Clinical commentary

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# Late adult-onset epilepsy in a patient with hemimegalencephaly

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**ABSTRACT** – Hemimegalencephaly is a malformation of cortical development that normally manifests in childhood with seizures and cognitive impairment. We present a case of hemimegalencephaly in a 55-year-old developmentally-normal woman who developed focal impaired awareness seizures with bilateral tonic-clonic spread. Her interictal EEG showed left-sided multifocal spikes, and ictal EEG showed seizures starting in the left hemisphere. Imaging showed hemimegalencephaly. This case may represent the oldest age for a first seizure in a patient with hemimegalencephaly.

**Key words:** focal epilepsy, malformations of cortical development, hemimegalencephaly

Hemimegalencephaly is congenital malformation characterized by overgrowth of one cerebral hemisphere (Wu *et al*., 2014). This malformation of cortical development normally manifests as an epileptic encephalopathy with cognitive impairment and focal seizures in infancy. It is rarely described in adults (Wu *et al*., 2014). There are two reported cases of 19-year-olds with a first seizure caused by hemimegalencephaly, but there are no documented cases of patients with a first seizure occurring at older than 19 years (Ikeda and Mirsattari, 2017).

Hemimegalencephaly can be associated with segmental overgrowth syndromes and somatic mutations in phosphoinositide 3-kinase (PI3K), protein kinase B (Akt), and mammalian target of rapamycin (mTOR) pathways (Lee *et al*., 2012).

## Case study

A 55-year-old, left-handed woman, the only left-handed person in the family, presented to the emergency department after a first seizure. She was at home when she had two bilateral tonic-clonic seizures of unknown onset. She would later develop seizures with ictal speech with reversion to Portuguese. With these seizures, she developed automatisms and then bilateral tonic-clinic seizure activity. She had a history of depression and hypertension and took medications for these conditions. She was born in Portugal and had normal development except that she first walked at the age of three. She moved to Canada at eight years old and completed high school and then worked in the healthcare field.

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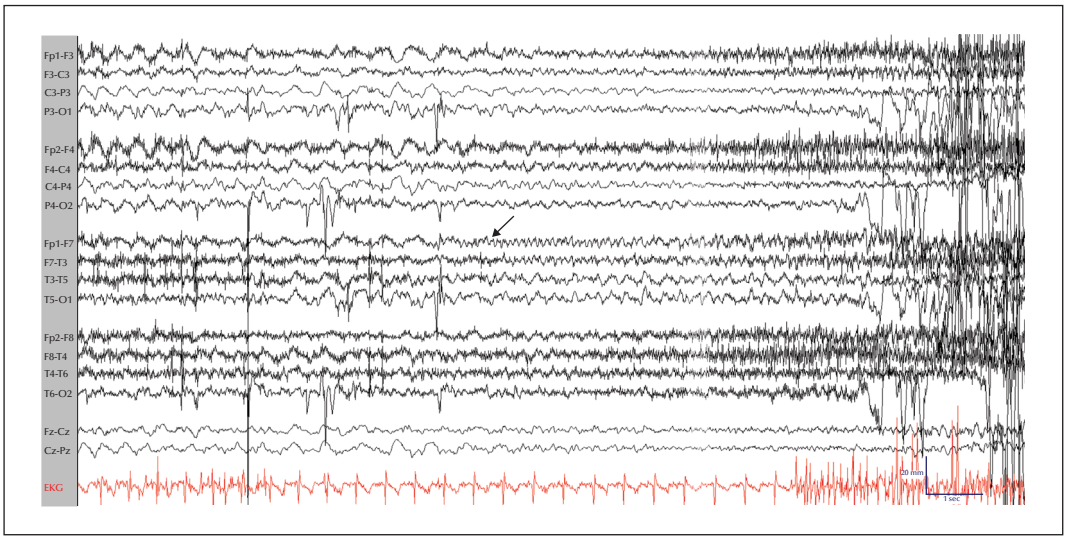


**Figure 1.** Ictal EEG recording of a developmentally-normal woman who had a first ever epileptic seizure at 55 years old (longitudinal bipolar montage). Rhythmic left frontal activity (arrow) is seen preceding clinical onset. During this event, she had speech reversion to Portuguese, followed by bilateral tonic-clonic activity. All EEG electrodes were placed using the international 10-20 system for electrode placement.
Low frequency filter: 1 Hz; high frequency filter: 70 Hz; notch off; sensitivity: 7 μV/mm; time base: 30 mm/s; sampling rate: 500 Hz.

Her examination was significant for right hand polyminimyoclonus and subtle loss of right-sided dexterity. She had full strength and symmetric reflexes. She had no skin abnormalities and no language abnormalities.

She was later admitted to the epilepsy monitoring unit for continuous video-EEG. Mild diffuse slowing, maximum in the left hemisphere, was identified. Frequent, interictal spikes were captured. Most spikes were left fronto-temporal or left diffuse, but rare independent right frontal-central or generalized discharges also occurred. Captured seizures revealed ictal speech reversion, left body turning, followed by bilateral tonic-clonic seizure activity. Rhythmic activity originated diffusely from the left hemisphere, but rapidly became generalized (*figure 1*).

T1-weighted magnetic resonance images revealed an enlarged left hemisphere (*figure 2*). A diagnosis of hemimegalencephaly was made.

Given her preserved language and motor skills, functional hemispherotomy was not offered. She underwent vagus nerve stimulator implantation and her anti-seizure medications were adjusted, but she still has occasional seizures.

## Discussion

Although hemimegalencephaly usually presents in children as an epileptic encephalopathy, this case demonstrates that adult patients with normal cognition can have late-onset seizures caused by hemimegalencephaly. This case may represent the oldest age at which a first seizure can occur in a patient with hemimegalencephaly. □

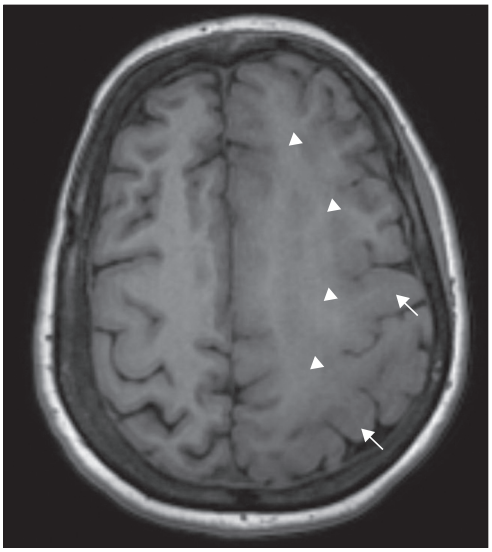

**Figure 2.** Axial T1-weighted MR images showing left-sided cortical thickening (arrows), deep white matter heterotopias, and abnormal white matter signal (arrowheads), consistent with hemimegalencephaly.

**Supplementary data.**
Summary didactic slides are available on the www.epilepticdisorders.com website.

**Disclosures.**
None of the authors have any conflict of interest to declare.

## References

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**TEST YOURSELF**

(1) What is the typical age for presentation of hemimegalencephaly?

(2) What are the imaging findings in hemimegalencephaly?

(3) Somatic mutations in which genetic pathways are associated with hemimegalencephaly?

*Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com, under the section "The EpiCentre".*