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CASE REPORT

Focal megalencephaly: intraoperative ultrasound imaging in epilepsy surgery

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Abstract Hemimegalencephaly is a rare neuronal migration disorder that can be defined as abnormal neural and glial proliferation localized to all or part of a cerebral hemisphere. Most patients demonstrate intractable epilepsy, with early onset before 1 year of age. Surgical resection is one of the treatment options. In recent years, many advanced intraoperative techniques have been used for brain surgery for various pathologies. Intraoperative ultrasonography is a time-saving and noninvasive method for intraoperative imaging. In this report, we present the use of intraoperative ultrasonography in a patient with focal megalencephaly as an anatomical navigation with the functional navigation system, electrocorticography. In this report, we present the use of intraoperative ultrasonography in a patient with focal megalencephaly as an anatomical navigation with the functional navigation system, electrocorticography.

Keywords Focal megalencephaly · Intraoperative ultrasound imaging · Epilepsy surgery

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Introduction

Hemimegalencephaly (HME) is a rare malformation of cortical development characterized by unilateral excessive growth and dysplasia of one cerebral hemisphere [1]. All or part of a cerebral hemisphere can be involved [2, 3]. The pathogenesis of HME has been associated with a variety of mechanisms, including abnormality in organization, proliferation, and neuronal migration [2]. Patients with HME usually present with developmental delay, psychomotor retardation, and intractable seizures. To control the seizures surgical resection may be required.

In recent years, intraoperative tools, such as intraoperative magnetic resonance imaging (MRI), image navigation systems, and electrocorticography (ECoG), have been used for brain surgery for various pathologies. Intraoperative ultrasonography (IOUS) is one of these techniques. With real-time images, IOUS can inform neurosurgeons about the location of the lesion and its relation to the eloquent areas. We present the use of IOUS in a patient with focal megalencephaly as an anatomical navigation to locate and reach the site indicated by ECoG. This is the first case of focal megalencephaly using IOUS in the literature.

Case report

A 12-month-old male presented with medically intractable seizures since he was 7 months old. During a seizure, his eyes deviated upward and there was distonic posture of the left arm. The frequency of seizures was initially 5–6 times/day. However, in 2 months it increased to 20–30 times/day. Neurological examination showed no pathology except for left hemiparesia, which developed following the increase in the frequency of seizures. The seizures could not be kept under control despite the use of many medications. During video monitoring electroencephalogram (EEG), interictal EEG showed slow baseline activity in the frontal area of the right hemisphere, and the ictal EEG showed that there were epileptiform discharges arising from the same area. The patient underwent 3-Tesla MRI. MRI showed focally enlarged frontal and anterior temporal lobes with expanded gyri and ill-defined gray and white matter differentiation. The findings were consistent with focal megalencephaly (Fig. 1). The intractable and frequent seizures forced the family and physicians to choose surgical intervention.

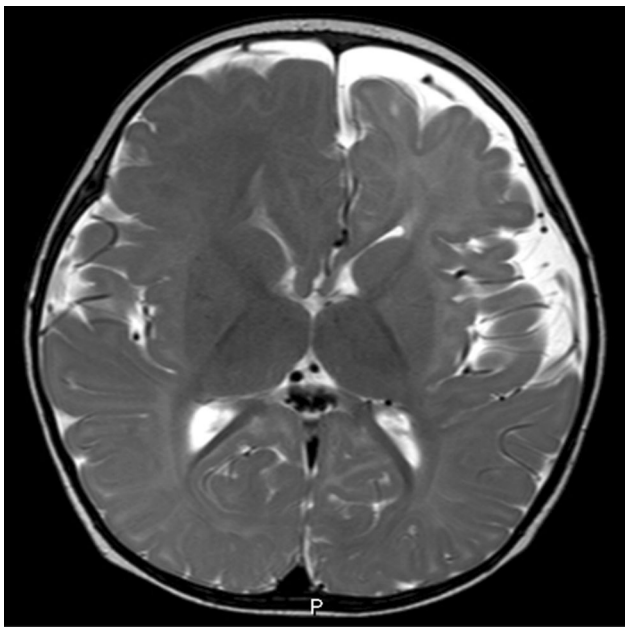

Fig. 1 T2-weighted axial 3T-MRI of focally enlarged right frontal lobe with expanded gyri and ill-defined *gray* and *white* matter differentiation

