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CASE REPORTS

Late-onset central hypoventilation caused by posterior inferior cerebellar arterial infarction

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Abstract

Central hypoventilation syndrome (CHS) is a rare condition caused by damage to the brainstem's respiratory centers, often following stroke. We report a case of a 65-year-old woman with a lateral medullary infarction presenting with dizziness, dysarthria, and dysphagia. Five days after admission, she experienced respiratory failure with apnea and CO2 retention, requiring mechanical ventilation. Timely intervention improved her respiratory condition, and she was discharged with home ventilation care for ongoing management. This case highlights the importance of recognizing CHS in brainstem stroke and the critical role of early ventilatory support in improving outcomes.

Keywords: Central hypoventilation syndrome, brainstem stroke, respiratory failure, mechanical ventilation

INTRODUCTION

Central hypoventilation syndrome (CHS), also known as acquired Ondine's curse, is a rare disorder that presents with respiratory failure during sleep or, if severe, during the awake state.[1] The term, first described in 1962, refers to the syndrome of sleep apnea with preserved voluntary control of respiration.[2] Hypoventilation occurs after damage to the medullary respiratory center. Rarely, brainstem stroke can produce CHS, characterized by the loss of automatic breathing during sleep and, in severe cases, leading to respiratory failure.[2,3] The respiratory neurons are located in the reticular formation of the medulla oblongata. However, central hypoventilation has also been reported following damage to the medulla from poliomyelitis, neoplasms, trauma, and other causes.[4] The anatomic proximity of the centers controlling breathing, sleep, and upper respiratory tract motility suggests a close physiological interrelationship. Hypoventilation occurs in the acute phase following the impairment of the medullary respiratory center.[3,4] However, delayed development of the syndrome after several days is unusual.

CASE REPORT

A 65-year-old woman with a history of hypertension, type 2 diabetes mellitus, and myocardial infarction presented to the emergency department (ED) with dizziness, dysarthria, and dysphagia. On arrival at the ED, the patient had a blood pressure of 130/70 mmHg, temperature of 36.1°C, respiratory rate of 25 breaths per minute, and pulse rate of 130 beats per minute. Neurological examination revealed spontaneous nystagmus to the right, left-sided facial droop, dysphagia, and dysarthria. Brain MRI revealed a left posterior inferior cerebellar infarction involving a lateral medullary lesion (Figure 1a, 1b). Chest radiographs revealed no acute abnormalities. Five days after admission, the patient's condition abruptly deteriorated, showing lethargy with apnea and, after agitation, chest discomfort and confusion. Shortly thereafter, the patient became cyanotic, requiring intubation. Arterial blood gas analysis showed pH 7.048, pCO2 84.1 mmHg, pO2 148, and HCO3 22.1 mmHg. CO_2 retention indicated possible central respiratory dysfunction despite the patient having no medical history of lung disease. Follow-up chest radiography and brain MRI revealed no interval changes.

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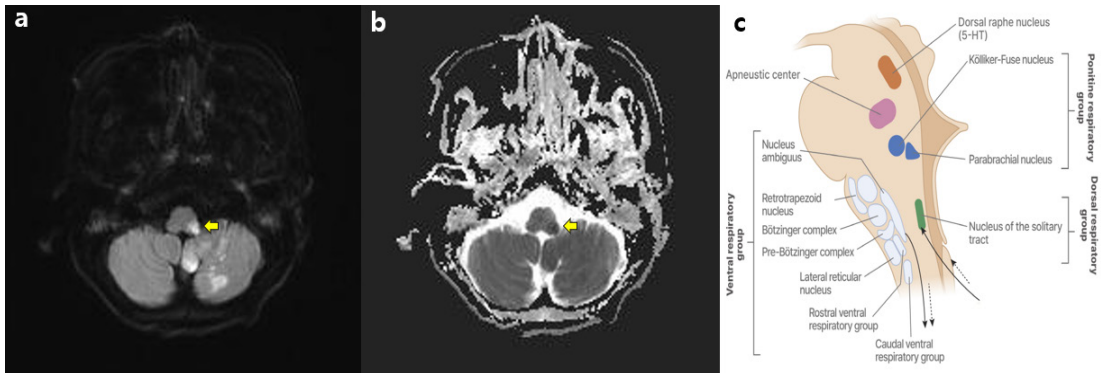

Figure 1. Axial diffusion-weighted magnetic resonance imaging (a, b) showing left posterior inferior cerebellar arterial territorial infarction (yellow arrow). Drawing of the brainstem (c), showing areas of the organization of respiratory center.

Mechanical ventilation was initiated, leading to gradual improvement in the patient's apnea. The patient was eventually stabilized and discharged with home ventilation care, allowing for continued management of her respiratory condition.

DISCUSSION

Breathing is controlled by voluntary and automatic mechanisms. The dorsal respiratory group, consisting of inspiratory neurons, is located ventrolateral to the solitary nuclei of the medulla. The ventral respiratory group, which consists of inspiratory, expiratory, and rhythm-generating neurons, comprises a long column of neurons extending from the cervical cord at C1 to just below the facial nuclei. The pontine respiratory group controls the switch between inspiration and expiration and is located dorsolateral to the upper pons. All three groups of neurons are bilaterally paired (Figure 1c). In the acute phase of brainstem stroke, they can result in abnormalities such as Cheyne-Stokes respiration, central hyperventilation, sleep apnea, apneustic breathing, and ataxic breathing.[5,6] Rarely, brainstem lesions can produce CHS, characterized by the loss of automatic respiration, which results in severe sleep apnea that can lead to respiratory failure. Bilateral damage is thought to be necessary to induce central hypoventilation because each side is believed to have the ability to independently drive diaphragmatic activity.[7] However, unilateral ischemic infarction of the lateral medullary tegmentum demonstrates that unilateral damage can cause CHS.[4,8] In our patient, a small dorsolateral medullary infarct resulted in delayed central hypoventilation syndrome and apnea, with gradual diminution of automatic and voluntary respiration during quiet rest, followed by apnea. The delayed development of the syndrome is unusual. The authors hypothesized that some form of secondary neuronal degeneration or an abnormality of local synaptic interconnections related to plasticity and remodeling contributed to the pathogenesis of the patient's disorder.

CHS is a rare disorder caused by various mechanisms and should be considered in patients with respiratory neural pathway deficits. Prompt recognition of CHS is critical for improving patient prognosis. Specifically, CHS may manifest later in smaller medullary lesions that affect a restricted portion of the lateral medulla. Administering appropriate treatment for assisted ventilation can have a significant impact on patient prognosis.

DISCLOSURE

Ethics: Written informed consent was obtained from the patient for the publication of this article.

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Conflicts of interest: None

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