CORRESPONDENCE

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Fatal Hydrocephalus in a Patient with Neurofibromatosis

To the Editor:—Anesthetic implications of neurofibromatosis may include airway, mediastinal and vertebral neurofibromas, associated endocrine syndromes, and intracranial masses in 5–10% of patients.[1] Hydrocephalus is a rare manifestation of neurofibromatosis and is commonly described during the first two decades of life.[2–4] We present a case of acute postoperative hydrocephalus in a previously healthy 21-yr-old patient with neurofibromatosis.

The patient presented for excision with bilateral flap closure of a 12-cm cutaneous neurofibroma of the posterior neck and back. He had another neurofibroma on a toe, cafe-au-lait spots, and no history of seizures or intracranial disease and was otherwise in good health. The 4-h anesthesia and surgery was uneventful, with careful prone positioning, and the patient was discharged, awake, from the recovery room. Over the next 6 h, the patient received hydrocodone (two tablets), promethazine (12.5 mg), morphine (4 mg), and promethazine (12.5 mg). One hour after the morphine-promethazine (12 h after surgery), the patient's mother reported that the patient had a headache, had vomited, and may have had a seizure. At assessment, the patient was found to be unresponsive and was immediately intubated and resuscitated by an anesthesiologist who was on the floor at the time. Pupils were fixed and dilated, computed tomography showed severe obstructive hydrocephalus with transtentorial herniation, severe diffuse cerebral edema, and an ill-defined area of decreased density in the right thalamus. Although a ventriculostomy was placed, the patient remained unresponsive and was declared brain dead the following day.

The computed tomography findings were compatible with a diagnosis of aqueduct stenosis, and the cause of the acute increase in intracranial pressure was speculated to have resulted from a seizure. In retrospect, the presumed nausea, necessitating perphenazine 1 h before arrest, may have heralded an increase in intracranial pressure.

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We present this case to alert anesthesiologists to the rare and potentially life-threatening association of obstructive hydrocephalus in patients with neurofibromatosis. The absence of symptoms does not preclude the diagnosis with late presentation described, and preoperative assessment should include a search for previous cranial imaging. If not available, as in this case, imaging should be considered before elective surgery in light of recommendations for routine imaging in these patients.[4]

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