During the operation, NIM-Eclipse version 3.5.350 (Medtronic XomedInc., USA) was used for ECoG recording. Following the anesthesia, based on the international 10–20 system, the reference electrode was placed at Fpz localization on the scalp and the ground electrode was placed on the right shoulder. Subdermal needle electrodes were used. After opening the dura mater, a 2 × 8 strip electrode (8 contact, 2 strip electrode 13 mm, 27G-Inomed) was placed on the right frontal and temporal lobes for ECoG recording. Identification of epileptic focuses was attempted by changing the location of the electrode from front to back. Slow wave, isolated sharp wave, spike, spike wave, and electrophysiological seizure records were identified as abnormal findings and they were defined as the area of epileptic focus (Fig. 2a, b).

Transcortical ultrasound was performed for the purpose of anatomic mapping, as well as functional purpose. IOUS helped to locate and show the affected areas detected with ECoG by real-time imaging. We performed IOUS using aLogiq p6 (GE Healthcare, Washington) and a linear transducer (7–11 MHz). The probe was placed on the frontal pole and displayed the abnormal frontal lobe parenchyma with homogeneously hyperechoic expanded gyri (Fig. 3). After resection, we controlled the residual parenchyma with ECoG and detected activation in the medial frontal lobe. This area was confirmed by IOUS as showing expanded gyri and hyperechoic parenchyma (Fig. 3c). After resection and hemostasis, IOUS and ECoG were used to confirm the resection (Fig. 3d). The resection

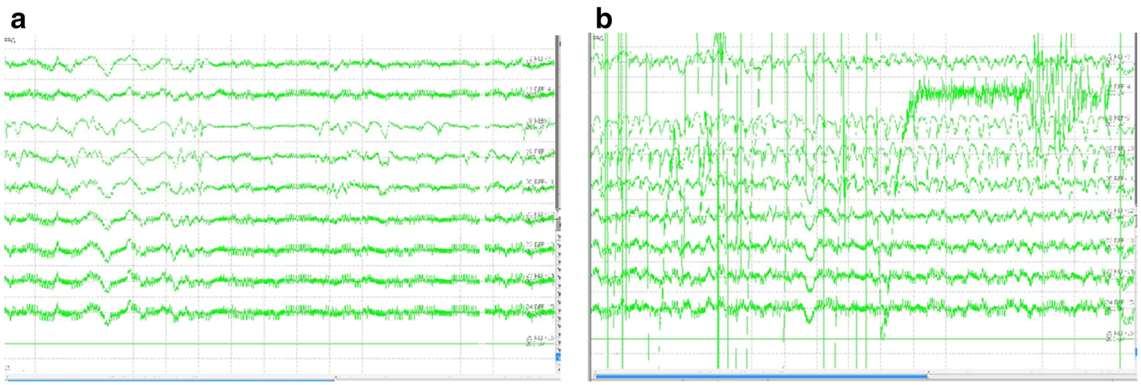

Fig. 2 Intraoperative electrocorticography a 2 × 8 strip electrode (8 contacts, 2 strip electrodes 13 mm, 27G-Inomed) was placed on the right hemisphere. Slow wave, isolated sharp wave, spike, spike wave (**a**), and electrophysiological seizure record were identified as abnormal findings, and these areas were identified as the epileptic focus (**b**)

