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Childhood-onset nonprogressive chronic encephalitis

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ABSTRACT

Purpose: The purpose of this study was to describe a series of patients with pathologically proven chronic encephalitis who had a nonprogressive course during a long follow-up, suggestive of a "benign" variant of Rasmussen's encephalitis (RE).
Methods: Four patients who were referred to our Comprehensive Epilepsy Program at London Health Science Centre in London, Ontario, were diagnosed with chronic encephalitis on a pathological basis after epilepsy surgery to treat their partial-onset seizures.
Results: None of our four cases followed the typical course of RE despite their childhood-onset seizures between ages 2 and 12 years. One was preceded by a mild head trauma and fever at onset. None had epilepsia partialis continua (EPC). Their long-term follow-up revealed a nonprogressive form of the syndrome with respect to the neurological examination, EEG, MRI, and neuropsychological findings.
Conclusion: These cases extend the spectrum of childhood-onset intractable epilepsy with chronic encephalitis to include nonprogressive variants of RE. The absence of EPC may be a prognostic indicator of a nonprogressive course.

1. Introduction

Rasmussen's encephalitis (RE) is an inflammatory neurological disease that manifests with medically intractable partial seizures and a progressive clinical course including unilateral cerebral atrophy, hemiparesis, and cognitive impairment [1]. The exact etiology of RE is unknown, although viral causes [2–4], autoantibodies [5–9], and cytotoxic T-cell mechanisms [10,11] have been proposed. Antiepileptic drugs (AEDs) are usually ineffective in treating the seizures in RE, while hemispherectomy is effective in childhood in arresting its progression. Since its first description by Rasmussen and colleagues in 1958 [2], a number of atypical features are reported that have widened the clinical spectrum of RE [12]. We present four cases of pathologically confirmed chronic encephalitis, all of which had childhood-onset intractable partial seizures with or without secondary generalization. None of them experienced significant neurological or radiological decline despite continued partial seizures.

Abbreviations: RE, Rasmussen's encephalitis; CE, chronic encephalitis; EEG, electroencephalography; MRI, magnetic resonance imaging; EPC, epilepsia partialis continua.

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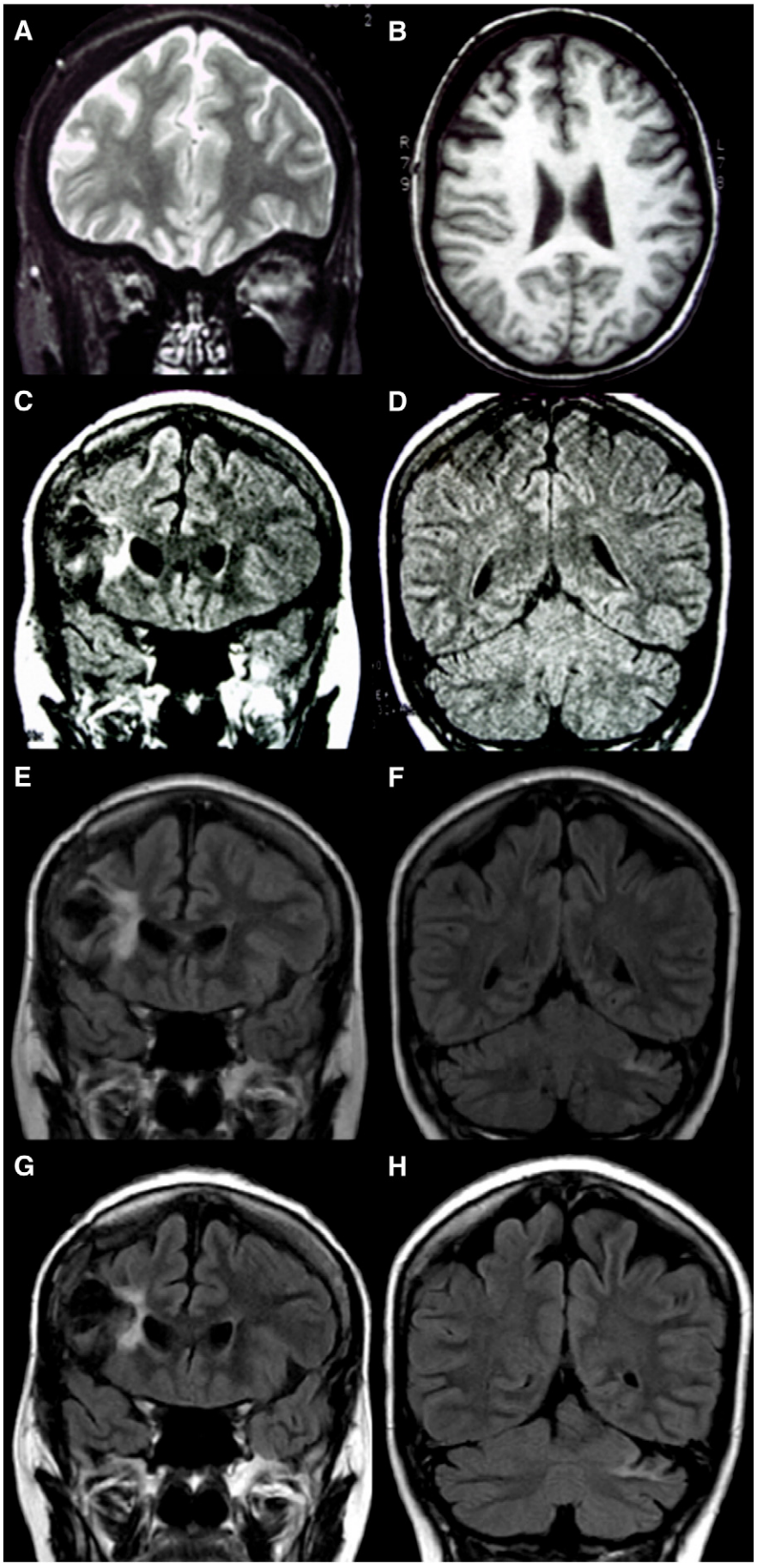
2. Case histories

Four patients referred to our Comprehensive Epilepsy Program were unexpectedly diagnosed with chronic encephalitis on a pathological basis after epilepsy surgery to treat their medically intractable partial-onset seizures.

2.1. Patient 1

This 38-year-old, right-handed woman began having seizures at age 12 years with multiple seizure types, which were refractory to AEDs. Her most frequent seizures consisted of facial twitching around her nose bilaterally with inability to speak while retaining awareness and comprehension, occurring up to 4–5 times per day. She also experienced motionless staring spells with retained awareness. Occasionally, she had head turning to the left with clonic jerking of the left hand and mouth with inability to speak but retained awareness followed by secondary generalization. Postictally, she was unable to move or talk for about 15 min. Preoperative EEG at age 24 years demonstrated right frontal spikes and seizures; her cranial MRI demonstrated right frontal cortical atrophy (Figs. 1A, B). Preoperative neuropsychological testing showed frontal lobe dysfunction maximal in the right hemisphere manifesting with difficulties in some executive abilities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