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Review

Pigmented ganglioglioma in a patient with chronic epilepsy and cortical dysplasia

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ABSTRACT

We report a rare case of a 22-year-old woman with biopsy-proven pigmented ganglioglioma. The patient initially underwent a right temporal lobectomy for intractable seizures at the age of 9 and remained seizure free for several years but subsequently developed complex partial seizures. Due to enhancement of a left mesial occipital lesion on preoperative MRI of the brain, the patient underwent a left subdural electrode placement and simultaneous biopsy of the left mesial occipital lesion. Biopsy results revealed a rare pigmented ganglioglioma, World Health Organization Grade I. The seizure focus was identified in the left mesial occipital lobe and the patient underwent tumor resection. An extensive literature search revealed that our patient is the fourth case of pigmented ganglioglioma described in the literature and was positive for *BRAF* V600E mutation by molecular studies.

1. Introduction

Gangliogliomas are rare primary central nervous system tumors occurring frequently in the first three decades of life in those with chronic focal epilepsy [1] and are generally World Health Organization (WHO) Grade I [2]. These tumors are largely benign with the most common cause of malignant transformation being radiation-induced [3]; there are rare reports of spontaneous transformation after surgery [2,4]. Pigmented neural cell tumors are well described in the literature but infrequent, especially when presenting as primary neuroepithelial tumors. The pigmented cells contain either melanin [5–11], neuromelanin [12–14], hemosiderin [15,16], or lipofuscin [13,14]. Reported cases of pigmented neuroepithelial tumors in the literature include ependymoma [5,13,17], subependymoma [17], ganglioglioma [6,9,15], pilocytic astrocytoma [12], pleomorphic xanthoastrocytoma [7,8,10,11], desmoplastic infantile ganglioglioma [18], desmoplastic non-infantile ganglioglioma [19], choroid plexus papilloma [20], choroid plexus carcinoma [21], medulloblastoma [22,23], primitive pineal tumor [24], and central neurocytoma [14,16]. Primary central nervous system tumors of melanocytic origin include diffuse melanosis, melanocytoma, and malignant melanoma. We report the fourth case of pigmented ganglioglioma described in the literature which occurred in a patient with biopsy proven cortical dysplasia.

2. Clinical summary

The patient is a 22-year-old woman with a history of intractable seizures beginning at 16 months of age. Her complex partial seizures consisted of staring spells and picking automatisms. In May 2001 at the age of 9, she underwent a right temporal lobectomy after monitoring revealed a corresponding epileptic focus. Her contemporaneous brain MRI revealed evidence of right hippocampal sclerosis and a small cystic area in the left mesial occipital lobe (Fig. 1). The left mesial occipital lobe lesion was followed with serial MRI over many years and found to be stable in size and character. Pathology from the right temporal lobectomy showed clusters of dysplastic neurons, consistent with cortical dysplasia (Fig. 2).

At the age of 11, the patient's seizures returned and despite continued medical therapy, her seizures continued. In 2008 she experienced an episode of confusion and underwent non-invasive monitoring that revealed seizure onset from the left temporal region. Subsequently, she began experiencing seizures almost daily. Her seizures progressed to involve staring spells with or without bilateral upper extremity automatisms. She also experienced ictal speech and speech arrest.

Due to the increase in seizure frequency, in August 2014 the patient was deemed an appropriate candidate for placement of a

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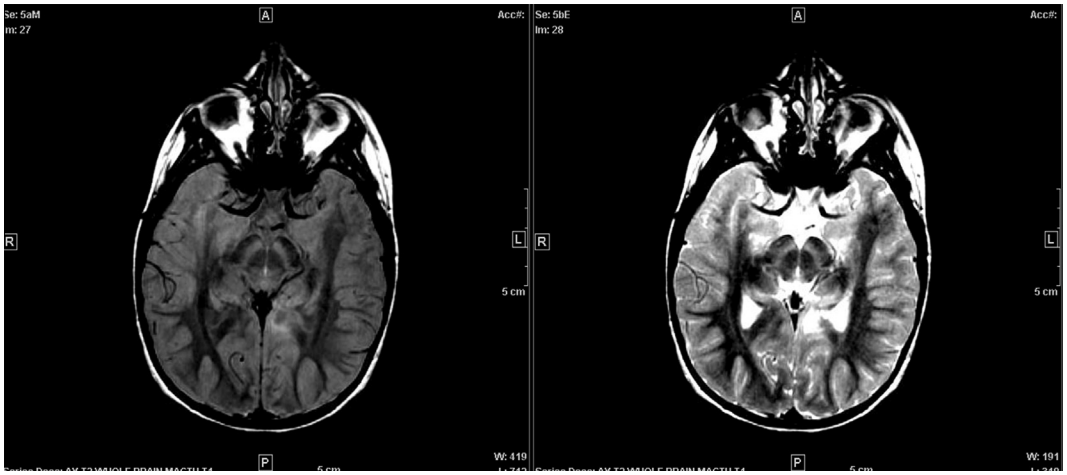

Fig. 1. Pre-contrast T1-weighted axial MRI (left) and T2-weighted axial MRI (right) revealing a left mesial occipital cystic lesion in January 2001.