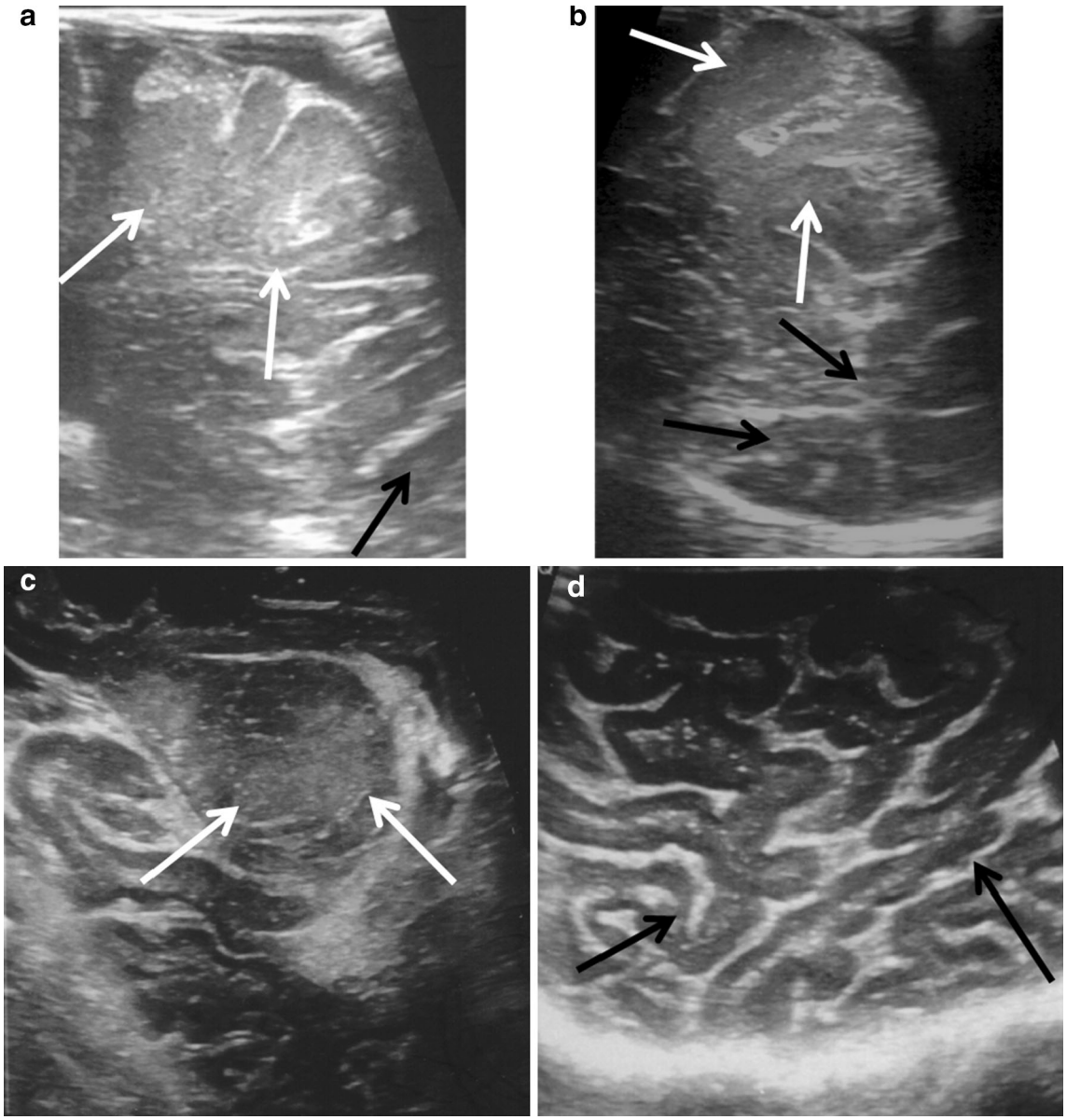

Fig. 3 IOUS images. (**a**, **b**) The cortical dysplasia area of the frontal lobe is characterized with homogeneously hyperechoic expanded gyri (*arrows*). Normal parenchyma (*black arrows*). (**c**) IOUS of the residual dyplastic parenchyma after resection in which activation was detected with ECoG (*arrows*). (**d**) IOUS of normal parenchyma (*black arrows*) after resection and hemostasis

was terminated after ECoG recording; it showed that there were no electrophysiological abnormalities. During the resection, the eloquent cortex was preserved anatomically by ultrasound guidance, and functionally by ECoG.

