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A Rare Cause for Migraine with Aura

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Letter

We report a 32-year-old anxious male who presented to us with history of intermittent throbbing unilateral headache for past 7 years. Headache was episodic and usually lasts for 6 to 24 hours. It was usually associated with nausea, vomiting, photophobia and phonophobia. Headache was relieved by sleep and analgesics. Most episodes of headache were preceded by visual aura. He was diagnosed as having migraine with aura but was not on any preventive medications for the same. Patient consulted us for his headaches. We got an interesting history from the patient which prompted us to evaluate him in detail. His father also had migraine at a young age and has suffered two ischemic strokes before the age of 45. Now his father has progressive cognitive decline and has been diagnosed of dementia. On examination, we did not find any focal neurological deficits.

Migraine is a common disabling neurological disorder in about 10-20% of general population. Migraine often runs in families and shows complex genetic traits. Clinical and genetic heterogeneity as well as the influence of environmental factors have hampered the identification of the gene responsible for migraine disorder [1]. Familial Hemiplegic migraine (CACNA1A gene mutations), Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) (Notch 3 mutation) and Mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS) are disorders which present with migraine and have a specific gene mutation. Based on history of patient we suspected CADASIL as there was a positive family history. We evaluated the patient with MR imaging of brain which showed diffuse periventricular white matter T2/FLAIR hyperintensity. The earliest and most frequent abnormalities are areas of increased signal on T2 /FLAIR appearing as punctiform lesions, in periventricular areas and in the centrum semiovale, which later become diffuse and symmetrical. Involvement of the external capsule and the anterior temporal lobes is highly suggestive of CADASIL but often occurs late in course of disease [2]. Simona Sacco, et al. analysed 34 studies on CADASIL which included 749 patients. 51.7% patients had presented with headache and among them 92% (356) had migraine [3]. Genetic testing is indicated if the patient has a characteristic clinical syndrome in combination with characteristic neuroimaging or a positive family history with no hypertension. NOTCH3 has 33 exons but all CADASIL mutations occur in exons 2-24, which encode the 34 EGFR (epidermal growth factor). Screening of these 23 exons that encode the 34 EGFR has 100% specificity and 90% specificity for exons 2-6 [4]. The need is more debatable if a patient without a family history has only migraine with aura and a few hyperintense signals on T2-weighted imaging. Unless there is a specific request from the patient, genetic testing is not indicated in such cases as white-matter abnormalities are common in migraine and up to 30 years can elapse between the onset of migraine and the first stroke or onset of dementia. As per patients request we proceeded with genetic testing. He was offered genetic counseling before the test and was positive for NOTCH 3 gene mutation sequenced from exon 2-6.

Diagnosis of CADASIL requires strong clinical suspicion because of varied presentation. Clinical and MRI alone may not help in confirming diagnosis and genetic testing although not widely available is the gold standard for diagnosis. Diagnosis is important for patient as well as family members for genetic counseling. The interesting feature of our case was he neither had characteristic MRI nor clinical features but we could make diagnosis by molecular genetics.

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