Ondine's Curse Caused by Brainstem Glioma

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Clinical Image

A 60 year old male presented to the emergency room from his nursing home with hypoxia. He had previously received levofloxacin for empiric treatment of pneumonia with no improvement. The patient had a history of high grade glioma of the 4th ventricle for which he underwent suboccipital craniectomy and resection approximately 10 months prior. The patient was intubated soon after arrival and admitted to the intensive care unit for further management. On initial evaluation, he was noted to have prolonged episodes of apnea on pressure support ventilation. Initial venous blood gas showed pH 7.39, pCO₂ 89, PO₂ 33, and HCO₃ 56, indicative of a well-compensated respiratory acidosis. Magnetic resonance imaging of the brain with intravenous contrast showed interval expansion of a hyperintense lesion within the brainstem consistent with recurrence of his glioma (arrow) (Figure 1). This clinical picture was consistent with the central hypoventilation syndrome known as Ondine’s curse, in which disruption of the central respiratory center causes impaired control of autonomic respiration. The patient was ultimately unable to be weaned from the ventilator succumbed to his disease during this hospitalization.

Figure 1: MRI of the brain at the time of admission reveals a T2/FLAIR hyperintense lesion within the brainstem at the level of the central respiratory center.