A case with hyperkinetic frontal lobe epilepsy presenting as a psychiatric disturbance

Humen Elmi[1], Ayşe Kılınçaslan[1], Mücahit Öztürk[2], Zuhal Yapıcı[3]

[1] Department of Child and Adolescent Psychiatry, and [3]Division of Child Neurology, Department of Pediatrics, İstanbul University İstanbul Faculty of Medicine, and [2]Department of Child and Adolescent Psychiatry, Yeditepe University Faculty of Medicine, İstanbul, Turkey

SUMMARY: Elmi H, Kılınçaslan A, Öztürk M, Yapıcı Z. A case with hyperkinetic frontal lobe epilepsy presenting as a psychiatric disturbance. Turk J Pediatr 2011; 53: 574-578.

We report a case emphasizing the association of epilepsy with psychopathology. The patient was first referred for impulsive and inattentive behavior at the age of 4, and was diagnosed with attention deficit hyperactive disorder. At the age of 7, intermittent daytime episodes characterized with sudden fear and purposeless running, jumping, clapping, and rocking started. Electroencephalography (EEG) recordings did not reveal any abnormality. Two years later, night-time episodes started, which were quite similar to the daytime spells. An overnight EEG showed bilateral frontal spike and wave discharges predominating on the right. Magnetic resonance imaging revealed gliotic changes in the anterior frontal areas, and neuropsychologic assessment findings were compatible with frontal lobe damage. Treatment with carbamazepine not only controlled hyperkinetic motor seizures but also helped interictal inattentiveness and behavioral impulsivity. In our case, ictal behavioral changes were masked by interictal behavioral disturbances and the normal findings in some of EEG records delayed the diagnosis.

Key words: frontal lobe epilepsy, attention deficit hyperactivity disorder, frontal lobe damage.

Although it has been more than 60 years since the first cases with frontal lobe epilepsy (FLE) have been reported, it is still one of the most complicated and least understood forms of epilepsy[1]. Frontal lobe seizures spread widely and rapidly so interictal discharges often appear bilaterally, which complicates accurate localization of the epileptiform foci[2,3]. The clinical spectrum of ictal and interictal behavioral phenomenology of the patients can be diverse accordingly. The electroencephalography (EEG) can be normal both in the ictal or interictal states[4] and the neuroimaging studies are often unrevealing[5].

The behavioral manifestations of frontal lobe seizures can be quite bizarre and are often mistaken for sleep disorders, pseudoseizures or other psychiatric disorders[1,6,7]. Clinical features include explosive onset, unusual movements (axial-pelvic torsion, bimanual and bipedal movements such as kicking and bicycling), bizarre posture and vocalizations (like screaming, swearing), and rapid postictal recovery. The behavior may seem purposeful, and in some cases, ictal or possibly postictal automatisms with aggressive behavior may be seen.

Here, we present a case of a 14-year-old girl, who had been followed with attention deficit hyperactivity disorder (ADHD) for years and was eventually diagnosed as FLE. Her presentation, differential diagnosis and two-year follow-up are discussed.

Case Report

The patient was the product of an unplanned pregnancy of a 16-year-old mother, and was born at the 38th week by an uncomplicated spontaneous delivery. Her birthweight was 3200 g and length was 52 cm. Her early motor development was unremarkable, and she spoke her first words at 12 months and

phrasal speech at 18 months. She was reported to have a period of 10 days with unremitting fever, and was hospitalized for 7 days when she was 18 months old. No extra-cranial focus for the fever could be detected, and lumbar puncture (LP) was planned. Because her fever started to decline on the day of LP with intravenous antibiotics, the doctors desisted from performing the procedure.

At 4 years old, she was referred to İstanbul Faculty of Medicine Child and Adolescent Psychiatry Clinic because of inattention and speech problems. She was reported to be very inattentive, could not focus on what she was doing, was unable to listen as long as her peers, and was distracted very easily. She spoke in short sentences and had difficulty in finding the right words, which led to many mistakes. She answered questions but could not converse to-and-fro on introduced topics. She scored 84 on the Stanford Binet IQ Test and was described as a "left-handed, unstable, meaninglessly smiling girl with tangential speech" by the tester. She was diagnosed as probable ADHD and was advised to attend a nursery school.

