■ Case vignette

Epileptic Disord 2022; 24 (2): 423-426

Epileptic Disorders

Adult new-onset status epilepticus secondary to en coup de sabre: a case report and review of the literature

Hera A. Kamdar, Jan Bittar, Sarita Maturu

Department of Neurology, The Ohio State University, Columbus, Ohio, USA

Received June 2, 2021; Accepted September 27, 2021

• **Correspondence:**
Sarita Maturu
Department of Neurology
The Ohio State University
395 West 12 Avenue, Columbus OH 43210, USA
<Sarita.maturu@osumc.edu>

doi:10.1684/epd.2021.1387

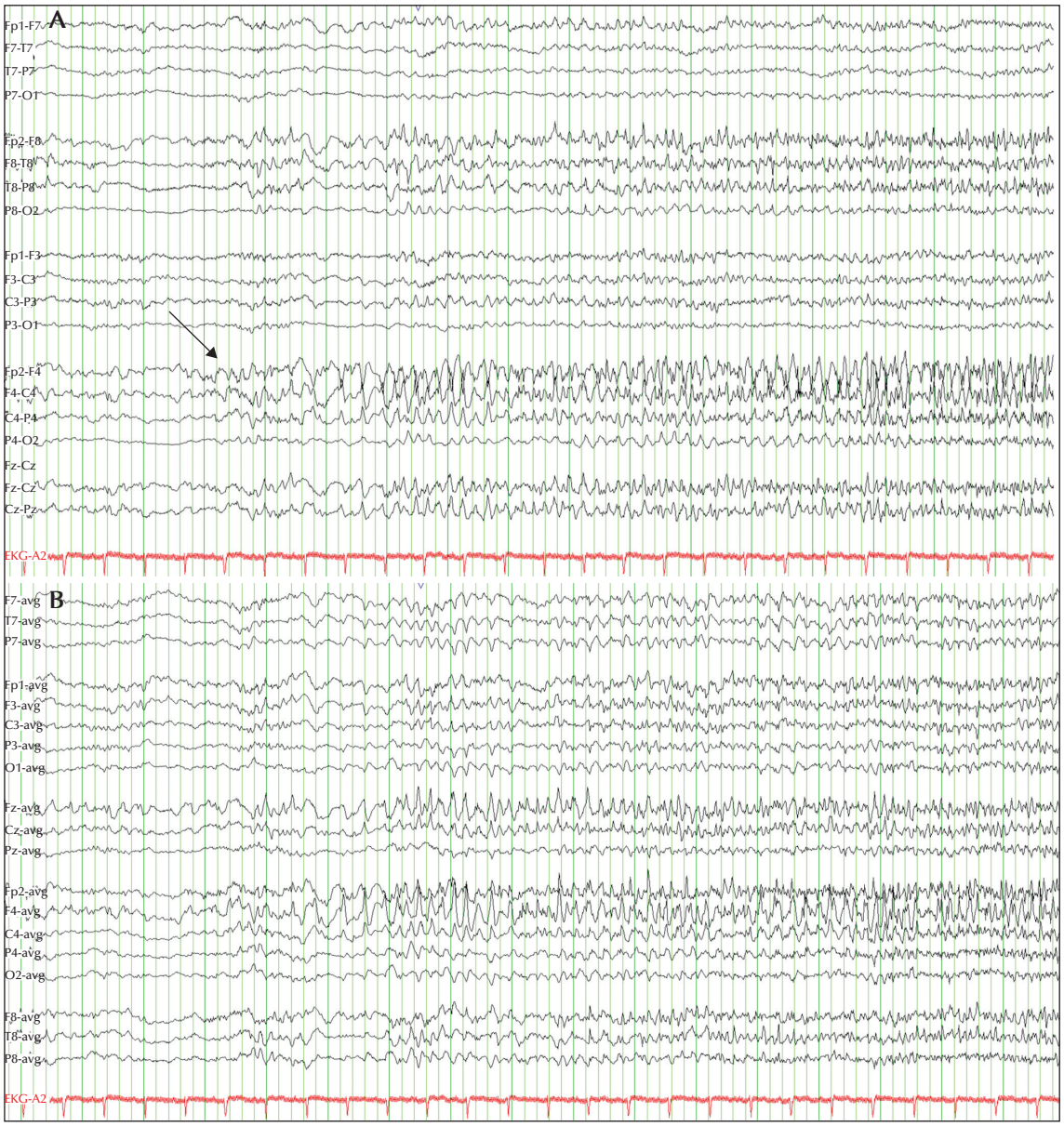
En coup de sabre (ECDS) is a rare form of linear scleroderma that forms a depressed groove on the scalp and linear alopecia [1, 2]. The neurological manifestations of ECDS are rare and typically recognized after dermatological presentation. Seizures are the most frequently reported neurological abnormality [3, 4]. Here, we report a unique case of ECDS presenting as new-onset status epilepticus (SE) in adulthood before the diagnosis of skin disease.

