Translational Research in Anatomy 5 (2016) 1–4

Contents lists available at ScienceDirect

Translational Research in Anatomy

journal homepage: http://www.journals.elsevier.com/translational-research-in-anatomy

Successful surgical treatment of intractable hemifacial spasm: A case report and review of cerebellar hamartomas of the floor of the fourth ventricle

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ARTICLE INFO

Article history:
Received 3 August 2016
Received in revised form
31 August 2016
Accepted 6 September 2016
Available online 8 September 2016

Keywords:
Cerebellar
Seizure
Epilepsy
Hamartoma

ABSTRACT

Introduction: Hamartomas involving the floor of the fourth ventricle and cerebellum are rare, but can be associated with medically recalcitrant hemifacial spasm. These lesions present early in the neonatal or infantile period and respond well to surgical excision.
Case Report: A 3-month-old white male presented with recurrent left hemifacial spasm, left eye deviation, and absent movement of the extremities. The patient was found to have a left eccentric lesion in the floor of the fourth ventricle and cerebellum. The patient showed no improvement with medical therapy by 6 months of age. He was taken to the operating room for suboccipital craniotomy and removal of the posterior arch of C1 followed by intralesional recording of epileptogenic activity and gross total resection of the lesion. After histologic analysis, the lesion was determined to be ectopic cerebral tissue consistent with a hamartoma. Postoperative MRI showed complete removal of the lesion, and the patient exhibited complete remission of his hemifacial spasm and associated symptoms.
Conclusions: Hamartomas involving the floor of the fourth ventricle can present with hemifacial spasm and respond well to surgical excision.

Contents

1. Introduction

Hamartomas are benign lesions composed of irregularly arranged mature tissue normally occurring in the respective location of the mass. These lesions may present with partial seizures and

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http://dx.doi.org/10.1016/j.tria.2016.09.001

hemifacial spasms that are poorly responsive to anti-epileptic drugs (AEDs). In addition to hamartomas, other lesions in the floor of the fourth ventricle, cerebellum, and brainstem have been associated with AED resistant seizures. The symptomatology of these lesions has even been described as “cerebellar epilepsy” [1–6]. The epileptic potential of these lesions has been described through intralesional electroencephalographic (EEG) recordings that demonstrated epileptic electrical activity, ictal single-photon emission computed tomography (SPECT) that demonstrated hyperperfusion of the lesion during the ictal onset, as well as gross total resection that resulted in cessation of the seizures [5–7]. Hamartomas of the floor of the fourth ventricle (HFFV) are remarkably rare but when symptomatic have a unique clinical presentation of recurrent partial seizures with hemifacial spasm [8–11]. A number of other symptoms may be present, which include eye blinking, eye deviation, and irregular breathing. As previously reported, these HFFV are generally resistant to AEDs but respond well to surgical therapy. Typically, surgical intervention for HFFV involves suboccipital craniotomy with gross total resection of the lesion [5,6]. Herein, we report a case involving a 6-month-old white male who underwent successful surgical resection of a HFFV.

2. Case Report

2.1. Initial hospital course

A 3-month-old white male who had multiple daily episodes of left hemifacial spasm involving the upper and lower face since birth was initially admitted to the neurology service. During each seizure episode, which would last approximately 10 s, the patient showed no movement in his upper or lower extremities while his eyes deviated to the left. Interictal neurologic examination demonstrated no abnormalities. Magnetic resonance imaging (MRI) performed shortly after birth at a previous hospital showed one non-enhancing left cerebellar mass that involved the floor of the fourth ventricle but spared the middle cerebellar peduncle and brainstem. No hydrocephalus was demonstrated on MRI. Subsequent computed tomography (CT) scan during his initial hospitalization revealed no changes. The patient's past medical and birth histories were remarkable only for hypospadias. The differential diagnosis for the lesion included ganglioglioma, ganglioneuroma, dysembryoplastic neuroepithelial tumor, hamartoma, or potentially Lhermitte-Duclos disease. The decision was made to continue with the AED Levetiracetam until 6 months of age and obtain an MRI of the lesion again at that time.

