Letter to the Editor

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Before Diagnosing CHANTER Syndrome, All Possible Differential Diagnoses Must Be Carefully Excluded

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We read with interest the article by Pandit et al. [1] about a series of three cases of cerebellar, hippocampal, and basal nuclei transient enemas with restricted diffusion (CHANTER syndrome). In patient-1 (23 year-old female), CHANTER syndrome manifested clinically as upstream hydrocephalus requiring suboccipital craniotomy and external ventricular drain placement [1]. Patient-1 also suffered cardiac arrest on hospital day-2 but was successfully resuscitated. CHANTER syndrome in patient-2 (60 year-old female) presented with headache and arterial hypertension, which were treated with intravenous antibiotics and mechanical ventilation [1]. Patient-3 (55 year-old female) suffered from seizures, arterial hypertension, and coma and was treated with naloxone, mechanical ventilation, and implantation of a percutaneous endoscopic gastrostomy [1]. Although this study is impressive, some points require further discussion.

First, research on the causes of the clinical presentation was inadequate for all three cases. Coma in patient-1, headache, arterial hypertension, and cerebral hypoxia in patient-2, and coma, seizures, and arterial hypertension in patient-3 require larger studies than previously reported. In this context, cerebrospinal fluid studies, electroencephalography recordings, information about cerebral perfusion MR angiography or CT angiography, MR venography, digital subtraction angiography, and MR spectroscopy are required in all three patients. In patients with headache at initial presentation (patient-2), it is imperative to rule out subarachnoid bleeding (SAB), reversible cerebral vasoconstriction syndrome, venous sinus thrombosis (VST), cerebral vasculitis, encephalitis, meningitis, seizures, and trauma. In patients with impaired consciousness (patient-1, patient-3), it is essential to rule out nonconvulsive status epilepticus, acute disseminated encephalomyelitis, acute hemorrhagic, necrotizing encephalitis, stroke, hyperglycemia/hypoglycemia, cardiac arrest, liver or kidney failure, and acidosis, in addition to SAB, VST, vasculitis, encephalitis/meningitis, and epilepsy. In patients with arterial hypertension (patient-2, patient-3), multifocal posterior reversible encephalopathy syndrome (PRES) must be excluded. In patients with seizures (patient-3), an epiphenomenon indicating a post-convulsive image abnormality must be excluded. Hypoxic brain injury must be excluded in patients with a history of cardiac arrest (patient-1) or respiratory failure (patient-2).

Second, patient-1 underwent a cerebral MRI after a suboccipital craniotomy [1]. The authors ruled out the possibility that the abnormalities observed on MRI were simply side effects of the operation and were independent of opiate intoxication.

Third, patient-3 did not undergo a cerebral MRI, making it difficult to verify the imaging abnormalities described in the case description.

Fourth, central nervous system complications of SARS-CoV-2 infection were not ruled out [1]. Because the three cases appeared to have been diagnosed during the pandemic, it is imperative to document that RT-PCR for SARS-CoV-2 was negative upon admission and that a history of recent anti-SARS-CoV-2 vaccination was negative.

Fifth, the indications for levetiracetam administration in patient-1 were not specified [1]. There was no evidence of seizures or a history of epilepsy. It is important to know why patient-1 received levetiracetam after surgery.

Sixth, the treatment and outcomes of three patients were inadequately described. Of particular interest are the

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antiepileptic treatment in patient-3 and the long-term outcomes of all three patients.

In summary, before drawing conclusions from the present cases, clinical genetic testing of the parents and other first-degree relatives should be performed, and the pathogenicity of the variant should be confirmed. Before CHANTER syndrome is diagnosed, differential diagnoses must be thoroughly ruled out.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Author Contributions

Conceptualization: Josef Finsterer, Investigation: all authors. Validation: all authors. Writing—original draft: Josef Finsterer. Writing—review & editing: Sounira Mehri.

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