At the nursery school, she was described as a puerile girl who wanted to play with other children on her own terms and cried a lot upon frustrations. She always wanted to boss other children and was rejected by them. She could stay on tasks for very short periods and always left them uncompleted. Her difficulties with receptive and expressive language continued. She was involved in a traffic accident with her family at the age of 6. Her parents reported no head trauma or symptoms associated with head trauma, but they noted some cuts on her lower extremities.

The patient had several problems adjusting to elementary school. At first, she did not write in the classroom, but could do her homework under the continuous supervision of her mother. She started writing only after several private lessons with her teacher. She could not listen to the teacher, looked around and blurted out answers or started to talk about irrelevant topics during the lesson. She was started on methylphenidate but could not use it because of skin eruptions after the first dose. She learned to write and read by the end of the year.

By the end of age 7, she started having daytime episodes. In the first one, she was reported to "become anxious" upon seeing two men fighting each other on television. She stood abruptly and started walking around, with vocalizations like "Aaa" or "Ay ay ay". It lasted about 45 seconds and ended with sleepiness, of short duration. During the first two years, the episodes all occurred at home, but the frequency differed from once in a month to several in a week. At the age of 9, episodes began to occur outside the house, like at school or a shopping center, and the frequency increased. Some of the episodes were provoked by an emotional or a physical change, like someone starting to speak loudly or after feeling that her father would be angry with her. There was no apparent provocation on many occasions. The episodes were quite similar to each other. She stood up suddenly, breathed deeply, and started running, jumping, and hitting her fists together, or sometimes remained sitting and rocked back and forth with vocalizations. She displayed intense feelings like fear, surprise and excitement, her face became pale, her lips turned purple and she yawned as if sleepy. She felt an urge to urinate and the episodes always ended in the bathroom. They were between 30-60 seconds but sometimes occurred in clusters and lasted about 30 minutes. She could remember the feelings and urination but could not remember other behaviors. She said she could hear others but could not answer or stop what she was doing. Her neurologic examination and two different daytime EEG recordings revealed no abnormalities. She was treated with sertraline and valproic acid but with very little success. Her problems with inattention, impulsivity and difficulties with her friends persisted.

By the age of 12, nocturnal episodes began. They were generally soon after falling sleep, mostly in 5-10 minutes and very similar in nature to the daytime spells. They could occur many times in a night and cause sleeplessness.

She was referred to the Pediatric Neurology Section where she underwent overnight sleep and wake EEG and magnetic resonance imaging (MRI) scans. EEG revealed no abnormality when awake but showed bilateral frontal spike and wave discharges predominating

on the right hemisphere in sleep. The MRI showed gliotic changes in the anterior frontal areas, which were larger on the left (Fig. 1). Results on the Wechsler Intelligence Scale for Children-Revised were as follows: verbal IQ: 78, performance IQ: 77 and total IQ: 78. Neuropsychologic assessment revealed problems in attention, inhibition and executive functions, with the preservation of memory and visuospatial skills.

The diagnosis of hyperkinetic FLE was given and she was started on carbamazepine 400 mg daily, and the dose was increased to 800 mg in two weeks. She first recovered from nocturnal episodes, and then the number of daytime seizures decreased to once or twice a week. She now has no hyperkinetic episodes; she only stands up and breaths deeply in her seizures. The frequency of spells dropped further with an increase of her drug to 1000 mg daily. Her mother describes behavioral improvement apart from seizure control after medication over two years.Although she still has some difficulties in sustaining attention and getting on well with friends, she can organize herself with respect to homework, ask questions at proper times, spend longer periods with her friends, and buy 4-5 items without forgetting.

Discussion

The presented case illustrates the complexity of the border between neurology and psychiatry. Early onset of problems with attention, impulsivity, interaction with peers, and relatively normal onset of speech led to the diagnosis of ADHD. The relation between ADHD and frontal lobe damage has long been highlighted[8,9], and localization of the brain lesion in our case might have contributed to both the behavioral problems and the neuropsychological findings. However, it is very difficult to detect the etiology and timing of the lesion. The unremitting fever at the age of 18 months is unlikely to be related with bilateral focal frontal lesions. Similarly, the traffic accident at the age of 6 is unlikely to have caused the symptoms because no head trauma or behavioral change was described after the accident, and the behavioral problems had started earlier.

