Endoscopic disconnection of hypothalamic astrocytoma causing gelastic epilepsy

Case report

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The authors report on a case of juvenile pilocytic astrocytoma (JPA) and concomitant hypothalamic hamartoma (HH) with gelastic epilepsy that was successfully treated with endoscopic disconnection. This 6-year-old girl presented with prolonged, medically intractable gelastic seizures that were often followed by generalized tonic seizures. An enhancing, low-grade hypothalamic tumor was identified on MR images obtained when she was 11 months old, but no surgical intervention was attempted at that time apart from bur hole drainage of a chronic subdural hemorrhage. In the first surgery, performed when she was 6 years of age, the authors attempted disconnection and tumor sampling; the lesion was revealed to be a JPA. A second endoscopic disconnection was performed 1 year later to improve seizure control and obtain a pathological specimen from the nonenhancing contralateral side. The pathological results after the second surgery revealed that the enhancing mass was a spontaneously regressing JPA and the contralateral nonenhancing mass was an HH. The HH was found as latent tumor and the JPA was the mass causing gelastic epilepsy. To the authors’ knowledge, this is the first report of a patient with a spontaneously regressing JPA and concomitant HH, both of which were treated by endoscopic disconnection. (DOI: 10.3171/2009.3.PEDS08129)

Key Words • gelastic epilepsy • pilocytic astrocytoma • hypothalamic hamartoma • neuroendoscopy • image guided surgery

Patients with HH and gelastic seizures are regarded as having “happy baby syndrome” because of the characteristics of this seizure type. The seizures are usually accompanied by HH-derived mental retardation and precocious puberty. Other pathological findings related to gelastic seizure have rarely been reported.3,6,8,18 One case of a hypothalamic astrocytoma with gelastic seizures has been reported previously, and in that case, a transcortical approach to surgery was used.6 Juvenile pilocytic astrocytomas have been known to regress spontaneously without malignant changes, which explains why these lesions are described as having hamartoma-like qualities.12,18 However, concomitant HH and JPA with gelastic epilepsy have not been previously reported.

Hypothalamic hamartomas with gelastic seizures are considered ideal lesions for endoscopic disconnection,15 but this procedure has not been attempted in a patient with a concomitant lesion. When an HH is present along with another pathological entity, gelastic epilepsy should be investigated to discover its true cause and identify the most effective treatment.

To our knowledge, a spontaneously regressing JPA with a concomitant HH in a patient without NF1 has not been reported previously. In this report, we describe our experience with such a patient suffering from gelastic epilepsy who underwent endoscopic disconnection.

Case Report

History and Presentation. This 6-year-old girl presented with long-lasting, medically intractable gelastic seizures followed by 2–3 episodes of generalized tonic seizures in a day. A low-grade hypothalamic tumor was diagnosed when the patient was 11 months of age, but

This article contains some figures that are displayed in color online but in black and white in the print edition.
no surgical intervention was attempted apart from bilateral bur hole drainage of chronic subdural hemorrhage (Fig. 1A). The enhancing tumor decreased spontaneously without resection, but the gelastic seizures worsened over time. Magnetic resonance images revealed the presence of a hypothalamic tumor with an area of solid enhancement (Fig. 1B).

The patient did not show the classic symptoms of precocious puberty, but her height (125 cm), weight (44.0 kg), and head circumference (57 cm) were in the 97th percentile for her age group.

First Operation and Postoperative Course. We obtained multiple biopsy specimens from both the enhancing and the nonenhancing areas in the hypothalamic tumor stereotactically. Biopsies from both areas revealed the lesion to be a pilocytic astrocytoma. We decided that the enhancing mass might regress spontaneously, so we focused on treatment of the intractable seizures rather than on tumor removal. We performed endoscopic disconnection of the tumor from the hypothalamus to control seizures in the first disconnecting surgery. Histological tests showed unexpected pathological results, namely, that the enhancing tumor and the nonenhancing mass on the right side of the hypothalamus were pilocytic astrocytomas characterized by a moderate cell density with hyperchromatic elongated nuclei, such as that found in the biopsy.

Considering the rate at which the lesion was regressing, we thought that it might regress spontaneously. We decided to disconnect the mass, which was causing the seizures, from the hypothalamus rather than remove the tumor.

After biopsy sampling, we inserted a depth electrode into the tumor mass using a stereotactic frame. Four contacts, each 2-mm-long and separated by 10 mm, were inserted using a Leksell frame. Three contacts were inserted into the hypothalamic mass, and the remaining contact was placed in the adjacent tissue (Fig. 2). After depth electrode insertion, the patient underwent video electroencephalography monitoring for 5 days. We found epileptic discharges, not only within the tumor mass, but also propagating into adjacent tissues. We performed endoscopic disconnection of the hypothalamic tumor to control the seizures. The Stryker navigation system (Stryker Instruments) was used for image guidance. Image-guided surgery uses real-time imaging to guide endoscopic placement and facilitate navigation (Fig. 3).

