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An Unusual Presenting Symptom of Tranverse Sinus Thrombosis: Gyratory / Rotatory Seizure

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ÖZET

Transves sinüs trombozunun alışılmadık bir başlangıç semptomu: jiratör/rotatuar nöbet

Serebral venöz tromboz (SVT) hayatı tehdit edici bir hastalıktır. Fokal ya da jeneralize nöbetler hastaların %12-15'inde SVT'nin ilk semptomu olarak görülür. Daha önce sağlıklı olan 36 yaşındaki bir kadın kendi ekseni etrafında dönme şeklinde tanımlanan öyküsü ile başvurdu. Acil serviste yapılan muayenesinde hasta tamamen alert durumdaydı ve herhangi bir nörolojik defisit saptanmadı. Manyetik rezonans görüntülemesinde (MRG) sol transvers sinüste tromboz ve sol inferior temporal girusta venöz enfarktı saptandı. Olgunun, nöbet semiyolojisi ve bu nöbet tipinin SVT'nin ilk ve tek semptomu olarak görülmesi nedeniyle sunulmaya değer atipik bir olgu olduğunu düşünmekteyiz.

Anahtar kelimeler: Sinüs trombozu, intrakraniyal, jiratör nöbetler

ABSTRACT

An unusual presenting symptom of tranverse sinus thrombosis: gyratory/ rotatory seizure

Cerebral venous thrombosis (CVT) is a potentially life-threatening disease. Focal or generalized seizures are seen in 12-15% of the patients as the first presenting symptom of CVT.A previously healthy -36-year-old woman admitted with a history which was described as 'she was rotating around her axis'. On examination, in the emergency service of our hospital the patient was fully alert and any neurologic abnormality was not detected. Her magnetic resonance-imaging (MRI) revealed a thrombosis in the left transverse sinus and a venous infarct in the left inferior temporal gyrus. We conclude that the case is atypical in terms of seizure semiology and for the reason that this type of seizure occurred as the first and the only symptom of CVT.

Key words: Sinus thrombosis, intracranial, gyratory seizures

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INTRODUCTION

Cerebral venous thrombosis (CVT) is a disease that occurs secondary to the occlusion of the dural venous sinuses, superficial cortical veins or deep veins of the brain. In about 30% of the patients the etiology is unknown. The most frequent clinical symptoms are headache, blurred vision, focal deficits and/or altered level of consciousness. Focal or generalized seizures are seen in 12-15% of the patients as the first presenting symptom of CVT (1). Here, we report a patient diagnosed as CVT presenting with unusual seizure semiology.

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CASE REPORT

A 36-year-old female patient was brought to the emergency room with complaints of 'confusion' and involuntary whirl around movement of her body. According to the patient's recollection and the testimony of her husband, while she was in a standing position in the bank, her mind suddenly became blurred, she had to discontinue the procedure, and then a nonsensical smile appeared on her face. She then experienced involuntary eye deviation to the left and she began whirling around leftwards; making three complete 360 degree turns around her axis. At that time her husband