Several reports have also found poorer performance on tests of attention, resistance to interference, response inhibition, and executive functions in subjects with FLE[1,10,11]. Thus, in our patient, it is reasonable to think that the addition of the seizures to the picture might have increased the problems. The beneficial effects of carbamazepine on seizure control and impulsivity might have helped to achieve the behavioral improvement reported in our case.

The addition of the daytime episodes increased the confusion about the diagnosis, and at the same time changed the route to a more neurologic origin. As described in many previous reports on FLE, EEG recordings revealed no epileptic focus, and the vague characteristics of the seizures in our case (stressors described before episodes, semi-purposeful behaviors, and abrupt termination with prompt return to responsiveness) delayed the diagnosis. Hyperkinetic daytime episodes

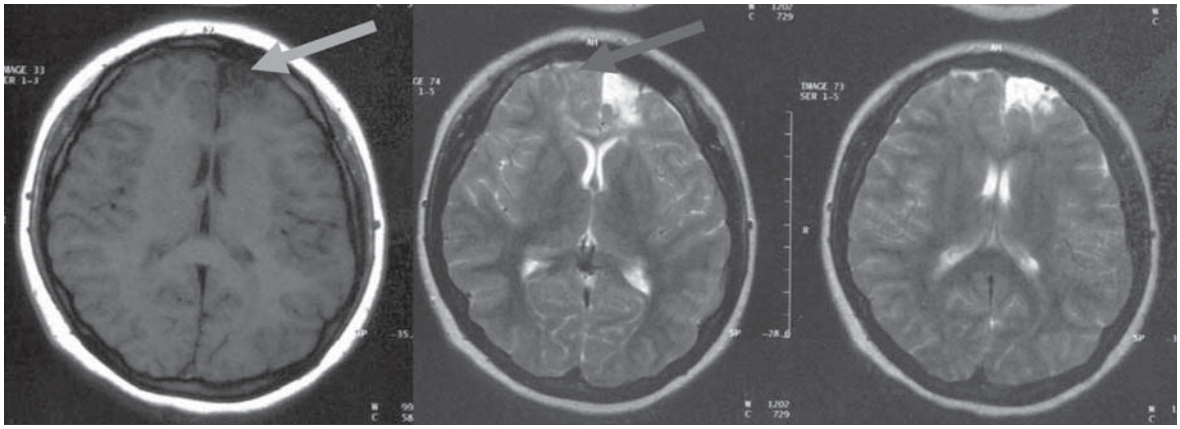

Fig. 1. Bilateral gliotic changes in the anterior frontal areas, larger on the left.

were commonly likened to ADHD symptoms, as in our case, in similar cases described in the literature[12].

Although FLE is predominantly characterized with nocturnal episodes, they emerged five years after the daytime spells in our patient. FLE can be mistaken for sleep terror because both can present at night with autonomic hyperactivity, crying and intense emotions like fear. A recent study on children[1] described the below features to suggest more commonly a frontal seizure rather than a sleep disturbance: 1) occurrence early in sleep, rather than in deep non-REM sleep as observed in sleep terror, 2) brief events of 1-2 minutes, as opposed to 20-30 minutes in sleep disturbances, 3) stereotypic events with little variability, which included prominent kicking and at times incontinence, 4) several events in a single night; sleep terror mostly occurs once in a night, 5) occasional occurrence of the events during the day, while awake or during nap, and 6) the patients often fall outside the usual age range for sleep terror, which is 2-10 years.

