Early Onset Schizophrenia: A Case Study

Cenk Erken Başlangıçlı Şizofreni: Bir Olgu Sunumu

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ABSTRACT

Early onset schizophrenia (defined as an onset of psychosis before 13 years of age) is a rare and severe form of the disorder which is clinically and neurobiologically concomitant with the adult-onset disorder. It is rarely reported under 6 years of age in the literature. Here we present a 5.5-year-old girl who developed psychosis and discuss the clinical and familial features, diagnostic and treatment processes and one year follow-up of this particular case. (Archives of Neuropsychiatry 2006; 45: 142-8)

Key words: Very early onset psychosis, clinical and familial features, diagnosis, treatment

ÖZET

Çok erken başlangıçlı şizofreni (nsikotik bozukluğun 13 yaşından önce ortaya çıkması olarak tanımlanmaktadır) oldukça nadir görülen ve klinik ve neurobiyolojik olarak erişkin başlangıçlı bozuklukla aynı zamanda oluşan bir olaydır. 5 yaş öncesinde çok nadir görülmektedir. Bu yüzden psikotik bozukluk gelişen ve çok erken başlangıçlı şizofreni tanı koyan 5.5 yaşındaki bir kız hastanın klinik özellikleri, tanı ve tedavi süreci ve bir yıllık takibi sunulmaktadır. (Nöropsikyatri Araştırmaları 2006; 45: 142-8)

Anahtar kelimeler: Çok erken başlangıçlı şizofreni, tanı, klinik özellikler, tedavi

Introduction

Early-onset schizophrenia (EOS) is a term used to indicate the onset of psychotic symptoms before the age of 18 years. Onset of the disorder rarely occurs before the age of 13 years. If the onset is earlier than 13 years, it is termed very early onset schizophrenia (VEOS) (1,2).

Genetic factors are believed to play an important role in the pathogenesis of schizophrenia. An increased family history of schizophrenia and schizophrenia spectrum disorders (e.g., schizotypal or paranoid personality disorders) have been found in patients with VEOS (3-5). Communication deficits are also often found in the families of children with VEOS (6).

The great majority of patients with VEOS have significant premorbid abnormalities in language, motor, and social development (3,4). Hallucinations, thought disorder, disorganized behavior and flattened affect all have been consistently found in EOS, while systematic delusions and catatonic symptoms may be less frequent (3). Developmental differences in language and cognition may affect the range and quality of symptom presentation (1). Symptom presentation in EOS or VEOS generally has prodromal, acute, recovery and residual phases.

The diagnosis of EOS or VEOS in children and adolescents is made using the same DSM-IV criteria as in adults (7). However, to make the diagnosis of VEOS, a comprehensive psychiatric and physical assessment is needed. General medical causes of psychotic symptoms should also be ruled out, and diagnosis should be confirmed in a longitudinal follow-up.

The treatment of VEOS requires therapies that are both psychopharmacological treatment, targeting the characteristic symptomatology (positive and negative) constituting the disorder, and the psychosocial and educational and social management of the child and family.

Case Presentation

Clinical Presentation:

A is a 5.5-year-old girl with a younger brother who presented with her biological mother. The main concerns of
the mother was her bizarre behavior and speech, not eating or dressing herself, loss of interest and social withdrawal. At the first interview, she did not respond to any attempt at communication and there was little random eye contact, with empty and meaningless looks. She did not have any meaningful words. Her affect was markedly flattened. She seemed to be unrelated and ungroomed. She was talking to herself silently and had smiling or other facial expressions sometimes as she was speaking with someone. She also had some hand movements or gestures as she was explaining or doing or throwing something.

History of Present Illness
The mother reported that she had shown strange behavior and speech during past three months. The first features to be recognized were talking to herself, alternating crying and laughing without any reason, having trouble in relations with her brother and an increased level of hyperactivity. This period lasted for one month. Then she started to say “there is a grave”, “my friend died, somebody killed her/him with a knife”. Following this stage, she started to lick her palms and swear frequently in an unusual way without any reason. The father hit the girl sometimes due to her excessive talking, hyperactivity and odd behavior during this time. Towards the end of the second month, she started to say things like “calling to a cat “ come pisi pisi and eat grandmother”, “there are bugs coming from your mouth” and “somebody ate my pudding in the kitchen”. She waved her hands in the air as if she was picking or throwing something. She was not sleeping and kept talking to herself until late hours. She was aggressive towards her toys and brother.