A 58-year-old female with no prior history of seizures, risk factors for epilepsy, or autoimmune disease, presented to the hospital after a first-time focal to bilateral tonic-clonic seizure. The patient was given lorazepam at 4 mg and loaded with fosphenytoin at 20 mg/kg. She remained in SE, and thus was loaded with levetiracetam at 40 mg/kg and intubated for airway protection. The patient was placed on continuous EEG monitoring which demonstrated frequent right fronto-central electrographic seizures with correlating forced left head deviation and left upper and lower extremity posturing (*figures 1A, B*). A versed drip was needed for the management of SE and lacosamide at 200 mg was administered.

An MRI of the brain with contrast was performed which revealed right frontal dural enhancement along with a right hemispheric cortical FLAIR hyperintensity with surrounding edema, mainly involving the right frontal region (*figure 2A, B*). A lumbar puncture was unrevealing other than a nonspecific mild protein elevation of 58 mg/dL. Careful skin examination revealed an atrophic scar on the scalp that was present since birth (*figure 2C*) overlaying the cortical FLAIR hyperintensity and edema seen on MRI. Further workup with skin biopsy and rheumatologic serum studies, including anti-scleroderma-70 (Scl-70) and anti-centromere antibodies, were nonspecific and unrevealing. Given her clinical presentation and dermatologic examination, she was diagnosed with ECDS.

The patient was discharged on levetiracetam at 1,500 mg twice daily, lacosamide at 100 mg twice daily, and phenytoin at 50 mg three times a day which was tapered off by 50 mg a week. She was also started on immunosuppression therapy with methotrexate.

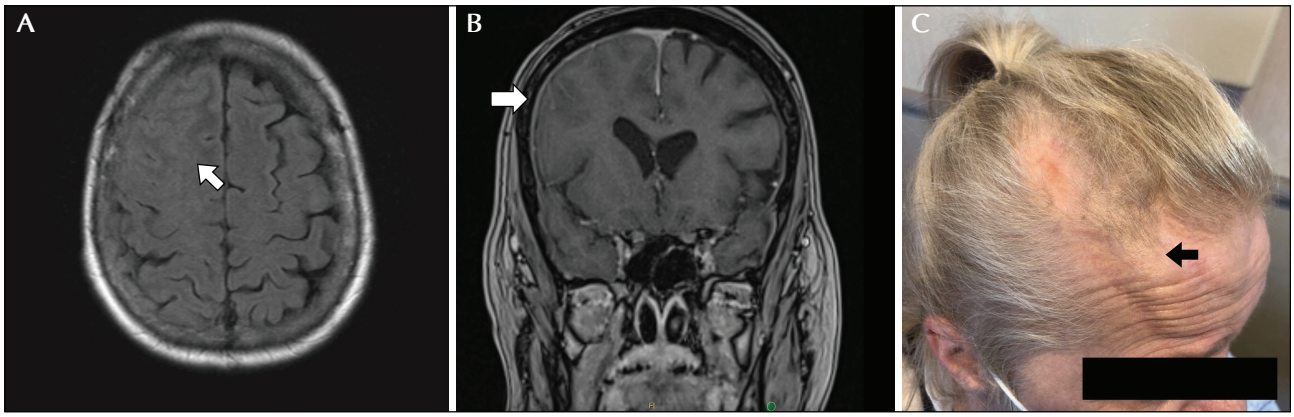
ECDS is a clinical diagnosis primarily defined by its characteristic scalp lesion that is described as an ivory-colored, sclerotic plaque present on the forehead [4, 5]. This condition mainly affects children, and previous studies report that 67% of patients are diagnosed prior to the age of 18 [6]. A common oral manifestation of ECDS is altered dentition [4]. Reported neurological manifestations include seizure disorder, muscle weakness, cranial nerve involvement, intracranial aneurysm, and migraine headaches [3]. Frequently seen ophthalmologic signs and symptoms include ptosis, exophthalmos, and ocular muscle atrophy [7]. Our patient presented with altered dentition, the



■ **Figure 1.** Right fronto-central electrographic seizures. (A) Bipolar longitudinal montage demonstrates rhythmic right fronto-central spike and wave discharges at onset of seizure (arrow). (B) The average referential montage shows maximum negativity over the right fronto-central region.

