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Hemiplegic migraine associated with Usher syndrome: a case report

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Introduction

Migraine is a common and disabling primary headache disorder. Migraine with aura is characterized by recurrent attacks of fully reversible visual, sensory, or other central nervous system symptoms that usually develop gradually and are followed by headache and associated migrainous symptoms. When aura includes motor weakness, the condition is classified as hemiplegic migraine. Usher syndrome is a rare autosomal recessive ciliopathy characterized by sensorineural hearing loss, retinitis pigmentosa, and sometimes vestibular dysfunction.

Case presentation

A 58-year-old woman presented with chronic migraine with aura since age 14. In 2005, she experienced paraplegia lasting 24 hours. In 2009, she developed recurrent focal neurological deficits, including paraplegia and hemiparesis, also lasting 24 hours. Between 2011 and 2012, further focal deficits occurred, resulting in persistent left-sided hypoesthesia. Between 2016 and 2018, she experienced headache episodes associated with aphasia and left facial paralysis. Her medical history included Usher syndrome (retinitis pigmentosa and left-sided sensorineural hearing loss), hypertension, dyslipidemia, myocardial infarction (2004), fibromyalgia, eosinophilic asthma, severe obstructive sleep apnea, irritable bowel syndrome, nutcracker esophagus, temporomandibular joint disorder, hepatic steatosis, Ménière's disease, and probable bilateral metachronous type 4 Ménière's disease. Family history was unknown, although her son had migraine. Neurological examination revealed mild divergent strabismus and reduced visual acuity. Brain MRI was unremarkable. Cervical spine MRI demonstrated cervical disc disease with an extruded C7–T1 disc herniation. MRI angiography showed a developmental venous anomaly in the right frontal lobe. Based on the recurrent reversible motor deficits and absence of structural lesions, a diagnosis of hemiplegic migraine was established. Current treatment includes lamotrigine, clopidogrel, and onabotulinumtoxinA every three months. Previous treatments included valproic acid, amitriptyline, nortriptyline, gabapentin, duloxetine, and topiramate. The patient currently has satisfactory headache control with botulinum toxin therapy.

Discussion

Migraine is frequently associated with comorbid conditions at rates higher than those observed in the general population. The coexistence of migraine and Usher syndrome appears exceptionally rare; a literature review identified only one previous case report describing this association. Recurrent focal neurological deficits in this patient were initially interpreted as cerebrovascular events but were later recognized as manifestations of hemiplegic migraine due to complete or near-complete symptom recovery and the absence of structural brain lesions.

Conclusion

This case illustrates a rare coexistence of hemiplegic migraine and Usher syndrome, highlighting the importance of considering hemiplegic migraine in patients with recurrent transient neurological deficits and normal neuroimaging findings.

Keywords:
Migraine
Hemiplegic migraine
Usher syndrome

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