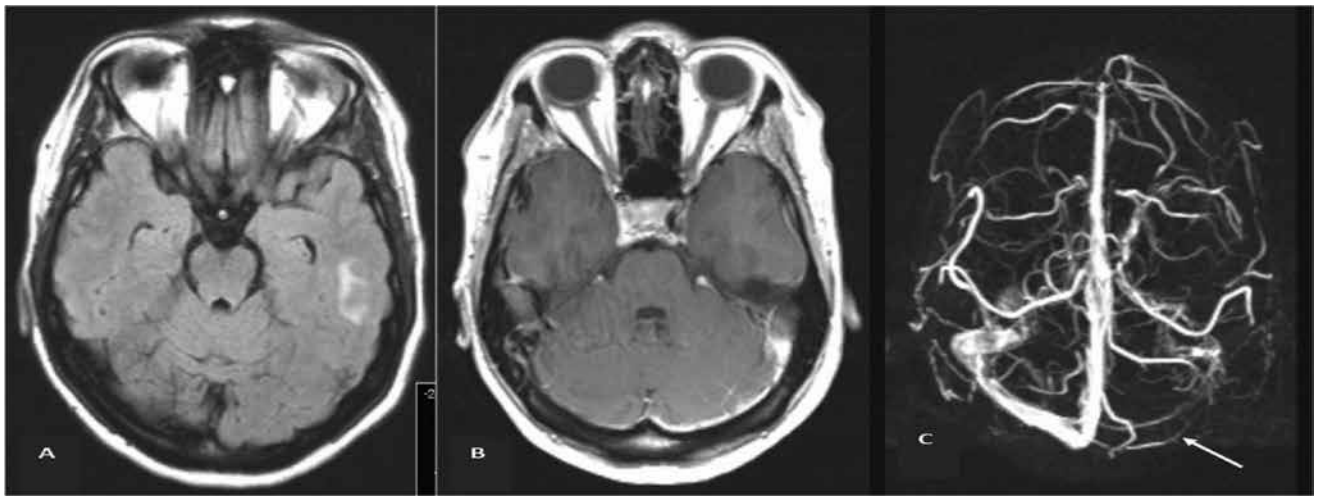

Figure 1: MRI of the case. Venous infarction was detected in the left inferior temporal gyrus in axial FLAIR T2 images (A). A left transverse sinus thrombosis in post-contrast axial T1 image (B) and the thrombosis was also shown in MR-venography (C).

had watched his wife without touching her. After the turns were completely over, the patient lost consciousness for about 10 minutes. There was no evidence of convulsion, incontinence or tongue biting. Her husband reported her to be confused and disoriented for about 30 minutes. The patient stated that she did not have any seizure previously. Past history was positive for using oral contraceptive drugs for three years and also for smoking 15 packs per year. Family history was noncontributory.

Vital signs were normal. There was no neck stiffness and her ocular fundus appeared normal. She was alert and became totally oriented soon after she was admitted to the hospital. Her speech was normal. The complete neurological examination was normal. Her basic laboratory findings and computerized tomography (CT) scan of the brain were normal. An interictal EEG was normal. The long term EEG for three hours and also video monitoring EEG (for three days) were normal. Her brain MRI revealed a thrombosis in the left transverse sinus and a venous infarction in the left inferior temporal gyrus of the left hemisphere (Fig 1 A,B) and MR venography confirmed the diagnosis of thrombosis (Fig 1 C). Her tests for vasculitic markers (ANA, Anti ds DNA, ANCA, antiphospholipid antidody tests) and hypercoagulability (factor V Leiden mutation, prothrombin 20210 G→A mutation, deficiencies of antithrombin III, protein C, protein S) were within normal limits.

The patient was prescribed anticoagulation therapy (heparin infusion 1000 U/h) and valproic acid (slow releasing 500 mg/day). She experienced no further seizures. Then, the anticoagulation therapy was initiated orally. The MRI scan, which was performed three months later, showed normal findings with recanalized sinus. Her oral anticoagulation therapy was discontinued after six months, while antiepileptic therapy was continued for two years. The patient did not experience any other seizure with the monotherapy.

DISCUSSION

Cerebral venous thrombosis (CVT) is a medical condition encompassing thrombosis of cortical and deep cerebral veins and the dural sinuses. The exact cause is unknown. The clinical presentation varies widely; according to the site and extent of thrombosis (1,2). Causes include hematologic disorders, hypercoagulable states, pregnancy and contraceptive medications. CVT may produce elevated intracranial pressure, headache, focal neurological deficits or seizures. Seizures are seen in 40% of all patients in any stage of the disease after admission. However, focal or generalized seizures are seen in 12-15% of the patients as the first presenting symptom of CVT (1). Recurrent seizures are reported to have occurred in less than 10% of patients and in those who experience seizures at the acute stage (2).

