cases. Deformation of the CN and NAc in the present case could be attributed to degenerative changes from recurrent ischemia or epileptic phenomenon considering that chronic ICA occlusion is significantly associated with progressive brain atrophy [6]. However, normal total gray and white matter volume, absence of ischemic lesion in the brain, and family history of unknown congenital anomaly indicate that the deformation was more likely to be a consequence of deterioration during the embryonic period due to genetic predisposition or external insults.

The deformation in her brain was associated with the extrapyramidal signs on the left side of her body. However, the current study had some limitations because it only included a single case. Further studies must be conducted to determine the clinical significance of the deformation in subcortical structures in AgICA.

Compliance with ethical standards

Conflict of interest The author declares no conflict of interest.

Informed consent Informed consent was obtained from the participant included in the study.

Statement of human rights The study has been approved by the Ethics committee of Chung general hospital and has been performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

References

1. Afifi AK, Godersky JC, Menezes A, Smoker WR, Bell WE, Jacoby CG (1987) Cerebral hemiatrophy, hypoplasia of internal carotid artery, and intracranial aneurysm. A rare association occurring in an infant. Arch Neurol 44:232–235
2. Vasovic L, Trandafilovic M, Vlajkovic S, Radenkovic G (2018) Congenital absence of the bilateral internal carotid artery: a review of the associated (ab)normalities from a newborn status to the eighth decade of life. Childs Nerv Syst 34:35–49
3. Dhiman R, Gorimanipalli B, Swamy DR, Sharma S, Garg A, Saxena R (2018) Congenital third nerve palsy associated with midbrain hypoplasia due to bilateral segmental internal carotid artery agenesis. J Neuroophthalmol 38:483–485
4. Garcia-Medina JJ, Del-Rio-Vellosillo M, Fares-Valdivia J, Aleman-Romero L, Zanon-Moreno V, Pinazo-Duran MD (2017) Optic nerve hypoplasia and internal carotid artery hypoplasia: a new association. Can J Ophthalmol 52:e173–e177
5. Fitzgerald RT, Zuccoli G (2012) Agenesis of the internal carotid artery: associated malformations including a high rate of aortic and cardiac malformations. Pediatr Radiol 42:1333–1338
6. Muller M, van der Graaf Y, Algra A, Hendrikse J, Mali WP, Geerlings MI, SMART Study Group (2011) Carotid atherosclerosis and progression of brain atrophy: the SMART-MR study. Ann Neurol 70:237–244