Alveolar Hypoventilation Syndrome in Brainstem Glioma With Improvement After Surgical Resection*

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A 3-year-old boy presented with brainstem astrocytoma and central alveolar hypoventilation syndrome. Contrast MRI of the brain showed that the tumor involved the cerebellum, with compression of brainstem, and resolved after surgical resection. Polysomnography performed before and after total tumor resection showed significant improvement in nocturnal respiratory rate, respiratory disturbance index, and oxygen desaturation. It is apparent that central alveolar hypoventilation syndrome secondary to brainstem tumor may improve after surgical resection for those with favorable anatomic location and histology. Serial polysomnography and MRI scans are useful for diagnosis and in the management plan, and to monitor progress.

(Submitted: January 26, 1999; Accepted: December 30, 1999)

Key words: brainstem tumor; central alveolar hypoventilation syndrome; childhood; polysomnography

Abbreviations: CHS = central hypoventilation syndrome; RDI = respiratory disturbance index

Central hypoventilation syndrome (CHS) in children is an uncommon but serious condition.1 Central alveolar hypoventilation caused by brainstem tumor is even rarer. There are only few case reports, and all cases were fatal.2–4 Herein we describe a child with CHS caused by brainstem astrocytoma with subsequent improvement in ventilation status after tumor resection. The improvement in ventilation status was demonstrated on serial polysomnography.

Case Report

A 3-year-old boy presented with chronic cough, hoarseness of voice, and left vocal cord paralysis. Contrast MRI of the brain revealed a 4-cm cystic mass in the posterior fossa. The mass filled up most of the fourth ventricle, extending to the inferior vermis posteriorly and to the medulla anteriorly, with compression of the dorsum of the pons and pontomedullary junction (Fig 1, bottom). Posterior fossa craniectomy was attempted, but only a small part of the tumor was removed. Histology showed pilocytic astrocytoma. The child later developed bulbar palsy and left facial nerve palsy. A tracheostomy was performed.

For the next 2 months, the child became oxygen dependent. When the child was awake, arterial blood gas measurement showed elevated CO₂ level at a partial pressure of 48.6 mm Hg and normal alveolar arterial oxygen gradient. He was drowsy in the daytime and had recurrent apneic spells necessitating resuscitation. A second craniectomy was carried out 3 months later, and the bulk of residual tumor was removed. A left ventriculoperitoneal shunt was inserted for hydrocephalus. The child had also developed left-sided hemiparesis. A repeat arterial blood measurement, performed when the child was awake, showed persistent CO₂ retention at a partial pressure of 49.4 mm Hg. End-tidal CO₂ level during sleep was elevated up to 84 mm Hg. Overnight polysomnographic evaluations were performed, using an Oxford Medilog 9200 (Oxford Instruments; Oxon, England), before the second operation and at 6 and 14 months after the second operation. The airflow sensor was placed at the tracheostomy site in all the studies. Central apnea was defined as cessation of airflow with no respiratory effort for >10 s. The initial study had total sleep time of 447.5 min, with sleep efficiency of 93.4%. The child's respiratory rate was 12 breaths/min during relaxed wakefulness, and it further declined to 6 to 8 breaths/min during sleep (Fig 2, top). There were multiple apneic episodes of central origin, with a respiratory disturbance index (RDI) of 83.8/h. The longest apneic episode was 30 s. The lowest arterial oxygen saturation was 72%. There were 383 episodes of oxygen desaturation <4%, with 155 events of desaturation <90%.

The second study had total sleep time of 466 min, with sleep efficiency of 79.3%. Respiration was more regular, with respiratory rate of 20 breaths/min. There was marked reduction in RDI to 9.9/h, and the longest apneic episode was 38 s. The number of episodes of oxygen desaturation <4% was reduced to 49, with 46 events of desaturation <90%. The third study had total sleep time of 391 min, with sleep efficiency of 45%. There was continuing improvement in the regularity and amplitude of breathing during sleep (Fig 2, bottom). The RDI was 3.4/h. The longest apneic episode was 14 s. The lowest arterial oxygen saturation was 83%. The number of episodes of oxygen desaturation <4% was further decreased to eight, with two events of desaturation <90%.

Contrast MRI scan of the brain was repeated 22 months after surgery (Fig 1, bottom). There was no evidence of enhancing tumor in the posterior fossa, and compression was relieved. The child has survived up to 3 years since initial presentation and has been discharged home. He has residual bulbar and left facial palsy and left hemiparesis. He does not receive ventilation in the daytime, but receives home ventilation at night. We plan to reduce his ventilation setting further, although previous attempts in taking him off night ventilation have resulted in retained secretions, atelectasis, and pneumonia.

Discussion

Brainstem tumor resulting in CHS has been rarely reported. The prognosis of brainstem tumor in children is poor, with a 5-year survival rate of 30%. Most reported cases of brainstem tumor with CHS were invariably fatal.2–4 Our patient is unique, in that not only did he survive after tumor resection, but also there was improvement in his CHS. The tumor was a low-grade pilocytic astrocytoma, with an anatomic growth pattern similar to that of a dorsally exophytic and cervicomedullary tumor, which has a better prognosis.5 One of the presenting features in our case was bradynea, which improved after tumor resection. Irregular respiratory pattern in a patient with a brainstem tumor had been reported previously.2

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CHEST 2000; 118:266–268

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The detailed mechanism of CHS is largely unclear. For secondary causes such as brainstem tumor, the extent of involvement of various respiratory centers would be important in determining the outcome. Our studies of cases of brainstem infarct suggest that unilateral involvement of pontomedullary reticular formation and nucleus ambiguus can result in loss of automatic respiration, and that associated lesion of the nucleus tractus solitarius may lead to more severe respiratory failure, involving both automatic and voluntary responses.6 In our patient, the tumor compressed the dorsum of upper medulla and the distal pons up to its mid-upper portion. The reticular formation and the pneumotaxic center are usually located just ventral to the floor of the fourth ventricle and may have been affected. Surgical resection of the tumor resulted in relief of compression.

Although there was significant improvement in his central alveolar hypoventilation, as shown on polysomnography, we were unable to wean this child off night ventilation. This could be a result of respiratory muscle weakness and abnormal ciliary function due to his neurologic complications. Another possible explanation is that although there was significant improvement in his central hypoventilation, the recovery was not complete.

In conclusion, we presented a unique case of brainstem pilocytic astrocytoma with improvement in CHS following surgical debulking. However, the long-term prognosis of this patient awaits further follow-up. A more invasive method for long-term ventilation, with the use of diaphragmatic pacing, is not without complications and should be reserved for future consideration.

References

Figure 2. Top: Polysomnographic tracing of rapid eye movement sleep of the patient before tumor removal, showing the slow respiratory rate and central apneas. Airflow was detected over the tracheostomy site. Bottom: Polysomnographic tracing of rapid eye movement sleep of the patient 14 months after tumor removal, showing more regular breathing at 13 breaths/min. Airflow was detected over the tracheostomy site. EOG = electro-oculogram; ROC = right outer canthus; LOC = left outer canthus; EMG = electromyogram.