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Case report

Early surgery of hamartoma of the floor of the fourth ventricle: A case report

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Abstract

Epilepsy from hamartoma of the floor of fourth ventricle (HFFV) is very rare, starting in neonate or infantile period, good response to surgery. We report a 3-month-old boy with episodes of medically intractable abnormal eyelid blinking and hemifacial seizure. The episodes began from the first day of life and were unresponsive to medication. A magnetic resonance imaging scan revealed mass lesion on the floor of fourth ventricle, with extended cerebellar peduncle and cerebellar hemisphere. Surgery had been performed two times previously for treatment; only a subtotal resection was performed due to severe bradycardia during the first operation. The patient underwent second operation for gross total removal of tumor. Complete resection of the mass after second surgery resulted in remission of seizures and histopathology revealed hamartoma with hemangiomatous vessel proliferation. Identification of characteristic semiology and associated HFFV can help prediction of intractability even in infant and favoring surgical treatment in early age. To our knowledge, this is the youngest patient who underwent early surgery just after progressing into status for HFFV, showing complete lesion resection resulting in successful seizure outcome.

Keywords: Hamartoma; Status epilepticus; Hemifacial spasm; Fourth ventricle; Cerebellar neoplasm

1. Introduction

Seizures originating from the brainstem, lesions surroundings to fourth ventricle or cerebellum are extremely rare entities. However, there are some cases in the literature reporting seizures associated with lesions in this anatomic region [1–3]. Partial seizures with eye blinking and hemifacial spasm are provoked by HFFV, are generally resistant to antiepileptic medication, may evolve into status epilepticus, and respond favorably to surgical therapy [1,2]. Recently, nearly 15 cases that were intractable to antiepileptic drugs were found to be associated with the fourth ventricle, brainstem, and cerebellum, and were subsequently reported as "cerebellar epilepsy" [1,2,4–6]. Intralesional EEG recordings of epileptic discharges and hyperperfusion on ictal SPECT (single-photon emission computed tomography) have suggested that seizure attacks may originate from the cerebellar parenchyma [1,2]. Early epilepsy surgery is recommended in children with medically refractory epilepsy for neurobiological, psychosocial, and learning perspective [7–9].

Here, we report the youngest infant who underwent epilepsy surgery of HFFV just after progressing into status, with successful seizure outcome after complete tumor resection.

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2. A case report

A 3-month-old boy was born at term by normal vaginal delivery after an uneventful pregnancy, the first child of unrelated parents. Since the first day of life, he had difficulty feeding, left hemifacial twitching, and eye blinking, with a left-sided deviation of the eyeball and lip for 20 s at 5–10 min intervals. There was no fever, cyanosis, or family history of seizure disorders. Phenobarbital and Sabril (gamma aminobutyric acid transaminase inhibitors), and Sentil (a benzodiazepine) were prescribed in a previous hospital, but these drugs did not decrease seizure activity. Eye blinking, hemifacial spasm, and irregular respiration during sleeping turned into status at 3 month of life. On presurgical evaluation, the neurological examination was normal between the attacks. Magnetic resonance imaging (MRI) scans revealed mass lesion on the left cerebellar hemisphere adjacent to the middle cerebellar peduncle, slightly hyperintense as compared to that of the cerebral cortex, with a bulging contour without definite enhancement and posterior displacement of the vermis (Fig. 1A). As a differential diagnosis, Lhemitte–Duclos disease and a low-grade astrocytoma were considered. A brain SPECT between the ictal and inter-ictal phases showed hyperperfusion around the cerebellar hamartoma and brainstem during seizure attacks (Fig. 2A). Video EEG showed non-localizing and non-lateralizing findings. A positron emission tomography (PET) scan showed hot fluorodeoxyglucose (FDG) uptake within the mass, and single-photon emission computed tomography co-registered to MRI (SISCOM) showed hyperemia within the mass and adjacent brainstem during seizures (Fig. 2A and B). Surgical intervention was warranted due to the increasing incidence of medically intractable seizures associated with autonomic symptoms (cyanosis, bradycardia). We did a midline suboccipital craniectomy and carefully advanced to the tumoral lesion under microscopy. The adhesive dorsal exophytic portion of the tumor was in the fourth ventricle, and another portion was attached to the brainstem. After exposure of the mass near the fourth ventricle, we put one depth electrode into the center of the mass and the other into floor of fourth ventricle to record the intralesional and adjacent electrical activity (Fig. 3). The mass generating electrical activities might be related with seizures. Because the cerebellar tumor was connected to the brainstem and the mass could not be distinguished from the normal tissue, we were not able to remove the tumor totally because of aggravated bradycardia during the lesionectomy showing electrical discharges (Fig. 3). The patient experienced worsening frequency of attacks after the first operation, uncontrolled by antiepileptic medication. A post-operative brain MRI demonstrated a subtotal removal of the cerebellar mass (Fig. 1B). A second operation was subsequently planned. We used intraoperative brain MRI for a more accurate and precise resection of the lesion attached to brainstem. Postoperative follow-up MRI revealed gross total removal of cerebellar peduncles and the floor of fourth ventricle portion of the tumor (Fig. 1C). Histopathology revealed hamartoma with vessel proliferation. No episodes had occurred since second surgery and he does not take any antiepileptic medication.

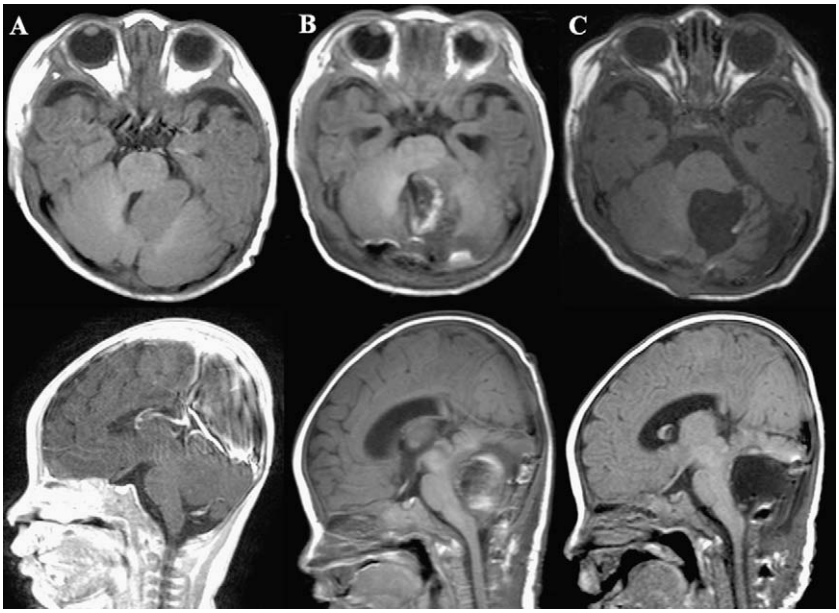

Fig. 1. Brain MRI. (A) Preoperative T1-weighted image was taken in axial and sagittal plan, (B) after the first operation, a heterotopic gray matter mass remained in left cerebellar hemisphere and middle cerebellar peduncle and (C) complete resection achieved after second operation.