Ondine's Curse

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Ondine was cursed and had to remember to breath. Acquired central hypoventilation is rare but potentially treatable.

A 60 year old woman was admitted to intensive care with increasing drowsiness and hypoventilation. She had initially noticed a dry mouth and slurred speech 7 weeks earlier, and subsequently developed postural hypotension followed by progressing incoordination and left sided deafness. On examination she was ventilated, and now fully aware, with upbeat nystagmus, a left 6th nerve palsy, with loss of lower limb reflexes and left sided ataxia. Extensive blood tests and MRI brain were normal. CSF was acellular with normal protein with positive oligoclonal bands. Nerve conduction studies found a mild sensorimotor axonal neuropathy. CT of chest, abdomen and pelvis were normal. A diagnosis was made and subsequently confirmed.

This subacute onset pontine-medullary syndrome is unusual but is characteristically associated with anti-Hu antibody. A sub-carinal node identified on PET scan was found to be small cell lung cancer.
She was treated with high dose steroids and her neurological syndrome resolved. She is undergoing chemotherapy.

Historical reports with late or post-mortem diagnosis indicate a poor prognosis in this syndrome. Greater awareness of the syndrome with earlier recognition and treatment may allow better outcomes.