Late Onset Subacute Sclerosing Panencephalitis: Presenting Psychosis

Psikoz ile Prezente olan Geç Başlangıçlı Subakut Sklerozan Panensefalit

Geç Başlangıçlı Subakut Sklerozan Panensefalit / Late Onset Subacute Sclerosing Panencephalitis

Yavuz Altunkaynak, Zerrin Yıldırım, Devrimsel Harika Ertem, Ayten Ceyhan Dirican, Sevim Baybaş
Bakırköy Ruh ve Sinir Hastalıkları Hastanesi Nöroloji Kliniği, İstanbul, Türkiye

Özet

Subakut Sklerozan Panensefalit (SSPE), nöbet, miyokloni, ataksi, davranışsal ve kişilik değişiklikleri, ekstrapiramidal sistem bozuklukları ve görme sorunlarının eşlik ettiği kızamık hastalığının uzun dönem komplikasyonu olan bir hastalıktır. Bu yazıda atipik psikotik bozukluk teşhisiyle psikiyatri polikliniğinde takip edilmiş 19 yaşında kadın olgu sunulmuştur. Psikiyatrik bulguları baskın olan bu hasta SSPE tanısı almıştır.

Anahtar Kelimeler

Subakut Sklerozan Panensefalit; Psikoz

Abstract

Subacute Sclerosing Panencephalitis (SSPE) is the late complication of measles and is characterized by seizures, myoclonus, ataxia, behavioral and personality changes, extrapyramidal dysfunctions and vision problems. A 19 year old female patient with SSPE who was followed up at psychiatry clinic with the diagnosis of atypical psychotic disorder was presented. While psychiatric signs and symptoms were dominant, she was diagnosed as SSPE.

Keywords

Subacute Sclerosing Panencephalitis; Psychosis

DOI: 10.4328/JCAM.2163 Received: 17.11.2013 Accepted: 30.12.2013 Printed: 01.10.2013 J Clin Anal Med 2013;4(suppl 2): 220-2
Corresponding Author: Devrimsel Harika Ertem, Bakırköy Ruh ve Sinir Hastalıkları Hastanesi Nöroloji Kliniği Bakırköy, İstanbul, Türkiye.
GSM: +905301428876 E-Mail: hkaozhan@gmail.com

Introduction

SSPE is a progressive central nervous system disease caused by a defective measles virus [1]. Although it mostly occurs during childhood and early adolescence, several late-onset SSPE cases were also reported in the literature [2-4]. The course of disease is divergent. Clinical features include behavioral or personality changes, progressive mental demolition, myoclonus, focal or generalized seizures, extrapyramidal dysfunctions and vision disorders [5]. The disease may be presented atypically and may show difficulties in diagnose.

In this case report, we presented a 19-year old female patient who had admitted to psychiatry clinic due to psychotic symptoms and followed up with a diagnosis of atypical psychotic disorder. In the following period, neurologic symptoms developed and the patient was referred to our neurology department. After a complete examination and laboratory investigation, electroencephalography (EEG) and cerebrospinal fluid (CSF) findings verified SSPE diagnosis.

Case Report

A 19-years-old female patient had admitted to psychiatry clinic due to behavioral changes, anxiety, aggressive behavior, emotional lability and received antipsychotic treatment with a diagnosis of atypical psychotic disorder. She had sudden falling attacks 1-year before her admittance to psychiatry but she had not mentioned about this complaint to the doctor. Under treatment, her psychiatric symptoms partially relieved but the frequency of fallings increased. Then she was referred to our department for neurologic examination and evaluation. On physical examination, there were several scars on the patellar skin bilaterally. Neurological examination revealed ataxia, generalised myoclonic jerks, decreased deep tendon reflexes, and positive Babinski reflex. She had intellectual deterioration and behavioral changes.

Routine laboratory investigations included hemogram, sedimentation, biochemistry, liver function tests, complete urine tests, Vitamin B12 level, Syphilis, HIV, chest x-ray, electrocardiography. No pathologic finding was found. Cranial magnetic resonance imaging (MRI) showed hyperintense lesions at the subcortical white matter of both cerebral hemispheres [Figure 1]. EEG showed slow wave paroxysms in both hemispheres, sleep-induced generalized epileptiform discharges and left frontal-central neuronal hyperexitability [Figure 2]. The CSF immunological study tested positive for IgG measles antibody in 20.6 titers (Normal value range is between 0-3.4).

A diagnosis of SSPE was made based on clinical and EEG findings and laboratory tests. Isoprinosine treatment was given and valproic acid was added to control fallings.

In the first year of treatment the patient was immobile and dependent medical care.

Discussion

In this paper, we report an 19-year-old female SSPE patient presenting with personality and behavioral changes, MRI and EEG findings, and high antibody levels in serum and CSF. In literature, there are previous cases presenting with atypical findings such as ataxia, encephalopathy, and vision loss [5].

The disease is characterized by seizures, myoclonus, ataxia, behavioral and personality changes, extrapyramidal dysfunctions and vision problems. In the literature SSPE cases presenting with psychiatric symptoms such as first episode schizophrenia, young-onset dementia were rarely reported [2,6,7,8,9,10,11].