The histopathologic features were compatible with cortical dysplastic changes associated with focal megalencephaly (Fig. 4).

Discussion

Epilepsy is one of the most common neurological disorders in the pediatric population. About 25–40 % of patients with epilepsy are refractory to medications [4]. In hemimegalencephaly, the extensive dysplasia usually involves an entire cerebral hemisphere but sometimes only appears over a partial area of one hemisphere, which is referred to as "focal megalencephaly" [5]. Marked hyperexcitability of the dysplastic cortex usually results in a catastrophic epileptic syndrome in patients. Epileptic seizures represent the main manifestations of HME, with seizures occurring in more that 90 % of the patients [2]. The treatment of epilepsy in HME includes pharmacological and surgical approaches. Although there are lots of available antiepileptic drugs, surgery may be appropriate for some of these patients after extensive investigations and careful selection.

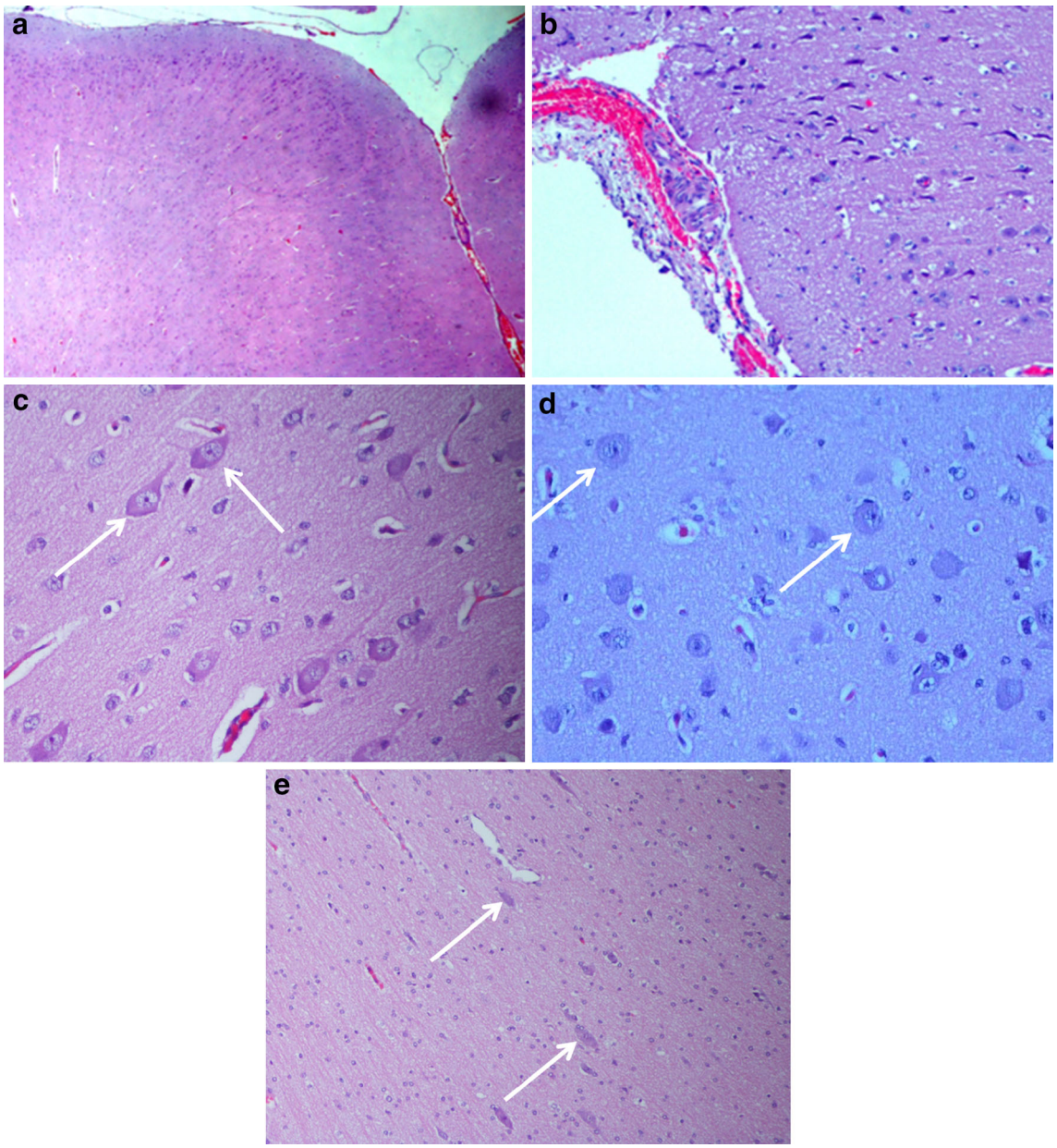

Fig. 4 **a** The cortex consists of poorly organizing neurons (×40, H&E). **b** At high power, the cerebral cortex can be seen to demonstrate disorganization of the normally laminar pattern of neuronal nuclei and abnormal dysmorphic neurons (×100, H&E). **c** In addition to enlargement of the neuronal perikarya, dysmorphic neurons (*arrows*) are characterized by increased density of cytoplasmic neurofilaments, which impart an irregular and hyperchromatic appearance to the neuronal cell bodies (×400, H&E). **d** Ballooned cells (*arrows*) can be appreciated by their abundant homogeneous glassy pink cytoplasm and eccentric large nuclei, sometimes with visible nucleoli (×400, H&E). **e** Dysmorphic neurons (*arrows*) are present in the white matter

To avoid suppressing the development of children, resection surgery should be performed as early as possible.

