

Non-convulsive status epilepticus presenting with Wernicke's aphasia

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ABSTRACT

تعتبر الحبسة (الخرسة) المنتبذة لدى البالغين ظاهرة نادرة. معظم الحالات الظاهرية التي تم الإبلاغ عنها تعاني من حبسة غير طليقة (Broca). الحبسة الطليقة من نوع (ويرنيك) (Wernicke) هي أقل شيوعاً. نستعرض في هذا التقرير حالة سيدة تبلغ من العمر 47 عاماً حضرت وهي تعاني من عوارض متكررة من نوبات صرعية لاإختلاجية على شكل حبسة من نوع (ويرنيك) (Wernicke) لمدة أسبوعين. أظهرت نتيجة تصوير الدماغ بالرنين المغناطيسي (MRI) وجود احتشاء دماغي قديم في المنطقة القذالية الجدارية اليسرى. أظهرت نتيجة تخطيط الدماغ الكهربائي (EEG) لفروة الرأس وجود موجات حادة دورية ومستمرة في المناطق الصدغية اليسرى مع انتشارها إلى كامل نصف الدماغ الأيسر، وفي بعض المناسبات تكون في الجانب الأيمن. يعقب ذلك فترات متغيرة من تباطؤ النوبة المنتبذة. تمت ملاحظة تكرار نموذج الانتباز الموصوف، كما تمت معالجة الحالة الصرع باستخدام عقار ديازيبام عبر الوريد، عقار فينيتوين وعقار فينوباربيتون. بعد المعالجة، تحسنت حالة المريضة من الناحية السريرية كما تحسنت نتيجة تخطيط الدماغ الكهربائي (EEG) مع غياب الإيقاع الانتبازي في الصدغ الأيسر وعودة تخطيط الدماغ الكهربائي (EEG) إلى الوضع الطبيعي. لهذا، فإن يتم القيام بتشخيص الحالة الصرعية الجزئية الغير اختلاجية (NCSE) كظاهرة حبسه (خرسه) منتبذة.

Ictal aphasia in adults is a rare phenomenon. Most reported cases manifest with non-fluent (Broca) aphasia. Ictal fluent (Wernicke) aphasia is less common. We report a 47-year-old, right-handed woman that presented with recurrent episodes of non-convulsive seizures in the form of Wernicke's aphasia for 2 weeks. An MRI of the brain showed an old cerebral infarction in the left parieto-occipital area. Scalp EEG revealed continuous periodic sharp waves at the left temporal regions with diffusion to the whole left hemisphere and at occasions to the right. This is followed by variable periods of post ictal slowing. Recurrence of the described ictal pattern was noted. Management of status epilepticus was started in the form of intravenous diazepam and a loading dose of phenytoin and phenobarbitone. After treatment, she improved clinically and the EEG improved

with disappearance of the left temporal ictal rhythm and normalization of the EEG background. Thus, establishing the diagnosis of non-convulsive partial status epilepticus manifesting as ictal aphasia.