It mostly occurs during childhood and early adolescence. The disease onset after 18 is rare. Several late-onset SSPE cases were also reported in the literature [2,4,9]. In our case it is remarkable that the disease onset was 19.

Psychotic symptoms and emotional lability of our patient together with ignored falling episodes led to admittance to psychiatry clinic and had delayed the diagnosis and treatment. Rare presentation of SSPE with psychiatric symptoms and late-onset of the disease might be the reason.

Despite vaccinating programs, SSPE is still a problem in our country and other developing countries. Therefore, diagnosing atypical symptoms in the early period may be important in disease course.

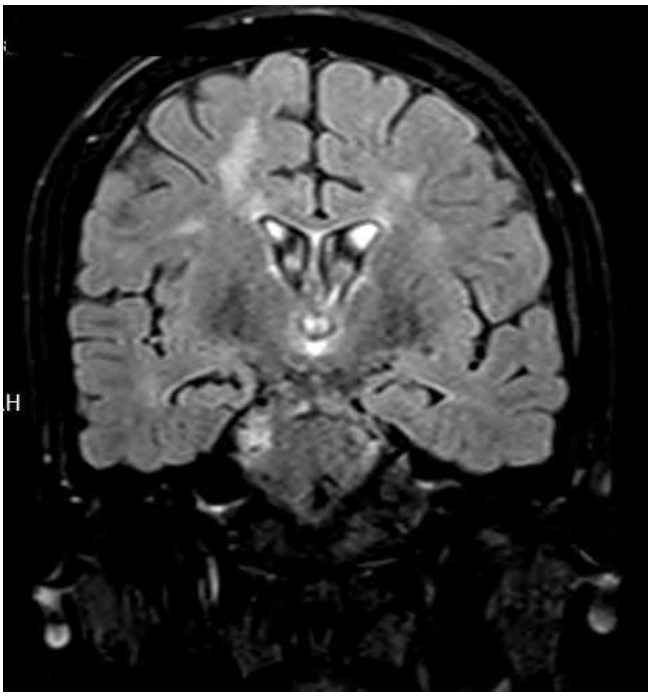


Figure 1. Bilateral subcortical white matter abnormalities on FLAIR sequence



Figure 2. Bilateral slow wave paroxysms, sleep-induced generalized epileptiform discharges and left frontal-central neuronal hyperexitability on EEG

Competing interests

The authors declare that they have no competing interests.

References

1. J. David Beckham, Marylou V. Solbrig, Kenneth L. Tyler. Bradley's Neurology in Clinical Practice, 6th ed Chapter 53B – Infections of the Nervous System : Viral Encephalitis and Meningitis; p:1254.
2. Tombul T, Anlar Ö, Sayın R, Beşiroğlu L, Dizkırıcı A. Geç Başlangıçlı ve Atipik Seyirli Bir SSPE Olgusu. Van Tıp Dergisi 2005; 12 (4):248-51.
3. Vilas D, Becerra JL, Lozano M, Soriano A, Matas L, Martro E, Dávalos A. Atypical presentation of adult-age onset subacute sclerosing panencephalitis Rev Neurol. 2012;1;54(1):60-1.
4. Singer C, Lang AE, Suchowersky O. Adult-onset subacute sclerosing panencephalitis: case reports and review of the literature. Mov Disord. 1997;12(3):342-53.
5. Gutierrez J, Issacson RS, Koppel BS. Subacute sclerosing panencephalitis: an update. Dev Med Child Neurol. 2010;52(10):901-7.
6. Duncalf CM, Kent JN, Harbord M, et al: Subacute sclerosing panencephalitis presenting as schizophreniform psychosis. Br J Psychiatry 1989; 155:557–59
7. Jähnel M.Paranoid-hallucinatory psychosis as primary manifestation of subactue sclerosing panencephalitis (SSPE) in a 19-year-old man. Psychiatr Prax. 2003;30 Suppl 2:70-2.
8. Kayal M, Varghese ST, Balhara YP. Psychiatric manifestation of SSPE. J Neuropsychiatry Clin Neurosci. 2006;18(4):560.
9. Baran Z, Hanağasi H, Uçok A. An unusual late presentation of subacute sclerosing panencephalitis with psychotic symptoms. J Neuropsychiatry Clin Neurosci. 2010;22(1):123.
10. Chakor RT, Santosh NS. Subacute sclerosing panencephalitis presenting as rapidly progressive young-onset dementia. J Pak Med Assoc. 2013;63(7):921-4.
11. Kartal A, Kurt AN, Gürkas E, Aydin K, Serdaroglu A. Subacute Sclerosing Panencephalitis Presenting as Schizophrenia With an Alpha Coma Pattern in a Child.J Child Neurol. 2013 Oct 18. doi: 10.1177/0883073813507484

How to cite this article:
Altunkaynak Y, Yıldırım Z, Ertem DH, Dirican AC, Baybaş S. Late Onset Subacute Sclerosing Panencephalitis: Presenting Psychosis. J Clin Anal Med 2013;4(suppl 2): 220-2.