Shunting the Diagnosis: A Case of Cavernoma Induced Hydrocephalus and Secondary Cluster-Like Headaches

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Abstract

Cavernomas are thin dilated vascular channels that lack smooth muscle or neural tissue. In this case report, the patient had headache attacks that resembled cluster headaches and cavernomas causing obstructive hydrocephalus requiring surgical intervention. Her cluster headaches did not improve with resolution of her hydrocephalus, but responded to treatment with verapamil and high flow oxygen. This case suggests that cavernomas may serve as a structural cause of secondary cluster headaches, and secondary cluster headaches can at times be responsive to primary cluster headache treatments like verapamil and high flow oxygen.

The intent of this study is to demonstrate cavernomas as potential structural causes for secondary cluster headache.

Keywords: Cluster Headache; Cavernomas; Hydrocephalus

Introduction

Cavernomas are thin dilated vascular channels that lack smooth muscle or neural tissue. Their incidence is 0.3–0.5% and is usually discovered incidentally in young adults. The most frequent presentation is seizure, but can present with focal deficits or headaches. Two case reports detailed a tectal cavernoma causing hydrocephalus requiring shunting and subsequent surgical resection [1-2]. There is also a recent case report of a 71 yr man who presented with cluster-like headaches that were thought to be related to a cavernous angioma in the left pontine area [3]. To our knowledge, we are not aware of other reports of cavernomas causing hydrocephalus and secondary cluster-like headaches. There are reports of other secondary causes of cluster-like headaches, including other vascular causes like aneurysms and thrombosis [4]. Cluster headaches are one of the primary headache disorders with a distinct phenotype. When the phenotype of cluster headaches occurs in the setting of a secondary disorder, they are considered secondary headaches. According to the ICHD-3beta, cluster headaches involve unilateral, orbital or supra-orbital severe pain that lasts from 15-180 minutes at a time along with autonomic features, which can include ipsilateral lacrimation, rhinorrhea and/or conjunctival injection [5]. This is a case report on a patient with cavernomas presenting with cluster-like headaches. The patient in this report consented to allowing us to publish the case and all identifying data was removed. According to ICHD3 beta criteria for secondary headaches, a causal relationship can be hypothesized when headache improves after treatment of the underlying lesion and/or when headache develops in temporal relation to the onset of the presumed causative disorder [5]. The intent of this study is to demonstrate cavernomas as potential structural causes for secondary cluster headache.

Case Report

A 36 year old woman with a history of right sided hemiplegia and hemisensory deficits of unclear etiology for as long as she can recall presented with neck pain and falls. An MRI of the brain revealed two cavernomas in the left thalamus extending to the midbrain and tectal region (Figure 1). She denied any family history of cavernomas or headache disorders. Two years later, she developed frequent nausea and headaches without any previous history of headaches. A repeat MRI demonstrated obstructive hydrocephalus, and a ventriculoperitoneal (VP) shunt was placed with resolution of hydrocephalus and improvement of nausea. Unfortunately, there was no improvement of headaches.

At onset, she was experiencing 3-4 headaches per day that occurred around the same time daily. They were located around the left temporal/parietal area, and individual headaches lasted about 45 minutes. The headaches were throbbing with a 7-10/10 intensity. They involved photophobia, phonophobia, nausea, vomiting, and cutaneous allodynia. During attacks she felt restless, and was unable to find a comfortable position. There was associated left unilateral eyelid drooping, left sided congestion, and rhinorrhea, but no other autonomic symptoms. Her neurological examination was unremarkable except for trace weakness of the right arm and leg, as well as trace decreased sensation on the entire right side of the body. After VP shunt placement, she was later started on a trial of verapamil, and reached a dose of 360 mg daily. On this dose of...
verapamil, her headache frequency improved to 1 headache per
day with a 5/10 intensity. For abortive therapy, she used oxygen
at 10 liters per minute (LPM) with a face mask, which typically
terminated a headache within 10 minutes.

Conclusions

Headaches with cluster-like features have been known to occur
in the setting brain tumors, infections, arterial dissections, and
pituitary lesions [4]. The patient’s cavernomas caused obstructive
hydrocephalus requiring surgical intervention. Initially, it was
unclear whether her hydrocephalus and/or cavernomas were
contributing to her headaches with cluster features. Subsequent
resolution of her hydrocephalus after VP shunt placement did not
lead to improvement of her cluster headache symptoms.

In the case of this patient, there is no specific treatment for
cavernomas. The cavernomas have likely been present since birth,
and contributed to her right sided hemiplegia and hemisensory
deficits. Her cavernomas may have also contributed to her
headaches later in life. It is possible however that one or both of her
cavernomas bled, which may have played a role in the development
of her hydrocephalus and later her cluster headaches, but due to
timing, the bleeding may not have been captured on MRI studies.

The trigemino-vascular system is thought to play a role in cluster
headaches; therefore the location of her cavernomas point towards
it being a cause [3]. In addition, this case suggests that verapamil
and oxygen can be effective treatments for secondary cluster headaches.

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