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BRIEF REPORT

# Cluster headache associated with moyamoya

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**Abstract** A 34-year-old man with right-sided cluster headache presented with a stroke from right-sided moyamoya. Following surgery on the right, both moyamoya and cluster headache remitted, but eighteen months later a cluster attack and symptoms of cerebral ischemia from moyamoya recurred on the left. Again, following surgery on the left, both moyamoya symptoms and cluster attacks disappeared. Cluster headache secondary to moyamoya has not previously been described.

**Keywords** Cluster headache · Moyamoya

## Introduction

Moyamoya disease is a chronic cerebrovascular occlusive disease characterized by stenosis or occlusion of the terminal portions of the bilateral internal carotid arteries and particular involvement of the circle of Willis and the arteries that feed it. The term moyamoya (Japanese for "puff of smoke") describes the appearance on angiography of abnormal collateral vascular networks that develop adjacent to the stenotic vessels. It may result in ischemic attacks, cerebral infarction, or cerebral hemorrhage. Headache is a common symptom, but these have until now uniformly been described as migrainous (Goadsby, Sakai, personal communications). Cluster headache secondary to moyamoya has not previously been described.

## Case report

A 34-year-old-man with cluster headache since age 18 had had cluster periods occurring for 4 months each year, usually in spring but occasionally during the autumn instead, characterized by three to seven right-sided cluster attacks per day that were ten out of ten in pain intensity, and associated with rhinorrhea, lacrimation, and ptosis. During an attack he would roll around on the floor, compulsively pound his head, and scream short phrases such as "why me?" Mostly nocturnal, the attacks lasted 1.5–2 h if untreated, but responded well to oxygen in 20 min and sumatriptan within minutes. Prophylactic medications such as ergotamine, propanolol, methysergide, verapamil, and divalproex were all ineffective, however. During a cluster period, sleep deprivation led to reassignment of duties when he was in the Army and decreased job performance in civilian life, but did not otherwise impact his personal relationships or academic performance. He has never had a fundoscopic exam. He has smoked 1.5 packs a day since age 19 and drinks a 12-pack of beer a week, but has never used any illegal drugs except for occasional marijuana prior to 1988 when he joined the military.

At age 34, he presented with unilateral left-sided restless legs syndrome (RLS) and numbness in the fingers

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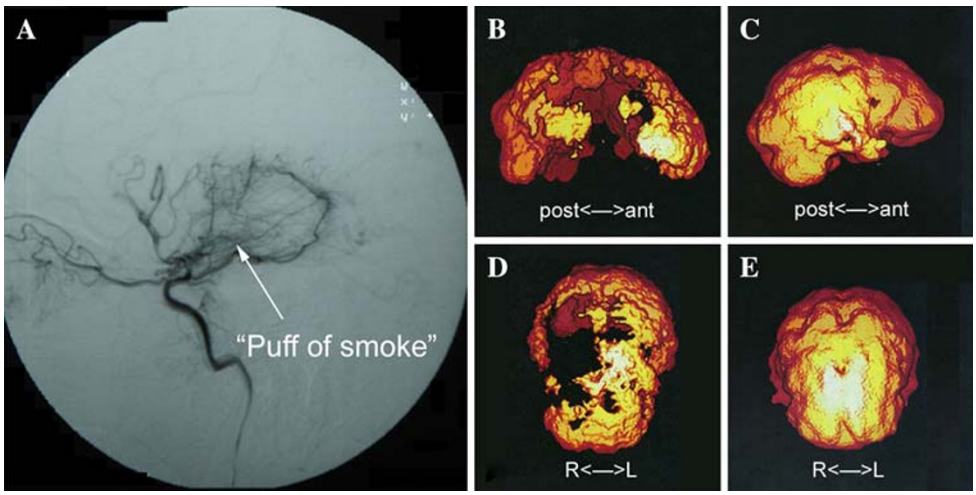


**Fig 1** **a** Lateral view from a selective right internal carotid angiogram shows occlusion of the supraclinoid ICA with "*puff of smoke*" from basal collateral vessels. **b, d** 3-D volume rendered SPECT images prior to surgical intervention demonstrate profound hypoperfusion in the right middle cerebral arterial distribution. **c, e** Images obtained following STA–MCA bypass show virtually normal and symmetric perfusion

of his left hand that was permanent in one finger but fluctuated throughout the day in the others. He was diagnosed with a stroke from moyamoya (Fig. 1a). Following a right-sided superior temporal artery (STA) to middle cerebral artery (MCA) bypass three months later, his cerebral perfusion improved (Before: Fig. 1b, d; After: Fig. 1c, e) and his RLS and cluster attacks remitted. Eighteen months later, however, he experienced his first, solitary cluster attack on the *left*—all previous attacks having been on the right—followed shortly thereafter by return of his RLS and finger numbness, this time on the right. Imaging confirmed a worsening of his left-sided moyamoya, and he underwent a left STA-MCA bypass (video 1). Six years later, he has had no further cluster attacks.

## Discussion

Although correlation does not imply causation, the temporal and lateral concordance of both cluster headache and moyamoya symptoms are consistent with such a relationship. Twice, cluster headache symptoms occurred contralateral to the moyamoya symptoms (in the ipsilateral hemisphere), and each time, cluster headache symptoms remitted with successful treatment of the moyamoya.

Secondary, or symptomatic cluster headache has been described from pituitary adenoma [1, 2], meningiomas of the sphenoid wing [3], upper cervical area [4], and undersurface of the tentorium cerebelli [5] (where inflammatory myofibroblastic tumor has also caused cluster headache) [6], aneurysms of the anterior cerebral [1], posterior cerebral [7], vertebral [8], and carotid arteries [1], dissections of the vertebral [9] and internal carotid arteries [10, 11], arteriovenous malformations in the middle cerebral artery [12] as well as occipital [13], frontal, temporal, and parietotemporal lobes [14–16], Wallenberg's syndrome [17], pituitary orbitosphenoidal aspergillosis [18], head and neck injury [19], ipsilateral enucleation [20], calcified lesions close to the third ventricle [21], cavernous sinus granulomatous tissue [22], and post carotid endarterectomy [23, 24].

Most symptomatic cases are from lesions located near the midline in the middle fossa of the skull base, although lesions involving structures that are innervated by branches of C2 can trigger cranial autonomic activation in humans [25] and animals [26], and C2 activation can sensitize trigeminal neurons that receive input from incracranial vessels [27]. Disturbance of sympathetic nerve function, either in the plexus of parasympathetic, sympathetic, and sensory fibers in the connective tissue of the cavernous sinus/hypophyseal region or in the walls of the internal carotid artery is known to generate cluster headache, and it is possible that the progressive intimal thickening in the carotid walls of this patient and consequent sympathetic disruption exacerbated his attacks.

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**Conflict of interest** None.

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