During the previous ten days she started not talking to anybody or responding to any comments. She became extremely withdrawn. She was not eating or dressing herself. She lost interest in her toys. She continued to be aggressive towards her brother. However, at this time she was only attacking his face when he approached to her. She kept talking to herself, but this time silently and only her lips moved.

Developmental History
The mother reported that the patient was born following a full-term pregnancy with normal delivery, normal weight and without any complications. She denied alcohol or substance use during pregnancy. Although the mother had no significant medical problems, she had had serious psychological stresses during the pregnancy.

The patient’s expressive language development was significantly late. She spoke her first meaningful words at two years of age and sentences at four years. Her early motor development was mildly delayed and her mother also reported some degree of clumsiness and hyperactivity in premorbidly. She was toilet trained at age four. She was responsive socially to her mother during and after infancy. She had some desire to communicate and play with her peers, although she had some difficulty in maintaining relationships. During premorbidly she was able to say everything she wanted to. She helped her mother at home. She also had imaginative and meaningful play with her brother. Her mother and other relatives described her as a talkative and overactive girl in her premorbid period.

Medical History
There was no significant medical history, no history of head trauma, seizure disorders or other neurological disorders.

Familial History
There was a third degree consanguinity between parents (parents are cousins) and a family history of schizophrenia (paternal aunt). The mother had a significant delay in language development and was not able to finish primary school. We also observed that she had difficulty in receptive and expressive language. There had been marital conflict between the parents throughout the marriage. The father has no regular job. The younger brother also has a mild delay in language and motor development.

Follow up
After a provisional diagnosis of VEOS, we ordered a medical and neurological work-up. The results showed a normal cranial MRI, normal sleep EEG, negative screening for metabolic disorders and normal blood tests (except a mild iron deficiency anemia). Routine pediatric and neurological examinations revealed no significant abnormalities.

We started risperidone treatment at 0.5 mg/day at bed time and referred her to a psychoeducational program. After one week, her sleep and hyperactivity improved significantly, but positive (hallucinations, bizarre behaviors) and negative (alogia, withdrawal, blunt affect, not eating or dressing herself) symptoms and aggressive behavior continued. We increased risperidone to 1 mg/day in the second week. At the next visit, two weeks later, significant changes were reported and observed. She began to say a few words, respond to stimuli, and made some level of eye contact for the first time. She seemed related with her environment, and her mother reported that she had started eating and dressing herself. Her aggressive behavior had also decreased. However, talking to herself, crying and laughing, hand movements, communication deficits and blunted affect continued. We increased risperidone to 1.5 mg/day in the eighth week. Although she developed nocturnal enuresis as a side effect, at the end of the third month her positive and negative symptoms and social communication significantly improved compared to the first presentation. She started to talk using short sentences according to her needs, be responsive to stimuli and make eye contact. We could not evaluate her thought content or process in terms of delusions as it was not possible to talk with her until the fourth month, either during clinical interviews or play therapy sessions. We increased risperidone to 2 mg/day as she tolerated medication well generally. In addition to previous improvements, her bizarre behavior (talking to herself, hand movements, facial expressions, crying and laughing) and negative symptoms (blunted affect, withdrawal, alogia) improved significantly by the fifth month of follow-up. She was also doing better in the special education program. We considered that she had a significant overall improvement, compared to the first clinical presentation, in the clinical global impression scale. The mother, grandmother and maternal aunt all stated that the child had improved more than fifty percent in comparison to
the premorbid condition. We gradually decreased risperidone to 1 mg/day. At the end of the one year period of treatment and follow up, she did not develop any other overt psychotic episodes, but deficits in social communication, cognitive functioning and play activities, reciprocal language, flattened affect and some temperament changes, compared to the premorbid condition, continued at some level.

**Discussion**

VEOS is a very rare childhood psychiatric disorder. Systematic and large sample studies on the diagnosis and treatment of this disorder in the literature are scarce. Any case under six years of age should be carefully questioned, and diagnosis should be confirmed in a longitudinal follow up. It is evident that the first issue with this case is the diagnosis. Differential diagnosis of VEOS should include; organic disorders (such as delirium, seizure disorders, central nervous system lesions, metabolic disorders, substance abuse and toxic encephalopathies etc), mood disorders with psychotic features, pervasive developmental disorders (PDD), sexual or physical traumas, nonpsychotic behavioral or emotional disorders (including dissociative disorder) (2).