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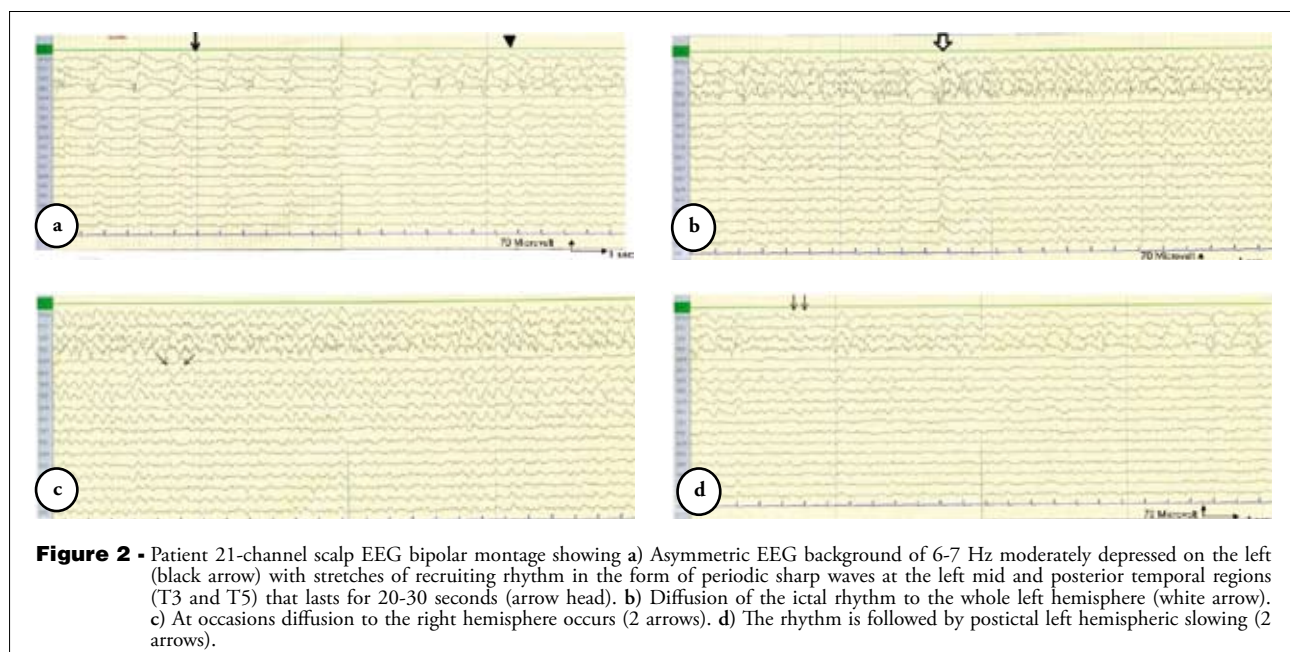
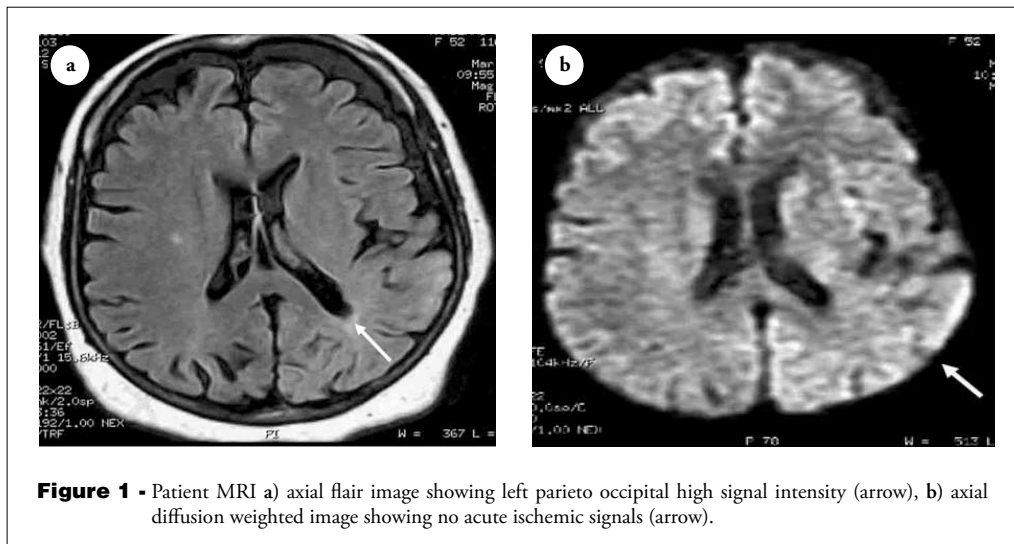
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Aphasia is defined as a breakdown of the 2-way translation that establishes correspondence between thought and language.¹ Broca's aphasia is characterized by intact comprehension language and impaired fluency of speech, naming, repetition, reading, and writing.¹ The lesion in Broca's aphasia (expressive aphasia) is in the speech area in the left inferior posterior frontal gyrus.² Wernicke aphasia is characterized by impaired comprehension with normal speech fluency. Naming, repetition, reading and writing are impaired, and the patients continuously produce jargon speech or word salad. The lesion is usually in the posterior position of the superior temporal gyrus sometimes extending to the inferior parietal lobe.³ Global aphasia is a summation of Broca's and Wernicke aphasia and the lesion is typically large involving both inferior frontal and superior temporal regions and often much of the intervening parietal lobe.⁴ Language disturbances manifesting as brief periods of speech arrest occur with seizures originating in the frontal or the temporal lobes.⁵ However, aphasia as the sole manifestation of non-convulsive status epilepticus (NCSE) is rare, and ictal aphasia is a simple partial seizure that originates from the language areas. This is different from the common speech arrest of temporal lobe seizure or vocalization that results from dominant hemisphere frontal or temporal lobe seizures, as the latter manifest

there is often clouding of consciousness and spread of the seizure activity to involve the ipsilateral and possibly the contralateral hemispheres.⁶ We report a patient with NCSE presenting with Wernicke aphasia and with normal fluency speech but impaired comprehension, naming, repetition, reading and writing. The objective of this case report is to highlight a rare form of partial seizure.

