P-107

Central alveolar hypoventilation (Ondine's curse) in Medullary infarct : A case report

Jieun Kim, M.D., Soo Jeong Han, M.D., Ph.D.
Department of Rehabilitation Medicine, Ewha Womans University Mokdong Hospital, Seoul, Korea

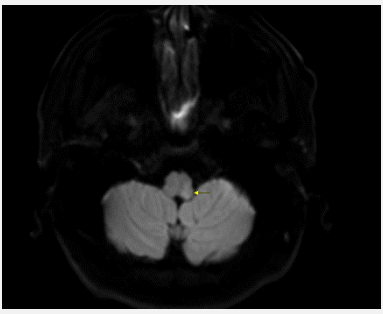
Introduction

- Central alveolar hypoventilation (CAH), also known as "Ondine's curse" is rare, presenting with respiratory failure.
- CAH is caused by a defect in the PHOX2B (paired-like homeobox 2B) gene. Occasionally, it occurs among adults, and common causes of CAH in adults include brainstem ischemia, mass, infection, demyelinating disease, or anoxic-ischemic damage.
- We present **a case of a 55-year-old man who revealed CAH as a sequela of stroke**.

Case

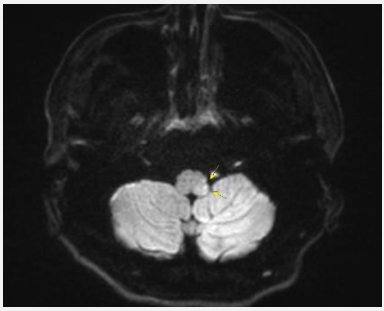
- A 55-year-old man without underlying medical conditions presented to the emergency room on July 4, 2023, complaining of headache and balance impairment. Brain magnetic resonance imaging showed **a tiny acute infarction in the left posterior medulla (Figure 1)**, so he admitted to the department of neurology.

Figure 1. On HD1 Brain MRI (MRI diffusion)



- On hospital day(HD) 20, he transferred to the department of rehabilitation medicine with nasal oxygen at a rate of 3L/min.
- On HD21, at 4:45am, **the patient became cyanotic and unconscious, requiring intubation with arterial blood gas showing partial pressure of carbon dioxide (pCO2) of 100.9 mmHg.** The patient was transferred to the Intensive Care Unit (ICU) and submitted to mechanical ventilation.
- His baseline pCO2 was around 50, and the remaining blood gas values were previously normal. A head CT with no contrast showed known ischemic infarction in the left medulla, with no evidence of intracranial bleeding or midline shift. The chest computed tomography(CT) scan was performed to determine the cause of the carbon dioxide(CO2) retention, **but no culpable lesion was identified.**
- **Brain magnetic resonance imaging was repeated and showed an extension of the previous infarction in left posterior medulla to high signal intensity in the posterolateral medulla(Figure 2).**

Figure 2. On HD21 Brain MRI (MRI diffusion)



- On HD28, the patient was extubated with the weaning parameters on a spontaneous breathing trial. On HD35, neurological examination was performed and there were no cognitive or functional sequelae compared to before CAH.
- He was able to breathe in room air and discharged.

Discussion

- Criteria for the diagnosis of CAH are not well established. However, CAH is the syndromic diagnosis, and that the criteria for diagnosing such syndrome should be flexible in some situations, and should be considered in patients who have deficits in respiratory neural pathways.
- **Recognition of CAH is important to improve the prognosis of patient in risk.**