We appreciate Dr Weisberg's concern that delayed enhancement at the operative site may simply represent recurrent tumor. We therefore provide clinical and radiographic follow-up data on patients in whom postoperative enhancement appeared along the line of resection (Table). Late CT scan observations when considered together with the clinical course suggest the following: (1) "benign" contrast enhancement can occur after brain tumor surgery; its appearance is delayed and it disappears spontaneously; and (2) when enhancement at the operative site persists for longer than 3 to 4 months, there is increasing concern that recurrent tumor is a contributing factor.

Frequent postoperative CT scans were the "methods of procedure" by which we analyzed patterns of contrast enhancement in the postoperative brain. Dr Weisberg has confused our method with our conclusion. We recommend a single postoperative scan, but we also recommend that this scan be performed on the third or fourth postoperative day. Postoperative enhancement in brain tumor patients is more than a radiological curiosity. It can mimic residual enhancing tumor leading to inappropriate treatment decisions and misinterpretations of response to treatment [2].

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References

1. Cairncross JG, Pexman JHW, Rathbone MP, DelMaestro RF: Postoperative contrast enhancement in patients with brain tumor. Ann Neurol 17:570–572, 1985
2. Cairncross JG, Pexman JHW, Rathbone MP: Post-surgical contrast enhancement mimicking residual brain tumour. Can J Neurol Sci 12:75, 1985
3. Weisberg L: Computed tomographic findings of intracranial gliosis. Neuroradiology 21:253–257, 1981

Episodic Tourette's Syndrome in a Patient with Citrullinemia

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We describe the case of a 13-year-old girl with citrullinemia who in the course of episodic metabolic imbalances developed symptoms of Tourette's syndrome. Citrullinemia was diagnosed at ten weeks of age and she has been the subject of previous communications in the literature [1]. Treatment consisted of protein restriction and supplemental essential amino acids. She did well on this regimen, although intercurrent infections triggered rare 1- to 2-day episodes of ataxia, irritability, and lethargy associated with hyperammonemia. At 11 years of age she developed episodes of additional symptoms of repetitive tics accompanied by involuntary vocalizations.

She was referred to the Mayo Clinic for an attack following a bout of gastroenteritis. Her parents described echolalia, but by the time of her evaluation she showed only lethargy, combativeness, and ataxia. Routine laboratory findings were normal. Abnormal laboratory values consisted of: ammonia, 165 μg/dl (normal, less than 50); plasma glutamine, 207 μm/dl (normal, 34 to 67); and citrulline, 203 μm/dl (normal 1.6 to 5.5). The patient was treated with intravenous fluids, protein restriction, supplemental arginine, and sodium benzoate, followed by rapid improvement. On the next day examination findings were normal. An electroencephalogram showed mild generalized slowing. Computed tomographic scan showed only a prominent sulcus in the left parietal cortex.

The impression was that she had an episodic form of Tourette's syndrome precipitated by hyperammonemia and elevated concentrations of certain amino acids. It was hoped that the patient would return during future attacks to allow further assessment, including video monitoring, but the parents elected to obtain medical care closer to home.

While we observed this child on only one occasion and are basing our impression of Tourette's syndrome on history alone, we believe it worthwhile to note this possible association. Citrullinemia produced by arginosuccinate synthetase deficiency is a rare disorder of the urea cycle, and elevated serum ammonia values is the hallmark. Cases are known [2] in which citrullinemia was associated with normal ammonia levels; therefore, ill effects on the nervous system may not always be, or all be, due to the hyperammonemia. The brain alters ammonia primarily by forming glutamine, with glutamate—a known excitatory neurotransmitter—as an intermediate metabolite [4]. It is of interest that Walser [3] cites cases from the Japanese literature in which patients with the late-onset type of citrullinemia show behavioral changes, with manic episodes, frank psychosis, and echolalia.

We speculate that some transient disturbance of neurotransmission was responsible for the episodic Tourette's syndrome described in this patient.

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References

1. Buist NRM, Kennaway NG, Hepburn CA, et al: Citrullinemia: investigation and treatment over a four-year period. J Pediatr 85:208–214, 1974
2. Scott-Emuakpor A, Higgins JV, Kohrman AF: Citrullinemia—a new case with implications concerning adaptation to defective urea synthesis. Pediatr Res 6:623–633, 1972
3. Walser M: Urea cycle disorders and other hereditary hyperammonemic syndromes. In Stanbury JB, Wyngaarden JB, Fredrickson DS, et al (eds): The Metabolic Basis of Inherited Disease. New York, McGraw-Hill, 1983, pp 402–438
4. Weil-Malherbe H: Ammonia metabolism in the brain. In Elliott KAC, Page IH, Quastel JH (eds): Neurochemistry. Springfield, IL, Thomas, 1962, p 321