We initially excluded any possible organic disorders with appropriate investigations and close monitoring during follow up. There was no history of substance abuse or intoxication. Given the possibility that severe physical or psychological trauma in this age group may trigger or mimic many psychiatric disorders, we paid special attention to any history of any kind of trauma. Although a dysfunctional family environment existed, her parents did not report any sexual abuse and we did not observe any signs suggestive of sexual abuse during clinical interviews or play therapy sessions. However, there was a history of physical abuse and neglect, as the father sometimes physically punished the child before and during the illness. However, environmental factors such as socioeconomic status and unusual psychological trauma do not appear to account clinically for the earlier age of onset in VEOS (2,5). Rather, environmental factors may potentially interact with biologic risk factors to mediate the timing of onset, course and severity of the disorder. Psychosocial stressors, including expressed emotion within the family setting, influence the onset and/or exacerbation of acute episodes, and relapse rates (APA, 1997).

A relatively acute onset of loss in social and language skills may prompt one to think about childhood disintegrative disorder (CDD). However there was no loss of motor skills or bladder and bowel control in favour of CDD. She only developed nocturnal enuresis as a side effect after we increased risperidone to 1.5 mg/day. The presence of overt hallucinations and delusional thinking are not expected in CDD (2,8). In addition, the onset and course of the symptoms (including hallucinations and possible delusional thinking), significant improvement with risperidone treatment and the family history of schizophrenia all suggest VEOS rather than CDD. The presence of delay in premorbid language development and subtle social difficulties may also suggest other autism spectrum disorders (ASD), but her premorbid language and social skills were sufficient to communicate easily. Furthermore, there had been no stereotypic behavior typical of ASD (we interpreted hand movements as part of psychotic symptoms rather than stereotypes).

On the other hand, the presence of excessive self-talking, increased level of hyperactivity, laughing or crying and not sleeping until late hours may all suggest a mood disorder with psychotic features. Furthermore, one may consider the subsequent transient picture of withdrawal and loss of contact and interest as a depressive episode of a bipolar disorder. So it may be difficult to differentiate both disorders in a crosssectional and symptomatic evaluation. However premorbid and family histories, onset and course of the symptoms are very helpful in the differential diagnosis and favour VEOS. McClellan and McCurry (1998) (4) found that social withdrawal and aberrant peer relationships, characteristics that equate to negative symptoms, differentiated the premorbid histories of a youth with EOS from those with bipolar disorder.

VEOS generally has an insidious onset (9), and most patients have a prodromal phase which may vary from an acute change (days to weeks) to chronic impairment (months to years) (10). Systematic delusions and catatonic symptoms may be less frequent in VEOS (3). In our case, VEOS seems to have an acute onset with a short prodromal period which included self talking, alternating crying and laughing, difficulties in relations with her brother and an increased level of hyperactivity for one month. This period was followed by overt hallucinations and bizarre behavior. Although she did not exhibit overt delusional thinking, her statement of “somebody ate my pudding in the kitchen” and “somebody killed my friend” could have been signs of delusional thinking. Later, a clinical picture resembling catatonia emerged. We saw her for the first time during this time. This period lasted almost four weeks and included not eating or dressing herself, loss of interest, extreme social withdrawal, decrease in psychomotor activation and sudden attacks on her younger brother when he approached her. Her overt psychotic symptoms (hallucinations, bizarre behavior and speech) continued for almost five months under treatment.

We would like to note some important points in this case. This is a very unusual case with an onset of VEOS before six years of age. Risk factors for such an early onset in this case may include a family history of schizophrenia, significant developmental delays and a dysfunctional and traumatic family environment. Differential diagnosis is another important and difficult part of this case. However, when we consider all factors together, the diagnosis of VEOS is more likely than other disorders. Family and premorbid history, onset and course of the symptoms, exclusion of organic disorders and treatment response all favour VEOS. Clinical symptomatology in this case is typical of childhood onset psychotic disorders in some aspects (e.g hallucinations including animals, bugs etc). However, the catatonic type of VEOS which we observed in this case is a very rare form of the disorder. The one month period of not eating or dressing...
herself, social withdrawal, loss of interest and decreased psychomotor activation was possibly a catatonic picture. Risperidone was significantly effective both for positive and negative symptoms, but she required higher dosage up to 2 mg/day which may be crucial in the treatment of EOS or VEOS (11). She generally tolerated medication well except for developing nocturnal enuresis after 1.5 mg/day and a weight gain of 3.5 kilograms within six months.

In conclusion, the diagnosis of VEOS requires a multidisciplinary approach, a detailed and careful differential diagnosis, with particular attention to risk factors for developing VEOS, exclusion of other neuropsychiatric disorders and a close long term follow up. The treatment of VEOS should include psychopharmacological, social and educational approaches for both the patient and family.

References