CLINICAL CASE - TEST YOURSELF | Adult neuroradiology

Atypical combination of gastrointestinal and neurological clinical symptoms

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PART A

History: A 57-year-old man was admitted to our Emergency Department with symptoms including ataxia, vomiting, dysphagia, left-sided hemiparesis, hemi-hypesthesia and Horner's syndrome, right palatal paresis, lower limb weakness, paresthesia on the left side of the body and face, and right central facial palsy. The patient had a history of chronic arterial hypertension, two earlier brain ischemic strokes (in 2005. and 2016.), and long-lasting type 2 diabetes mellitus. As part of the diagnostic evaluation, a brain MRI was performed.

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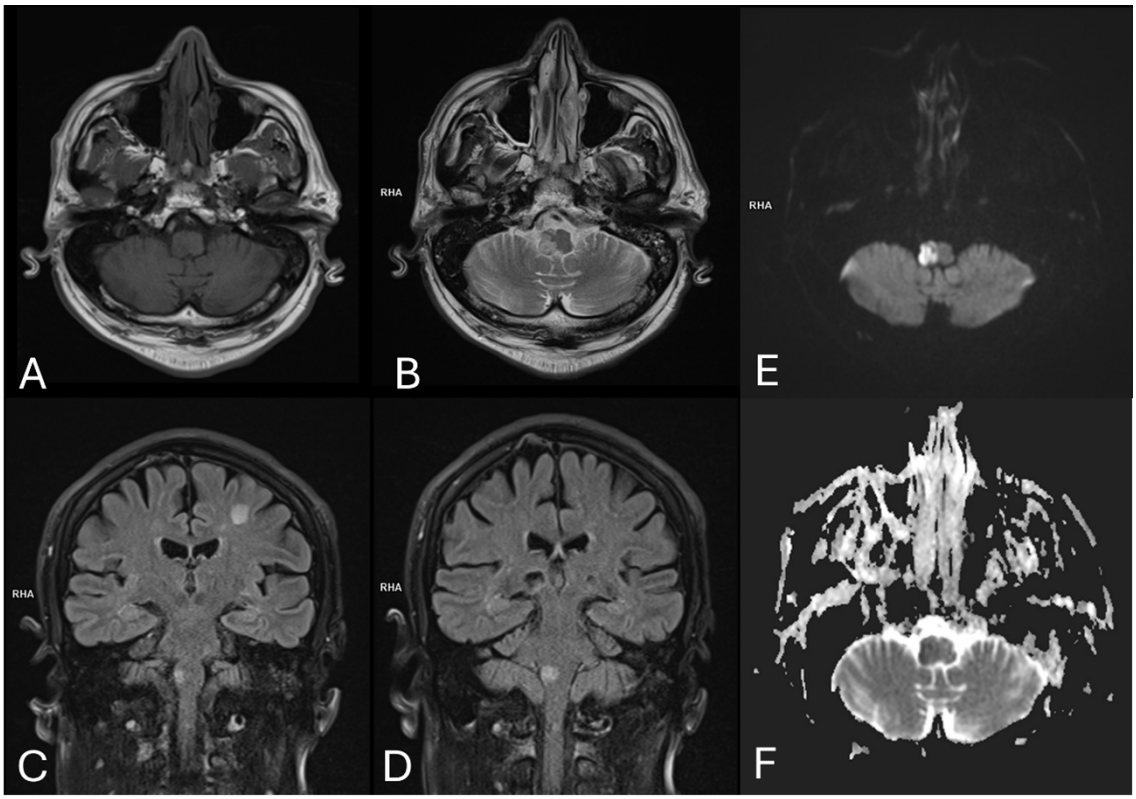

Figure 1. *A. T1W, axial section. B. T2W, axial section. C, D. FLAIR sequence, coronal section. E, F. B1000 DWI, and ADC maps*

PART B

Diagnosis: Wallenberg syndrome due to acute ischemic changes located in the right half of the medulla.

History: Wallenberg syndrome, also known as lateral medullary syndrome or posterior inferior cerebellar artery (PICA) syndrome, is associated with various clinical and neurological symptoms due to the damage of the lateral segment of the medulla, posterior to the inferior olivary nucleus [1,2]. It was described by Adolf Wallenberg in 1895 and is the most typical posterior circulation ischemic stroke syndrome in clinical practice by its presentation [1,2]. The arterial lesions of the lateral medullar syndrome are brain stem infarcts mainly caused by atherothrombotic vertebral and basilar arteries that supply the PICA, followed by dissections and embolisms (most commonly as a complication of long-standing hypertensive arteriosclerosis) [1]. In particular cases, the syndrome was described to be associated with syphilitic vascular changes, primary tumors, secondary metastases, or encephalitis in the region of the dorsolateral medulla oblongata [1].