characteristic scalp lesion, underlying edema of the cerebral cortex, and seizures justifying a diagnosis of ECDS. Interestingly, she was an adult during her time of presentation and her initial symptom was SE. The pathogenesis of ECDS is not well understood, but there have been proposed hypotheses. One hypothesis suggests that there is an inflammatory response within the brain parenchyma and meninges that causes a local perivascular inflammation or vasculitis [4]. Certain infectious diseases, such as Lyme disease, have been reported to incite ECDS. There have also been theories about genetic contributions [4]. Our patient reported no infectious symptoms prior to her presentation, had a negative infectious work-up, and denied any family history of rheumatologic or neurologic disease.

There are no current diagnostic laboratory tests for ECDS, however ANA, anti-single-stranded DNA



■ **Figure 2.** (A) Axial FLAIR image on brain MRI revealed right frontal FLAIR hyperintensity and edema. (B) Coronal T1 image post contrast on brain MRI demonstrated right frontal pachymeningeal dural enhancement. (C) Skin examination showed correlating atrophic scar on the right scalp with grooved appearance and surrounding alopecia.

antibodies, and more specific antibodies preceding systemic sclerosis such as anti-Scl-70 and anti-centromere are still routinely checked. In our case, these serological studies were negative, which can further explain the patient's lack of systemic disease [4]. Definitive treatment for ECDS remains difficult. Corticosteroids can be used for cutaneous skin lesions to reduce inflammation. Vitamin supplements, immunosuppressants, and UV-A therapy can be used if systemic symptoms are present and more aggressive therapy is needed [5]. This patient was started on oral methotrexate as she was diagnosed with a severe neurological manifestation.

We present a unique case of ECDS for two salient reasons. First, ECDS is known to be most commonly diagnosed in childhood, however, our patient was diagnosed after serious neurological manifestations in adulthood. Second, this case highlights the importance of a careful skin examination in patients who present with unexplained new-onset seizures. ■

Supplementary material.
Summary slides accompanying the manuscript are available at www.epilepticdisorders.com.

Disclosures.
Dr. Sarita Maturu is on the Executive board of My Epilepsy Story. Drs. Hera Kamdar and Jan Bittar have nothing to disclose.

References

1. Careta MF, Romiti R. Localized scleroderma: clinical spectrum and therapeutic update. *An Bras Dermatol* 2015; 90(1): 62-73.

2. Stone J, Franks AJ, Guthrie JA, Johnson MH. Scleroderma "en coup de sabre": pathological evidence of intracerebral inflammation. *J Neurol Neurosurg Psychiatry* 2001; 70: 382-5.

3. Flores-Alvarado DE, Esquivel-Valerio JA, Garza-Elizondo M, Espinoza LR. Linear scleroderma en coup de sabre and brain calcification: is there a pathogenic relationship? *J Rheumatol* 2003; 30(1): 193-5.

4. Holland KE, Steffes B, Nocton JJ, Schwabe MJ, Jacobson RD, Drolet BA. Linear scleroderma en coup de sabre with associated neurologic abnormalities. *Pediatrics* 2006; 117(1): e132-6.

5. Peña-Romero AG, García-Romero MT. Diagnosis and management of linear scleroderma in children. *Curr Opin Pediatr* 2019; 31(4): 482-90.

6. Peterson LS, Nelson AM, Su WP, Mason T, O'Fallon WM, Gabriel SE, *et al*. The epidemiology of morphea (localized scleroderma) in Olmstead Country 1960-1993. *J Rheumatol* 1997; 24: 73-80.

7. Gambichler T, Kreuter A, Hoffmann K, Bechara FG, Altmeyer P, Jansen T. Bilateral linear scleroderma "en coup de sabre" associated with facial atrophy and neurological complications. *BMC Dermatol* 2001; 1: 9.

TEST YOURSELF

(1) Diagnosis of en coup de sabre is based on which of the following?

A. Positive anti-Scl-70 and anti-centromere serology

B. Local sclerosis and calcium deposition based on skin biopsy of the lesion

C. Clinical examination and history with presence of characteristic forehead/scalp lesion

D. EEG with focal to bilateral tonic clonic seizures

(2) At what age does en coup de sabre typically present

A. Neonates

B. <18 years old

C. 25 years old

D. >55 years old

(3) Which of the following is the most common neurological manifestation of en coup de sabre?

A. Trigeminal neuralgia

B. Facial palsy

C. Muscle weakness

D. Seizure disorder

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com.