The surgical results of diffuse cortical dysplasias are worse than those of focal cortical dysplasias. Successful outcomes of surgeries depend on the complete resection of the lesion. For that reason, many intraoperative tools are used during resection, such as ECoG recording, image navigation systems, and intraoperative imaging. However, these tools have their own respective disadvantages. ECoG recording is a functional navigation, and recordings only provide a two-dimensional representation of the irritative zone with variable relation to the lesion [6]. Brain shifting is the main problem of neuronavigation based on preoperative MRI images. After opening the dura, the brain shifts because of cerebrospinal fluid drainage or leaks and tissue resection [7]. Intraoperative MRI takes a lot of time during the operation and the resolution and details are not as good as those in preoperative studies.

IOUS is used especially in intra-axial brain tumors in neurosurgery [8]. It has the advantages of continuous and real-time visualization of intracranial lesions. It is easily accessible, inexpensive, widely available, and provides the experienced user with real-time information. It provides three-dimensional information about the extension of the lesions to the neurosurgeon, while protecting the eloquent areas, independent of the brain shift.

Miller et al. [9] and Lee et al. [10] presented cases with focal cortical dysplasia type IIb and type IIa, respectively, in which they used IOUS. We used IOUS with EcoG to define the lesion and identify the shortest way to reach it by real-time imaging in epilepsy surgery for a case of focal megalencephaly. In the future, IOUS can be used as an anatomical navigation tool for the detection of dysplastic cortex intraoperatively. The extent of the usage of IOUS requires further comparative studies. The sensitivity and specificity for different types of dysplastic changes and other pathologies need to be determined. We should work on various images of dysplastic cortex to improve the capability of the differential diagnosis and interpret the findings accurately.

Conflict of interest There are no financial or other relations that could lead to a conflict of interest.

Ethical standard This article does not contain any studies with human or animal subjects performed by any of the authors.

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