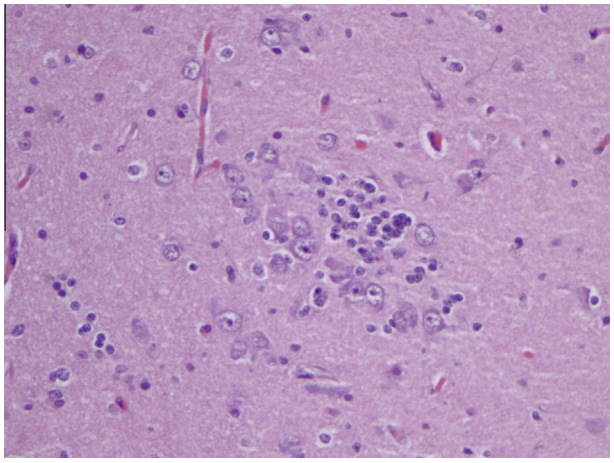

Fig. 2. Cortical dysplasia (original resection from 2001). The cortex displays clusters of dysplastic neurons and glial cells (hematoxylin and eosin, original magnification ×40). This figure is available in colour at http://www.sciencedirect.com/.

responsive neurostimulation (RNS) device. Preoperative evaluation included a new brain MRI which, when compared to her original MRI in 2001, showed interval enlargement of the left mesial occipital cystic lesion (Fig. 3). Plans for RNS device placement were subsequently aborted and the patient underwent biopsy of this lesion and placement of subdural electrodes for monitoring in the occipital interhemispheric fissure. Frozen section at time of surgery was consistent with low grade glioneuronal tumor. Electroencephalogram monitoring from the subdural electrodes revealed epileptiform activity from the area of the lesion and 3 days later she underwent resection. Final pathology returned as pigmented ganglioglioma, WHO Grade I.

3. Materials and methods

3.1. Intraoperative findings

Our patient was brought to the operating room and underwent general anesthesia. A lumbar drain was initially placed perioperatively to allow cerebrospinal fluid drainage for additional brain relaxation and minimal retraction on brain parenchyma. She was positioned with her left side down to allow gravity to assist in retracting the left occipital lobe from the midline and minimal retractor use. A left-sided paramedian linear scalp incision was used to expose the occipital bone and a craniotomy was performed to expose the underlying occipital dura just off midline. The dura was opened and reflected medially against the superior sagittal sinus allowing the occipital interhemispheric fissure to be dissected. An abnormality was visualized in the deep mesial

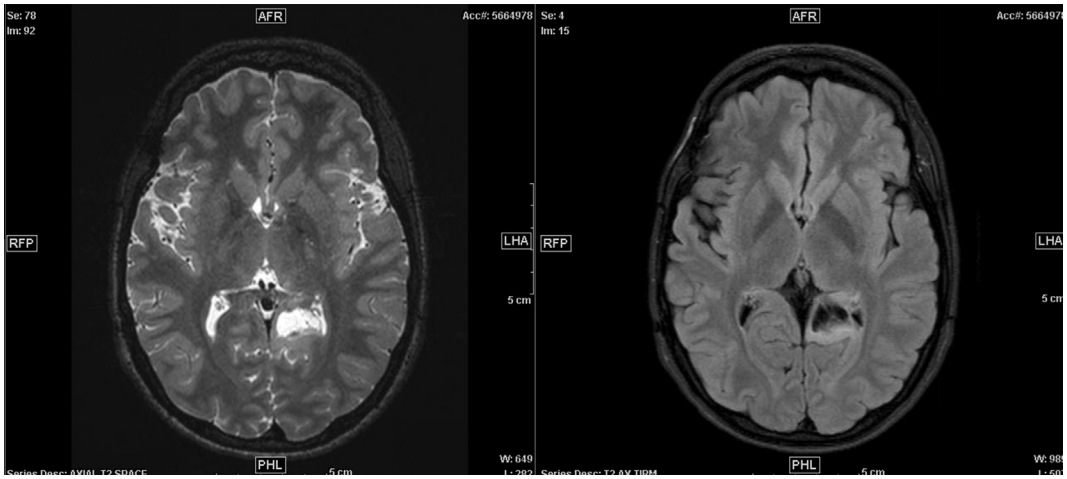

Fig. 3. T2-weighted axial MRI (left) and T2 axial fluid attenuated inversion recovery MRI (right) revealing a lobulated cystic area in the left posterior mesial occipital lobe.