The seizure of our case is called complex partial seizure according to clinical classification and simple

motor seizure - gyratory (synonymously rotatory, circling and volvular) seizure according to semiology (3). Patients that have gyratory/rotatory seizures rotate around the axis of their bodies during a seizure at least 180 degrees for once or more times; just as our patient did (4.5). The complex circling behavior occurs in a "ballet-like" fashion and is usually observed at the beginning of the episode when the patient is in a standing position (4). Gyratory seizures can be mistaken for unidirectional turning of the whole body which is defined as the rotation of the trunk (with head and limbs) in one direction by more than 90 degrees, lasting for 10 seconds. Moreover, it is reported that unidirectional turning of the whole body is a new lateralizing sign in the temporal lobe complex partial seizures with a good predictive value for epileptogenic focus contralateral to the direction of turning (6). It may be the sole expression of the seizure or preceded or followed by other epileptic manifestations. The direction of the rotation usually remains constant from one attack to another in the same patient. Consciousness may be preserved or may be lost during the episode (4). It is concluded that the direction of the rotation is not of importance for lateralization (7). On the other hand, it has been reported that in gyratory seizures without a preceding gyratory forced head version, the direction of rotation is towards the side of seizure onset (8).

The patients mostly had neoplasm, abscess or gliotic lesions in cortical or in thalamic area (4,7). Yet, to our knowledge, none of them had an etiology of CVT. Gastaut et al. defined two types of gyratory seizures (9). The common form which was seen during childhood or adolescence and associated with generalized seizures, shows bilateral, rhythmic, symmetrical, or asymmetrical 3-Hz EEG slow-wave discharges. This form is related to a family predisposition, normal cerebral structure, and good response to treatment and thought to be a benign variant of primary generalized epilepsy. The other form, occurring in infants or adults and associated with focal EEG and demonstrable pathology, is a relatively rare condition (9). Recently, some authors divide the seizures into two groups; juvenile onset (< 20 age) and adult onset. The juvenile onset group represents a manifestation of benign generalized epilepsy, while the latter group represents focal pathologies in the basal ganglia or thalamus (7).

It has been suggested that gyratory seizures are associated either with a manifestation of frontotemporal automatism or with a direct effect of -asymmetric activation of related part of the- extrapyramidal system connected with the substantia nigra to which cortical discharges spread (10,11). Vercueil et al. reported that gyratory seizure onset is possibly from the temporal lobes followed by a spread to the basal ganglia or deeper structures (5). Saka et al. also suggested that cortico-basal ganglionic-thalamo-cortical circuit is related with rotating movements (12). Also, there are some reports of the gyratory seizures originating from a frontal lobe, occipital lobe or in contrast, a functional imbalance in generalized cerebral excitability (11,13). The latter is supported by rat studies suggesting an intrinsic asymmetry of the nigrostriatal dopamine system (13). It has been reported that circling movements could be observed with unilateral dopamine injection into the striatum in rodents, and in cats by stimulating caudate nucleus, various thalamic nuclei, or the subthalamic region and the pulvinar-lateralis posterior nucleus complex (14). It has also been suggested that vestibular cortex could be responsible for such a complex motor activity, at least during focal seizures (5).

Our patient had venous infarction in the inferior temporal gyrus due to sinus thrombosis. The origin of the epileptic discharges in our case may have been in the left temporal lobe, and the discharge may have spread to amygdala causing the nonsensical smile and confusion, and then to the nigrostriatal system causing the gyratory seizure. Our case did not fall down or experience a generalized tonic-clonic seizure. Unfortunately, her EEG in the emergency room was not able to be done. We could not demonstrate any pathology either on routine scalp EEG or on the longterm EEG. Furthermore the video-EEG study was normal. Although grid implantation is one of the best tools available for localization of epileptogenic foci in the brain, invasive EEG monitoring was not made because of complications related to grid electrodes.

Our case is atypical and rare for the seizure semiology, seizure type and symptomatology of venous thrombosis. To our knowledge there have been no publications on this subject. Moreover, the patient has a potentially alarming disease which is highly suspicious and presents a symptom which can easily be misdiagnosed.

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