Central neurogenic hyperventilation (CNH) is characterized by sustained tachypnea inspite of an elevated arterial PaO₂, pH and a low arterial PaCO₂. CNH is common in patients with brainstem injury accompanied by a decreased level of consciousness but this also has been described in some alert patients with an invasive brainstem tumor. We report one case with CNH resulting from a unilateral pontine infarction. His consciousness was clear, and CNH spontaneously disappeared without any medication.


Key Words: Central neurogenic hyperventilation, Unilateral pontine infarction

Central neurogenic hyperventilation (CNH) is seen in some cases of brainstem injury which usually shows a decreased level of consciousness. This syndrome also has been described in awake patients who have invasive brainstem tumor. We describe a patient with unilateral pontine infarction showing an episodic CNH during wakefulness.

Case

A 61-year-old man was admitted to our hospital for sudden onset of episodic dyspnea with tachypnea. One day prior to the admission, he developed two episodes of hyperventilation which persisted about 20 minutes without any precipitating factor and resolved spontaneously. He also developed mouth angle deviation and dysarthria which resolved spontaneously with hyperventilation. On the admission day, those episodes recurred five times. One episode occurred during sleep. There was no peripheral stimulations to respiration and specific causative drug medication.

Physical examination revealed fast (40/min), regular and shallow breathing which persisted about 20 minutes and spontaneously normalized (18/min). The chest auscultation during shallow breathing showed clear lung sound. The cardiac auscultation showed no specific murmur or irregular rhythm and there was no neck vein distension. He was alert. Speech was mildly sluggish. His eyes showed conjugate deviation to the left side and his face showed a peripheral type facial nerve palsy in the right side. Motor, reflex, sensation, and gait were all normal. Initial ABGA showed elevated pH (7.734), PaO₂ (122.9 mmHg) and decreased PaCO₂ (6.8 mmHg). After 20 minutes, follow-up ABGA showed normal range. The brain MRI showed an unilateral pontine infarction.
involving right paramedian pontine tegmentum (Fig. 1). MRA showed a diffuse stenosis in proximal and middle segment of basilar artery. The high resolution chest CT scan (Fig. 2) and perfusion lung scan (Fig. 3) showed normal findings. Serum D-dimer measurement was done to diagnose pulmonary embolism and it was normal range. Pulmonary function test (PFT) was normal. And transthoracic echocardiography showed normal heart configuration.

**Discussion**

CNH is characterized by sustained tachypnea in spite of elevated arterial PaO$_2$ and a low arterial PaCO$_2$. This is a rare clinical encounter in the setting of a normal level of consciousness. Most cases of reported CNH are related to metabolic or hypoxic encephalopathy and with decreased level of consciousness.¹ There are several reports of CNH with normal level of consciousness in brainstem tumor such as diffuse infiltrating astrocytoma, glioma and lymphoma.² - ⁴ However only one case had been reported with normal level of consciousness in ischemic stroke. In that case bilateral medial thalamic small infarction was responsible for the CNH.⁵

The mechanism of CNH is not clear. In their initial report, Plum and Swanson⁶ hypothesized that a medial pontine structural lesion was responsible for CNH.

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**Figure 1.** MRI of the patient. Acute stage of infarction at right paramedian pontine region (A), DWI, (B), T2WI

**Figure 2.** High resolution chest CT scan showing normal finding
Subsequently, Krendal et al. described this in the setting of diffuse lymphoma with acidification of CSF. This finding raised the possibility that local lactate production by tumor cell could stimulate medullary chemoreceptor and cause hyperventilation.

The precise nature of the structural lesion necessary to cause CNH also remains unclear. All reported awake patients with CNH have diffuse infiltration of the pons by tumor, either glioma or lymphoma, with relative sparing of medullary respiratory center. The thalamus may play an important role in regulating ventilation. The mediodorsal nucleus of thalamus normally inhibits respiration and ventilation. The vagus and afferents of the phrenic nerve project signals to cortex through the thalamus. Plum and Swanson suggested that the medial pontine dysfunction results in unopposed stimulation of the medulla by interrupting inhibitory signal from the thalamus. Our case showed episodic central hyperventilation with concomitant facial nerve palsy and horizontal conjugate eyeball deviation which are the symptoms consistent with unilateral pontine lesion in brain MRI. And diffuse basilar artery stenosis may aggravate bilateral pontine tegmental ischemic change which interrupts whole inhibitory signal from thalamus transiently. These may explain the pathophysiology of our case similar to those of Plum and Swanson. The alertness depends on the integrity of reticular activating system (RAS). Pontine RAS is relatively sparse compared with the thalamic or midbrain RAS. For this reason, we hypothesize that our patient may keep alertness.

There are many causes of episodic or chronic hyperventilation. Psychogenic problem such as tardive dyskinesia, heart failure, hyperthyroidism, portal systemic encephalopathy, meningitis and SAH can elicit hyperventilation. Our patient had hyperventilation during the sleep which excludes psychogenic cause. Normal LFT and TFT, medical history and imaging study exclude other remained possible causes of hyperventilation. Normal PFT, perfusion lung scan and chest CT ruled out primary lung problem.

Figure 3. Perfusion lung scan showing normal finding.
Our case is the first domestic report of CNH in alert patient with unilateral pontine infarction compared to previously reported case with bilateral median thalamic infarction. Extensive work-up can differentiate many relevant causes of CNH.

The mechanism and nature of brain lesion which causes CNH are unclear. In the future, many other cases may emerge to clarify the pathophysiologic process.

REFERENCES