Diagnosis: We report a rare case of acute transient Wallenberg's syndrome with a dominant atypical gastrointestinal clinical presentation including vomiting and dysphagia, and discuss its mechanisms and clinical-radiological correlation. A 57-year-old man was admitted to the Emergency Department with symptoms including ataxia, vomiting, dysphagia, left-sided hemiparesis, hemi-hypesthesia and Horner's syndrome, right palatal paresis, lower limb weakness, paresthesia on the left side of the body and face, and right central facial palsy. The patient had a history of chronic arterial hypertension, two earlier brain ischemic strokes (in 2005 and 2016), and long-lasting type 2 diabetes mellitus. His current symptoms appeared mainly during physical activity and lasted from five to thirty minutes. Deviations in laboratory biochemical values were: fibrinogen 4.3 g/L, glucose level 27.7 mmol/L, creatinine values 91 µmol/L, total cholesterol levels 7.08 mmol/L, LDL cholesterol 5.3 mmol/L, sodium 131 mmol/L, CRP 8.8 mg/dL. Brain computerized tomography (CT) and CT angiography showed chronic lacunar ischemic changes in the brain tissue, right vertebral artery hypoplasia in its V4 segment, and mild stenotic changes of vertebrobasilar circulation arteries. Furthermore, brain magnetic resonance imaging (MRI) including diffusion-weighted (DWI) imaging was performed. It showed typical MRI changes for acute ischemia in the right half of the medulla with the T2W and FLAIR sequence hyperintensities and diffusion restriction on DWI (Figure 1). MRI examinations were performed on a 1.5T MRI machine (Siemens, Erlangen, Germany), and an 8-channel head coil was selected. Transverse axial T1WI, T2WI, FLAIR, sagittal T1WI, and diffusion-weighted imaging (DWI) were routinely scanned. The dispersion sensitivity coefficients (b-value) were 0 s/mm^2 and 1000 s/mm^2 respectively and the acquisition matrix was 512 × 512, the FOV was 23 × 23 cm, the layer thickness was 5.5 mm, the layer spacing was 1 mm, and the deflection angle (FA) was 90°.

Magnetic resonance imaging (MRI imaging) is far superior imaging modality compared to CT in visualizing medullar lesions (MRI can clearly depict the infarcted area, but CT rarely delineates this small medulla area mainly due to reduced sensitivity and artifacts from adjacent bony structures). MRI with DWI is the best optional diagnostic test to detect and/or confirm the infarct located in the lateral medulla. The infarcted area as a hypercellular change has a high B1000 DWI signal and a low ADC map signal. Clinicians and radiologists should keep in mind that in approximately 15-20% of subjects, associated concomitant cerebellar infarcts are present (as a result of PICA circulation vascular compromise). More often, the infarct is usually limited to a limited area of tissue in the dorsal lateral medulla, as a consequence of the occlusion of direct VA branches that irrigate this vascular territory.

Main differential clinical-radiological diagnoses of lateral medullar infarcts (Wallenberg syndrome) are medial (Dejerine) and hemimedullary (Reinhold) medulla oblongata syndromes, the Babinski-Nageotte and Cestan-Chenais syndromes, Opalski syndrome, Tapia syndrome, other classical pontine stroke syndromes (Marie-Foix syndrome, Millard-Gubler syndrome, locked-in syndrome, Brissaud-Siccard syndrome, facial colliculus syndrome, Gasperini syndrome, Raymond syndrome, etc.), caudal fastigial nuclei inactivation,

