Cluster like headache in an elderly patient with lateral medullary infarct — does the clue lie somewhere else?

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Abstract

Cluster headache is a relatively uncommon primary headache. The exact aetiology of cluster headache is yet unknown. There are rare case reports of cluster like headache in patients who have had vascular insults, either in the form of a dissection or an ischaemic infarct. The case of a 73 year old man is presented, who had a transient ischaemic stroke with dizziness, vomiting, left leg weakness and non-specific occipital headache that resolved in one day. Two days later, he developed features of partial Wallenberg syndrome which was confirmed on magnetic resonance imaging. One day after the onset of Wallenberg syndrome, he developed typical features of cluster like headache ipsilateral to the stroke, site. The headache was treated with traditional therapy of cluster headache including high flow oxygen and verapamil. The patient responded well to the treatment. This case suggests a possible link of lateral medulla to cluster like headache etiology and further emphasizes that semiology of cluster headache can be secondary to central lesions.

Keywords: Cluster headache, Lateral medullary infarct, Wallenberg syndrome, Stroke and headache

Introduction

Headache is a common but underemphasized feature of lateral medullary syndrome. Previously, headache has been reported in 54 to 76% of patients with Wallenberg syndrome.[1] The headache described by patients is usually non-specific and may or may not occur on the side of the site of the stroke. Symptomatic cluster headache (CH) cases have been described in association with different kinds of lesions located in the middle cranial fossa, near the sellar or parasellar structures including lesions of internal carotid artery.[2] Involvement of the hypothalamus has been suggested to explain the cyclic aspects of cluster headache.[3] There is one reported case of continuous cluster like headache (CLH) associated with lateral medullary infarct.[1] Headache mimicking cluster like headache has also been

reported with vertebral artery dissection.[4] We report the case of an elderly man with episodic (CLH) ipsilateral to the side of lateral medullary infarct. Complete remission of symptoms occurred with usual treatment of CH including verapamil and high flow oxygen.

Case Report

A 73 year old man with prior history of diabetes, and hypertension presented to emergency room with one day history of headache, dizziness, vomiting and left leg weakness. Headache was sudden in onset, more in the occipital region and gradually increased in severity. It was associated with 10 to 12 episodes of vomiting which were non projectile, containing food particles with no blood. He simultaneously developed left leg weakness which improved within few hours. There was no history of loss of consciousness, urinary or bowel incontinence and speech problems. On arrival to emergency room his blood pressure was 160/90 mm Hg; pulse 90/minute and regular. On examination there was weakness of left leg with a power of 4/5 and he was dragging the left leg while walking. Rest of the neurological examination including higher mental functions, cranial nerves, deep tendon reflexes and sensory examination were normal. His plantars were down going bilaterally. CT scan of head without contrast showed old subcortical strokes with no evidence of acute haemorrhage or infarct. The weakness improved and headache resolved till next day. After two days, he developed ataxic gait with tendency to fall towards left side. MRI brain showed tiny acute infarct in left posterolateral medulla (Figure-1a and 1b). A diagnosis of partial Wallenberg syndrome on left was made. His EKG and echocardiography were normal.

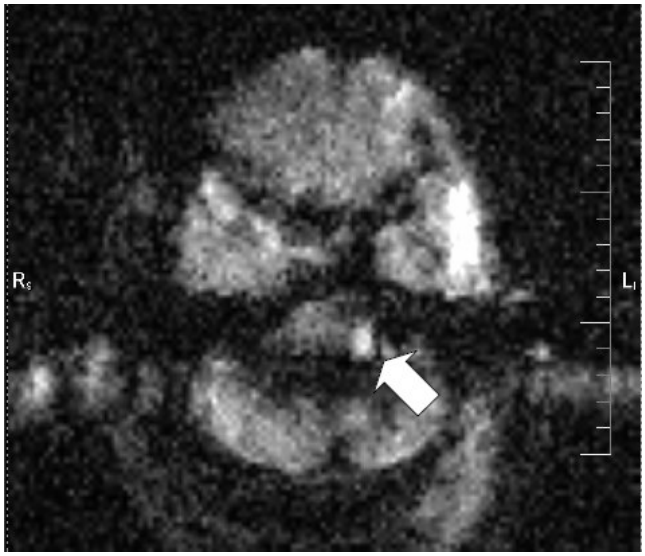

Figure-1a: Magnetic resonance diffusion weighted imaging (DWI) of brain showing acute infarct in left lateral medulla.