Case Report. We present a 47-year-old, right-handed mother of 10 daughters. She had a medical history of pre-eclampsia, eclamptic fits, and parieto occipital infarction complicating the course of pre-eclampsia with subsequent post stroke epilepsy

manifesting with recurrent generalized tonic clonic seizures in remission on phenytoin 300 mg daily for 23 years. She presented to the emergency room with a 2-week history of episodes of headache, confusion, and talking nonsense, which became continuous for 2 days prior to presentation. Physical examination revealed an afebrile, normotensive lady with normal general and visceral systemic examination. The neurological examination showed a conscious lady with Wernicke aphasia. Her comprehension was impaired with no response to verbal stimuli. She had inappropriate fluent jargon speech (word salad), the ability to repeat, write, or read could not be tested. There were no motor or cerebellar signs and the sensory system could not be



tested. Blood profiles including full blood count, urea and creatinine, liver function tests, thyroid function tests, coagulation profile, serum glucose, serum lipids, autoimmune screen, anticardiolipids, antiphospholipid, proteins, C, S, Factor V, and urine drug screen were within normal limits. An MRI of the brain was carried out (Figures 1a & 1b) and confirmed the presence of an old ischemic left parieto occipital lesion, there were no acute ischemic changes on diffusion images. A CSF analysis was performed and showed normal cell count, normal protein and glucose, and herpes simplex virus polymerase chain reaction (PCR) was negative. A 21-channel scalp EEG for 30 minutes was performed (Figures 2a-2d) and showed asymmetric background of EEG 6-7 Hz moderately depressed on the left with stretches of recruiting ictal rhythm in the form of periodic sharp waves at the left mid and posterior temporal regions (T3 and T5) that lasts for 20-30 seconds with diffusion to the whole left hemisphere and at occasions to the right hemisphere followed by left hemispheric slowing with 4-6 Hz transients at the left temporal regions for 5-10 seconds. This left temporal pattern repeated itself throughout the recorded EEG. The diagnosis of NCSE was made, and management of status epilepticus was started immediately with intravenous diazepam of 10 mg followed by loading dose of intravenous phenytoin at 20 mg per kg at 50 mg/min with no improvement. A loading dose of intravenous phenobarbitone 15 mg/kg was given slowly over 3 hours to avoid any respiratory suppression. Electrically, the NCSE subsided after the phenobarbitone loading dose as the scalp EEG revealed left hemispheric theta transients of 4-7 Hz and eventually normalization of the EEG background with excessive beta activity of 20-25 Hz bilaterally, more in the frontal regions. Clinically, after the phenobarbitone loading dose she became drowsy but could respond to simple verbal commands. Her speech and comprehension gradually improved over 3 days to normal. She was discharged and maintained on phenytoin 300 mg once daily, and phenobarbitone 100 mg once daily with adequate phenytoin and phenobarbitone serum levels.

Discussion. This case demonstrates a NCSE manifesting with Wernicke's aphasia in a patient with old left hemispheric infarction. The association of the Wernicke aphasia, the EEG finding, and the clinical and electrical response to antiepileptic drugs support the diagnosis. Prolonged episodes of aphasia and concurrent dominant hemisphere epileptic activity have been recognized in the past, and are still increasingly recognized in recent literature.⁷ In most cases, there is underlying recently acquired focal pathology with

lateralized neurological findings.^{7,8} The EEG patterns reported in these cases vary from paroxysmal dominant hemisphere spike and sharp activity to periodic lateralized epileptiform discharge (PLEDs) patterns, and the aphasic disorders have been predominantly non-fluent or mixed character.⁹ Our case demonstrates a NCSE presenting with Wernicke aphasia and suggests that focal seizure should be added to the differential diagnosis of acute Wernicke aphasia, or in cases of worsening Wernicke aphasia in stroke patients. It is also important to emphasize that the term ictal aphasia and aphasic seizures are not quite synonymous. Ictal aphasia should refer to seizures in which some degree of aphasia is present such as dominant temporal lobe seizures in which vocalization, speech arrest, or paraphasic errors are only part of ictal semiology, and such seizures are relatively common.¹⁰ By contrast aphasic seizures, namely, seizures where aphasia is the predominant or the only ictal symptom of a partial seizure, as in this case report, are relatively rare as described above.^{8,11}

In conclusion, non-convulsive seizure manifesting as Wernicke aphasia may occur and should be distinguished from acute vascular events as the former is reversible with appropriate antiepileptic drugs.

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