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Electroencephalographic features of moyamoya in adults

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HIGHLIGHTS

- Patients with moyamoya often have transient neurologic events such as altered mental status, aphasia, limb shaking, or facial twitching, which lead to referral for EEG.
- In the inpatient setting, particularly post revascularization surgery, epileptiform discharges were seen in 24% and seizures were seen in 12%.
- Seizures should be considered, along with transient ischemia, on the differential diagnosis of adult moyamoya patients.

ABSTRACT

Objective: Electroencephalography is useful for evaluating transient neurological events in the setting of moyamoya disease.
Methods: EEG findings of adults with moyamoya seen at a large moyamoya referral center are summarized. Patients were identified by retrospective chart review.
Results: EEGs were ordered after cerebral revascularization for altered mental status, aphasia, limb shaking, or facial twitching. Among the study population of 103 patients having EEGs, 24% of adults with moyamoya had a history of clinical seizures. Ischemic or hemorrhagic strokes were associated with a twofold relative risk of seizures. Overall, 90% of EEGs were abnormal, most commonly focally (78%), or diffusely slow (68%). Epileptiform EEG discharges were seen in 24%. Whereas hemispheres with an ischemic stroke had a 19% risk of epileptiform discharges and an 8% risk of seizures on EEG, hemispheres with hemorrhagic stroke had a 35% risk of epileptiform discharges and 19% risk of seizures on EEG. Focal amplitude attenuation was seen in 19%, breach rhythm in 15%, rhythmic delta in 14%, and electrographic seizures in 12%.
Conclusions: Seizures and epileptiform EEG changes are common in patients with moyamoya disease.
Significance: Transient events in patients with moyamoya can result from seizures as well as ischemia.

1. Introduction

Moyamoya disease is a chronic occlusive vasculopathy involving the distal supraclinoid internal carotid arteries, presenting with ischemic or hemorrhagic symptoms (Pandey et al., 2011). Transient neurologic events can pose diagnostic dilemmas in people with moyamoya. Given the pathology of this progressive cerebral vasculopathy, etiologies of neurologic symptoms, often recurrent and stereotyped, naturally include transient ischemic attacks (which can manifest as limb movements), ischemic strokes, and recrudescence of prior strokes during intercurrent illness or hemorrhage (Kraemer et al., 2012). Migraines are also seen, which are sometimes associated with aura or hemiplegia (Zach et al., 2010). Movement disorder is an uncommon presentation (Pandey et al., 2010). Seizures can be seen in 6–12% of pediatric moyamoya cases (Scott et al., 2004; Nakase et al., 1993). In adults, the prevalence of seizures may be higher, in one series reaching 18% of revascularized hemispheres (Jin et al., 2011). The high prevalence of seizures in adults with moyamoya might reflect a higher prevalence of

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intracranial hemorrhage (Kuroda and Houkin, 2008), given that the relative risk of seizures is higher with intracranial hemorrhage than with ischemic stroke (So et al., 1996).

Electroencephalography (EEG) can be useful in the differential diagnosis of TIA-like symptoms (Ali et al., 2006), but EEG findings have not systematically been surveyed in adults with moyamoya. One small mixed series (Shoukat et al., 2009) of seven patients under age 12 years and six over age 12 reported EEGs in six patients: four with diffuse slowing and two with epileptiform activity, but the age of the patients receiving EEGs was not specified. Similar observations were made in an Italian series of 27 pediatric and 7 adult cases with moyamoya. EEG showed slowing in 58% and spikes in 33%, but the study did not specify how many of those with epileptiform activity were adults. Accordingly, we reviewed the features associated with clinical seizures and epileptiform EEG activity in 103 adult patients with moyamoya seen at a large moyamoya referral center in the western United States, comprising 90 who underwent revascularization procedures on 128 hemispheres. We do not here address the use of EEG for intraoperative monitoring in patients with moyamoya (Lopez, 2009; Vendrame et al., 2011).