A single bur hole was made 1 cm anterior to the coronal suture and 2 or 3 cm lateral to the midline. After lateral ventricle tapping, a 14 Fr peel-away catheter (Cook) was inserted into this tract. The sheath of a peel-away catheter is useful in protecting the cortex around the endoscopic tract during the procedure. After advancing the endoscope through the foramen of Monro and across the lateral ventricle, we could see the protruding mass from the third ventricle. A surgical margin was created between the thalamus and the tumor mass. We then placed the depth electrodes under navigation guidance. Disconnection was performed at the pia mater of the mass adjacent to the interpeduncular cistern at the shortest distance without causing injury to the pia, thalamus, or brainstem. We used a monopolar electric coagulator and microforceps to disconnect the mass from the hypothalamus; radiofrequency ablation and lasers were not used in this case.

Pre- and 1-month postoperative laboratory test findings related to adenohypophysial hormones were as follows (preoperative finding/postoperative finding): adrenocorticotropic hormone 17.88/32.95 pg/ml (normal 7–56 pg/ml), thyroid-stimulating hormone 8.87/1.85 µIU/ml (normal 0.3–4.0 µIU/ml), growth hormone 0.07/0.01 ng/ml (normal 0.28–8.70 ng/ml), luteinizing hormone 4.55/3.50 µIU/ml (normal 1.1–8.8 µIU/ml), follicle-stimulating hormone 5.27/4.87 µIU/ml (normal 1.8–13.6 µIU/ml), prolactin 18.32/12.74 ng/ml (normal 1.40–14.6 ng/ml), T3 85.85/160.56 ng/dl (normal 80–200 ng/dl), free T4 0.77/0.54 ng/dl (normal 1.0–1.8 ng/dl), cortisol 240.06/16.75 µg/dl (normal 3.80–18.4 µg/dl). The responses of adenohypophysial hormones to the triple stimulation test were normal pre- and postoperatively (thyrotropin-releasing hormone 0.5 mg, luteinizing hormone–releasing hormone 0.1 mg, and growth hormone–releasing factor 0.1 mg).

There was no postoperative increase in sodium levels, nor did diabetes insipidus arise. Mental retardation (IQ 69) was determined preoperatively. The patient showed marked improvements in behavior, school performance, and quality of life at 1 year postoperatively.
Second Operation. For better seizure control and so that a pathological specimen could be obtained from the contralateral side, a second endoscopic disconnection was performed in the same manner as the first, 1 year later. The first disconnection and tumor specimen were on the left side, and the second disconnection and tumor specimen were on the right (Fig. 1C and D). The sample obtained from the contralateral nonenhancing mass was confirmed to be an HH (Fig. 4).

Postoperatively, the patient remained free of disabling seizures, and the frequency of gelastic seizures decreased by about 80%. Magnetic resonance images obtained 1 month after the second endoscopic disconnection demonstrated disconnection between the tumor and the bilateral hypothalamus, and no further growth of the enhancing mass (Fig. 1D).

Discussion

Gelastic seizures are typically provoked by HHs, although other causes have been reported rarely.8,14 Epileptogenic zones were reported in the frontal, temporal, and parietal lobes as well as the hypothalamus.8,9,18 Although the pathophysiological mechanisms of gelastic seizures are still controversial, stimulation of various anatomical regions including the hypothalamus, anterior cingulate gyrus, orbitofrontal cortex, basolateral temporal cortex, and the supplementary motor area may elicit laughter. The balance of the current literature seems to indicate that the anterior cingulate region is involved in the motor aspects of laughter, while the basal temporal cortex is involved in the processing of mirth.8,20 Theoretically, irritation or an epileptogenic mass in these neuronal pathways could cause gelastic seizures.

There are various pathological causes of gelastic seizures, including HHs, cortical dysplasia, tuberous sclerosis, and tumors.2,6,8,19 Hypothalamic hamartomas are the most common pathological entity associated with gelastic epilepsy. These lesions are nonneoplastic masses of ectopically located tissue resembling gray matter and containing neurons and glia arranged with no discernible pattern or organization.19 Another cause of gelastic seizures is a hypothalamic astrocytoma, previously reported by Coppola and colleagues;6 however, these authors used a transcal-
Conclusions

In our case, typical HH and JPA lesions arose at the same time in a patient, one in a spontaneously regressing tumor, the other in an aggravating symptomatic mass. Spontaneously regressing hypothalamic astrocytomas can be endoscopically disconnected from the hypothalamus. Like HHSs, in spontaneously regressing JPAs with concomitant HH and gelastic epilepsy, treatment should be targeted at seizure rather than tumor control. In cases of stable or spontaneously regressing tumor, endoscopic disconnection can lead to freedom from seizures. To our knowledge, this is the first case of a spontaneously regressing JPA and concomitant HH treated by endoscopic disconnection.

Disclaimer

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