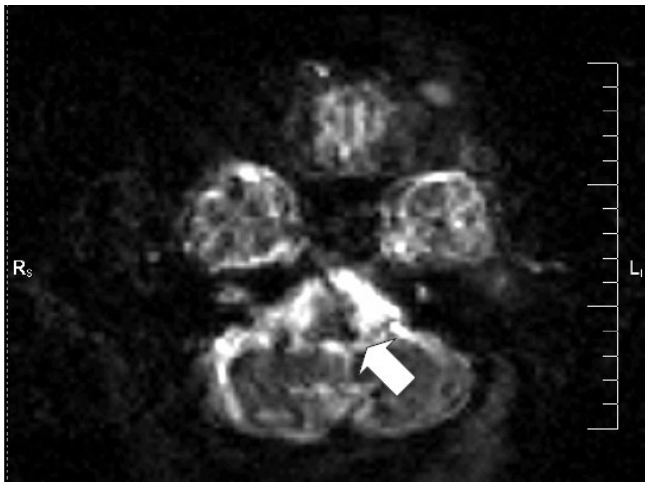

Figure-1b: Apparent diffusion coefficient (ADC) map imaging showing low signal in left lateral medulla corresponding to high signal on DWI.

One day after the onset of partial Wallenberg syndrome, he developed a new type of headache. It was sudden in onset, episodic, unilateral on the left side of face lasting for 10 to 30 minutes. It was severe in intensity and was associated with pain and watering of the left eye. He had 4 to 5 episodes of similar headache in a day. A diagnosis of CLH was made. His acute attacks were managed with 100% oxygen by facial mask, whereas he was started on verapamil 80 mg twice a day for prophylaxis of his headaches. Initially he showed partial response to oxygen but as the dose of verapamil was increased to 160 mg twice a day, the headaches decreased in frequency and rapidly responded to oxygen therapy. He was discharged on secondary stroke prophylaxis and verapamil. At one month follow-up his headaches had resolved completely and verapamil was gradually tapered off. At one year follow-up he had no recurrence of stroke or headache, and had no residual neurological deficits.

Discussion

According to international classification of headache disorder, 2nd edition (ICHD-II) the diagnosis of CH requires at least 5 attacks of severe or very severe, unilateral orbital, supraorbital and/or temporal pain that last for 15 to 80 minutes, occur once every other day to 8 times a day, and are accompanied by ipsilateral conjunctival injection/lacrimation, or nasal congestion and/or rhinorrhea, or eyelid oedema or forehead and facial swelling, or miosis and/or ptosis, or a sense of restlessness or agitation.[5] It affects young adults, predominantly males.[3] Treatment of CH with high flow oxygen inhalation at the time of symptom onset results in rapid pain relief and has been proven in randomized, placebo-controlled trial.[6] The mainstay in the prophylactic treatment of cluster headache

is high-dose verapamil ranging from 360 to 720 mg.[7]

It is considered that CH is a type of neurovascular headache. The exact mechanism and site of origin of headache is debated. The pathophysiology of CH is imperfectly understood and treatment has so far been mainly empirical.[8] The individual attack involves activation of trigeminal-autonomic reflex.[9] Activation of hypothalamus has been associated with CH.[3] Neurostimulation of hypothalamus has resulted in successful treatment of this condition.[11] Central disinhibition of trigeminal nociceptive system has also been suggested to be one of the mechanisms involved in CH.[10] The complexity of the pathophysiological mechanisms and possible involvement of multiple areas of brain make it unclear as to what is the exact site of origin. Our patient did not have functional neuroimaging like PET scan or functional MRI, however, conventional MRI with FLAIR and DWI sequences did not show any acute lesion in the hypothalamus.

With vertebral artery dissection CLH has previously been reported.[4] This suggests a possible relationship between medullary strokes and CLH and also supports the idea that the semiology typical of CH can be secondary to pure central lesion located in the lateral medulla.[1]

The patient in our case, with no prior history of headaches developed CLH following lateral medullary infarct which suggests that there might be some trigger zone in lateral medulla which leads to the initiation of CH. There was no lesion found in hypothalamus in our case on conventional MRI which leads to the assumption that there are other areas in brainstem that may contribute to the pathophysiology of CH.

High flow oxygen is the mainstay of treatment of acute attacks of CH.[6] The acute attacks of our patient responded very well to high flow oxygen. The response to this therapy also essentially excluded other causes of unilateral headache like paroxysmal hemicrania, hemicrania continua and short-lasting, unilateral neuralgiform pain with conjuctival injection and tearing (SUNCT syndrome), which do not respond to oxygen.[11]

Conclusion

After lateral medullary infarct CLH is a rare but important association which suggests a possible trigger zone in lateral medulla.

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