2. Materials and methods

The clinical informatics and EEG databases at Stanford University (including both hospital and outpatient clinic data) were queried for patients with a diagnosis of moyamoya, ICD9 437.5. This diagnostic code captures patients with either moyamoya disease (idiopathic moyamoya) or syndrome (associated with another condition). From this patient list, the Stanford STRIDE clinical informatics database was again queried for clinical documents with "EEG report" in the title or description, as well as procedural codes for routine, extended, ambulatory, and continuous EEG (CPT 95812, 95813, 95816, 95818, 95819, 95822, 95950, 95951, 95953, 95956). STRIDE (Stanford Translational Research Integrated Database Environment) is a research and development project at Stanford University used to create a standards-based informatics platform supporting clinical and translational research (Lowe et al., 2009). EEGs performed on patients who had not yet attained 18 years by the time of the study were excluded, as they were not part of the adult hospital database. Location, gender, race, age, indication for EEG, history of seizures, and history of seizures were obtained in the following fashion. The EEG report, the most recent hospital history and physical examination, hospital discharge summary following the EEG, and most recent outpatient neurology or neurosurgery clinic note prior to the study were reviewed in turn. EEG findings containing any of the following phrases were collected for review: diffuse slowing, focal slowing, focal amplitude decrease, epileptiform activity, electrographic seizure, frequent seizure, periodic epileptiform discharge, pseudo-periodic epileptiform discharge, stimulus-induced rhythmic periodic ictal or interictal discharge, rhythmic delta, evolving rhythmic discharge or breach rhythm. Phrases were used only as search items for clinic notes and EEG reports in identified moya-moya patients, with criteria for meeting the terms of the phrase left up to the creator of the original note. Per institutional protocol, EEG response to hyperventilation is not routinely assessed on adults with suspected cerebrovascular disease, given concern for provoking cerebral ischemia. Therefore, presence of the "re-build up" phenomenon with EEG slow-wave paroxysms after hyperventilation could not be assessed. The Stanford Institutional Review Board approved this retrospective study.

3. Results

A cohort of 540 patients with moyamoya and at least one EEG was identified with methods detailed above. Among all adult patients with a diagnosis of moyamoya (ICD9 437.5), 103 had an EEG outside of the operating room setting. Of these, 74 patients had an inpatient short-term EEG (20–40 min), 36 had an inpatient long-term EEG (greater than 12 h), 8 had an outpatient short-term EEG, and 1 had an ambulatory 24-h EEG. A flow of patient categories and numbers is given in Fig. 1.

3.1. Cohort demographics

Demographics are summarized in Table 1.

Females comprised 68% of the population. Racial background was distributed as 48% White, 34% Asian, 6% African–American, 5% Latino, and 7% unspecified/other. Mean age was 43.7 ± 11.3 years (median 42 years). To determine if the demographics of patients receiving EEGs were similar to those of other adult patients with moyamoya disease, the Stanford STRIDE electronic medical record cohort discovery tool was used to abstract de-identified data from the Stanford Hospital and Clinics. There were 750 unique adult patients with a diagnosis of moyamoya disease, of whom 515 (69%) were female. Ethnic and racial data were only available for 620: 375 (60%) White, 170 (27%) Asian, 45 (7%) Hispanic or Latino, 30 (5%) Black.

3.2. Clinical history of cohort

Prior to obtaining an EEG, 53 of 103 (51%) had a history of at least one clinical seizure. A clinical stroke preceded the EEG in 86 (83%) and an intraparenchymal hemorrhage in an overlapping group of 33 (32%). Eleven patients had neither a stroke nor a hemorrhage. Of patients receiving revascularization, 83 of 103 (92%) had a prior stroke, hemorrhage or clinical seizure. Indications for obtaining the EEGs were obtained from the EEG report or where necessary physician progress notes and are summarized in Table 2.

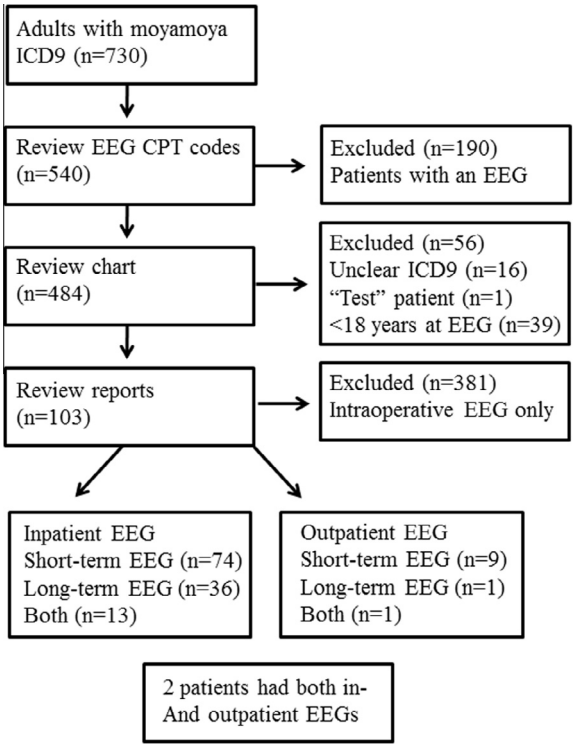

Fig. 1. Patient recruitment diagram. *Abbreviations:* EEG = electroencephalogram; ICD9 = International Classification of Diseases, version 9; CPT = current procedural terminology.