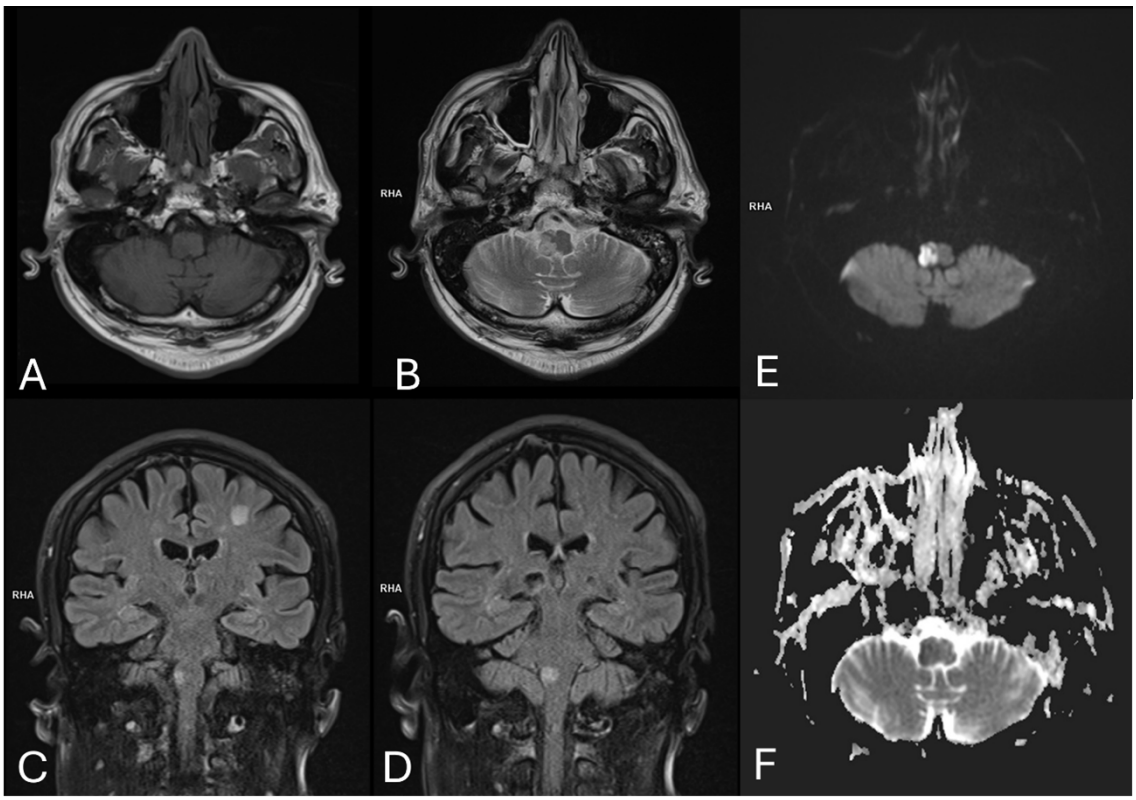

Figure 1. A. T1W, axial section, an inhomogeneous area iso-to-hypointense compared with the surrounding tissue, located in the right half of the medulla. B. T2W, axial section, a hyperintense area in the right part of the medulla. C, D. FLAIR sequence, coronal section, an inhomogeneous area hyperintense compared with the surrounding nervous tissue, vaguely discerned, located in the right half of the medulla. E, F. B1000 DWI and ADC map showing diffusion restriction in the right half of the medulla – acute ischemic changes.

acute demyelinating plaques (most commonly multiple sclerosis), pontine neoplasm (diffuse intrinsic pontine glioma being most common), pontine hemorrhage, central pontine myelinolysis, osmotic demyelination syndromes, pontine infarction, pontine metastasis, vigabatrin toxicity, etc. Clinical-radiological diagnosis of Wallenberg syndrome should be always made in a correlation between different radiological (CT, MR, CTA, MRA, perfusion analysis, etc.) and clinical data (patient history - anamnesis and clinical symptoms – so-called pontine stroke specific symptoms). In addition to previous patients with Wallenberg syndrome can have a wide variety of clinical symptoms (motor hemiparesis, sensorimotor hemiparesis, ataxic hemiparesis, balance difficulty, double vision, swallowing difficulty, dizziness, numbness, dysarthria-clumsy hand syndrome, loss of coordination and sensation, nausea, difficulty articulating words – slurred speech, vertigo – spinning sensation, etc.) due to different neural centers anatomically located in the pons.

Furthermore, according to the study conducted and published by Kim JS (to the very best of our knowledge one of the biggest studies examining lateral medullary syndromes), MR angiograms performed in 130 consecutive respondents with pure lateral medullary infarction detected vertebral artery (VA) disease in 67% and posterior inferior cerebellar artery (PICA) disease in 10% (the presumed pathogenetic mechanisms included large vessel infarction in 50%, arterial dissection in 15%, small vessel infarction in 13%, and cardiac embolism in 5% - mainly in patients with isolated PICA disease) [3]. Previous findings have very important clinical implications, practically meaning that in subjects with medullary infarcts vertebral arteries should be detail scrutinized and examined in routine clinical examination.

Hemodynamic instability due to the arterial vessel damage from chronic arterial hypertension and type 2 diabetes mellitus, combined with the right vertebral artery hypoplasia could lead to acute transient Wallenberg's syndrome in our patient. Neurological insult affecting neuronal networks for swallowing in the brainstem (infarction of the swallowing centers in the rostral dorsolateral medulla which occurs in lateral medullary infarction including nucleus ambiguous) could lead to severe dysphagia and at the very best of our knowledge, only a few cases with Wallenberg syndrome presenting with dominant atypical gastrointestinal clinical presentation have been reported in the literature so far [4,5]. Dysphagia is a potentially important clinical manifestation mainly because it could be related and further complicated by aspiration pneumonia, malnutrition, immunosuppression, increased morbidity and mortality, and prolonged hospitalization [5,6]. Dysphagia mostly has a good clinical prognosis in Wallenberg syndrome, however, it has been reported in some cases that it was associated with a very poor prognosis requiring tubes even for years, implying the versatility of this complex syndrome [5]. In line with that, *Nakao M.* et al. published a retrospective, observational, multicenter study with 35 subjects and concluded that lateral medullary infarction impairs the sequence of swallowing events [7]. Our case could widen the knowledge of this complex syndrome and bring a new vision about a possible topography causing such a complex clinical presentation (combination of neurological and gastrointestinal clinical symptoms). It could benefit clinicians of different specialties (radiologists, neurologists, cardiologists, and gastroenterologists) so that diagnosis and treatment for patients with similar clinical manifestations could be promptly recognized. **R**

KEY WORDS | Wallenberg syndrome, medulla oblongata, MRI, dysphagia, and lateral medullary syndrome

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