2.2. Operation

At 6 months of age, the patient showed no improvement with medical therapy and his parents reported approximately 200 seizure episodes per day. MRI again showed the non-enhancing left cerebellar mass with no significant interval change (Fig. 1, Fig. 2). A positron emission tomography (PET) scan was performed and demonstrated increased fluorodeoxyglucose uptake within the mass. At that time, surgical intervention provided the best possible solution for treating his intractable seizures. The patient was taken to the operating room and ventriculostomy was placed at Kocher's point on the right side. The patient was then placed in the prone position and an incision was made from the inion down to C2 and removal of the posterior arch of C1 performed. A suboccipital craniotomy was performed and the dura opened. Using a surgical microscope, the cerebellar hemispheres were retracted to expose the subependymal lesion that was present on the medial aspect of the left cerebellar hemisphere. A 4-probe intralesional electrode was placed and showed epileptogenic activity. Biopsies obtained

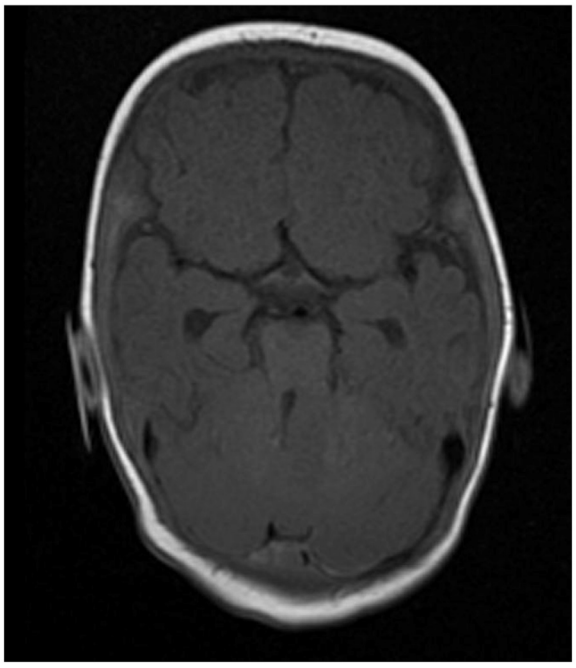

Fig. 1. Preoperative axial T1 MRI of the left-centered HFFV involving the cerebellum.

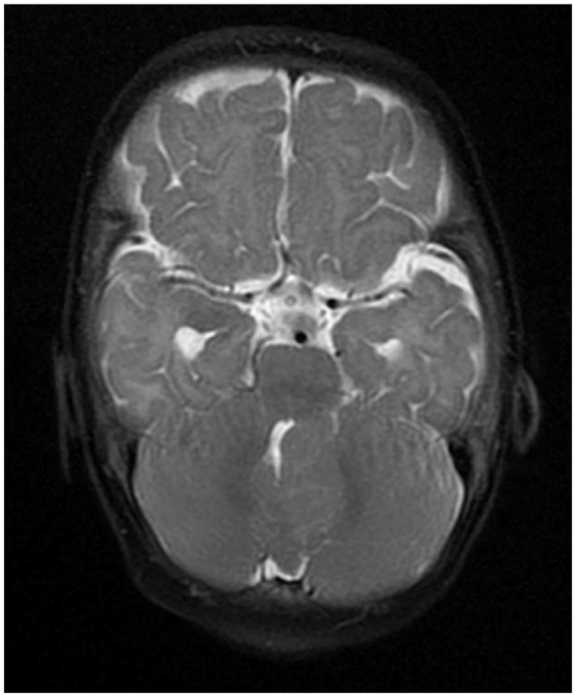

Fig. 2. Preoperative axial T2 MRI of the left-centered HFFV involving the cerebellum.

exhibited mature pyramidal neurons, with occasional dysplastic neurons showing abnormal processes and clumped Nissl substance, surrounded by reactive astrocytes with pink cytoplasm and spider-like processes (Fig. 3). Staining for glial fibrillary acidic protein, Neu-N, and neurofilament protein were positive. These findings were consistent with a hamartoma. To ensure complete resection of the lesion, measurements were made that corresponded with the dimensions of the lesion itself. In addition, an electrode was placed just within the middle cerebral peduncle