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Review article

A fourth ventricular ganglioneurocytoma representing with cerebellar epilepsy: A case report and review of the literature

Adnan Dagcinar[a,*], *Ahmet Hilmi Kaya*[a], *Haydar Ali Taşdemir*[a], *Enis Kuruoglu*[a], *Zafer Sabancilar*[a], *Aydin Sav*[b]

[a]*Medical Faculty, Department of Neurosurgery, Ondokuz Mayis University, Kurupelit, Samsun, Turkey*
[b]*Faculty of Medicine in Istanbul, Marmara University, Turkey*

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ABSTRACT

Fourth ventricular low-grade tumoral or dysplastic neuronal lesions have been reported as an epileptic focus for recently described cerebellar epilepsy in the form of repetitive and stereotyped attacks of hemifascial spasm, eye blinking, fascial movements, head deviation and dysautonomic manifestations. The case of a 3-month old infant having fourth ventricular mass with similar symptoms such as paroxysmal facial movements, eye blinking, eyelid contractions and abnormal head posture is reported in this article. After a few days of her admission, her attacks displayed a new form with altered consciousness and left limb jerks which were unresponsive to medical therapy. Following the surgical excision of the lesion 10 months ago, attacks disappeared and she is still seizure free. Histopathological diagnosis was *ganglioneurocytoma*. The seizures (which may be intractable in cerebellar epilepsy) are thought to have arisen from subcortical structures such as cerebellum, brain stem nuclei or the lesion itself. In the case of intractable episodes, surgical excision may prevent further seizures and help patients have a normal cognitive and motor development.

Contents

*Corresponding author. Tel.: +90 533 2314558; fax: +90 362 4576029.
E-mail address: adnandagcinar@yahoo.com (A. Dagcinar).
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1. Introduction

Seizures, resistant to medications associated with fourth ventricular hamartomas or low-grade tumors, have been reported in infants named as cerebellar epilepsy.[1–6] Formerly, these attacks were accepted as hemifacial spasm,[7–11] but in 1996, Harvey et al.[1] introduced the term of cerebellar epilepsy in a case of fourth ventricular hamartoma with the help of intraoperative recording. They postulated that the surrounding cerebellar tissue was causing the attacks and surgical excision would prevent episodes. By then, some identical cases were reported which tried to explain the nature of such seizures.[2–6] In this article, a 14th case of fourth ventricular mass lesion with intractable seizures where the patient became seizure free after surgical excision will be reported.

2. Case

The patient was a female and born as the first child of unrelated parents after a normal pregnancy. Since the first day of her life, her parents had noticed brief attacks of tonic contractions of the right eyelid, blinking of both the eyes and abnormal head postures. Attacks were occurring spontaneously, 30–40 times a day, recurring in clusters and lasting for a few minutes. Some clusters of attacks were also said to have been observed during sleep. Carbamezapine with valproic acid was prescribed in another center, but did not change the condition, and when the child was 3 months old, her parents decided to take medical care at our institution. Neurological examination was normal between the attacks. Several scalp video-electroencephalographic (EEG) recordings did not show any specific findings. Phenobarbital was added to previous medical therapy concluding no clinical improvement. Computerized brain tomography disclosed fourth ventricular mass that was isodense with the cerebellar tissue. Magnetic resonance imaging (MRI) scans revealed a mass arising from the left middle and superior cerebellar peduncles without accompanying hydrocephalus. The mass showed an isointense signal with cerebellar cortex on T1- and T2-weighted sequences and had no gadolinium enhancement (Fig. 1a and 1b).

After 4 days of admission, attacks gained a continuous pattern and were unresponsive to any medical application including infusion of midazolam. Tonic deviation of head and both eyes to the left side, blinking of the left eye, clonic contraction of the left arm and altered level of consciousness continued without any interruption. In this situation, surgical exploration was decided and the patient was taken to the operating room. After the completion of midline suboccipital craniotomy, uvulatonsillary approach was performed and fourth ventricle was reached. Space occupying lesion growing from left middle and superior cerebellar peduncles was disclosed. It was totally subependymal and no clear-cut border was detected. Mass lesion resembled the normal tissue and only the anatomical landmarks helped us to resect it without injuring the brainstem and peduncles. The immediate postoperative period was uneventful. No ictal episodes occurred. Patient became fully alert without any neurological abnormality. Antiepileptic drugs—except phenobarbital with decreasing doses—were ceased. Ten months after the operation, she is still seizure free and her development is appropriate to her age. Postoperative MRI scans have showed gross total resection of tumor.

3. Histopathological findings

Histopathological investigations showed the tumor as *ganglioneurocytoma*. During microscopic examination, intensive cellular aggregations were observed in neurophiles (Fig. 2a). These nodules consisted of small cells with narrow cytoplasms. Within these nodules, there were a few mature ganglion cells which had positive immunoreactions in pericarions with chromogranin (Fig. 2b), S100, neurofilament protein and negative paintings with synaptophysin. Glial fibrillary acidic protein exhibition was also positive in tumor nodules. Previously mentioned small cells are usually a characteristic finding of ganglioneurocytoma, and they are named as *neurocysts*. It may not be possible to differentiate the hamartomatous lesions from this pathology because of identical findings such as giant atypical neurons. However, nodular pattern of lesion, obvious mass formation of these nodules, and lastly very little but countable MIB-1 ($<1\%$) index made us to accept it as a tumoral lesion. If ganglioneurocytoma is compared to hamartomatous neuronal lesions from the prognostic point of view, it is important to follow up the former one with MRI for recurrences. Effects of surgery on seizures have been excellent for both of the lesions.

4. Discussion

To our knowledge, the total number of cases of the fourth ventricular lesions with cerebellar-type epilepsy that have been reported including the case that has been discussed is 14. In previous studies, variety of histological designations were described such as ganglioglioma,[1,6,8,9,11] fibrillary astrocytoma[7] and hamartomatous lesions.[2–5]

Frequent clinical findings of previously reported patients were hemifacial contractions, nystagmoid jerks of both eyes, eye blinking, upper limb involvement and autonomic dysfunction.[1–3] Historically, these attacks were classified as hemifascial spasm.[7–10] In 1996, Harvey et al.[1] localized the attacks in the cerebellum and they introduced the concept of cerebellar epilepsy. Delalande et al.[2] concluded that the seizures arose from inside the lesion and they are special kinds of noncortical seizures similar to gelastic seizures of hypothalamic hamartomas (HHs).

Like the fourth ventricular low-grade lesions, HHs have the potential of epileptogenecity.[2,12] They are frequently presented with gelastic or dacrystic seizures which may evolve into different kinds of seizures by time and lead to epileptic encephalopathy.[12] Intralesioner EEG recordings have revealed that the epileptic discharges are produced by lesion itself. Although the propagation of discharges may follow different