Motor manifestations are common in FLE and occur in 90% of the cases[13]. Seizures from the supplementary sensorimotor area consist of sudden and explosive tonic posturing of limbs often with contraversion of the eyes and head, vocalization or speech arrest[14,15]. Other hypermotor seizures may appear with extreme motor restlessness, complex motor automatisms and agitation, intensely affective vocal and facial expression associated with powerful bimanual-bipedal and axial activity, and repetitive rhythmical and postural movements. Seemingly purposeful aggressive activity and speech during seizure and sustained violent behavior in the interictal period may be misdiagnosed as conduct disorder. Less aggressive, hyperkinetic presentation and, at times, lack of alteration of consciousness may present as ADHD. Odd motor activity and abrupt termination with prompt return to responsiveness can be considered as a pseudoseizure, whereas bizarre movements and prolonged confusion may seem like a psychotic disorder.

There may be an aura comprised of psycho-sensorial, affective, cognitive, and neurovegetative symptoms. Following abrupt cessation of normal behavior, looking in surprise with eyes wide open (staring reaction) is frequent. Sudden emotional outbursts consisting of intense fright and facial expressions of fear associated with shouts and aggressive verbalizations are frequently reported. Autonomic phenomena such as pallor, tachycardia, mydriasis, forced urination, and apnea are common. Seizures are characteristically stereotypical and of brief durations. Several seizures may come one after another in clusters, and the risk of status epilepticus is high.

In conclusion, FLE is poorly understood and often unrecognized by healthcare providers working with children. The condition is often missed or misdiagnosed as a psychiatric problem/pseudoseizure or a sleep disorder. EEG recordings and neuroimaging are usually normal. The vivid psychiatric symptomatology seen in these cases highlights the necessity to consider multiple etiologies and evaluate them in a multidisciplinary team.

REFERENCES

1. Sinclair DB, Wheatley M, Snyder T. Frontal lobe epilepsy in childhood. Pediatr Neurol 2004; 30: 169-176.
2. Rasmussen T. Characteristics of a pure culture of frontal lobe epilepsy. Epilepsia 1983; 24: 482–493.
3. Shulman MB. The frontal lobes, epilepsy, and behavior. Epilepsy Behav 2000; 1: 384–395.
4. Williamson PD, Spencer DD, Spencer SS, Novelly RA, Mattson RH. Complex partial seizures of frontal lobe origin. Ann Neurol 1985; 18: 497–504.
5. Swartz BE, Halgren E, Delgado-Escueta A, et al. Neuroimaging in patients with seizures of probable frontal lobe origin. Epilepsia 1989; 30: 547–558.
6. Saygi S, Katz A, Marks DA, Spencer SS. Frontal lobe partial seizures and psychogenic seizures: comparison of clinical and ictal characteristics. Neurology 1992; 42: 1274 –1277.
7. Mukaddes NM, Bilge S, Polvan Ö. Psychiatric symptomatology in frontal lobe epilepsy: case report. Bull Clin Psychopharmacol 1999; 9: 222-226. (Turkish)
8. Mattes JA. The role of frontal lobe dysfunction in childhood hyperkinesis. Compr Psychiatry 1980; 21: 358-369.
9. Faraone SV, Biederman J. Neurobiology of attention-deficit hyperactivity disorder. Biol Psychiatry 1998; 44: 951–958.
10. Auclair L, Jambaqué I, Dulac O, LaBerge D, Siéroff E. Deficit of preparatory attention in children with frontal lobe epilepsy. Neuropsychologia 2005; 43: 1701–1712.

11. Shulman MD. The frontal lobes, epilepsy, and behavior. Epilepsy Behav 2000; 1: 384–395.
12. Powell AL, Yudd A, Zee P, Mandelbaum DE. Attention deficit hyperactivity disorder associated with orbitofrontal epilepsy in a father and a son. Neuropsychiatry Neuropsychol Behav Neurol 1997; 10: 151-154.
13. Commission on Classification and Terminology of the International League Against Epilepsy. Proposal for revised classification of epilepsies and epileptic syndromes. Epilepsia 1989; 30: 389-399.
14. Laich E, Kuzniecky R, Mountz J, et al. Supplementary sensorimotor area epilepsy. Seizure localization, cortical propagation and subcortical activation pathways using ictal SPECT. Brain 1997; 120: 855-864.
15. Weinstock A, Giglio P, Kerr SL, Duffner PK, Cohen ME. Hyperkinetic seizures in children. J Child Neurol 2003; 18: 517-524.