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Case report

Unusual progression of an adult-onset subacute sclerosing panencephalitis (SSPE) in Turkey

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1. Introduction

Subacute sclerosing panencephalitis (SSPE) is a late complication of measles infection due to defective cell-mediated immune response in the primary measles infection [1]. The incidence is approximately 0.06 per million in the USA, on the contrary, 0.461 per million in Turkey [1,2].

The disease has a progressive course and starts with nonspecific symptoms and signs like a decline in school performance, mood changes, and behavioral problems. Later on, it manifests itself with myoclonic jerks, seizure, pyramidal, and extrapyramidal signs. Eventually, it may progress to a vegetative state and finally to death [3]. Adult cases present with a more atypical presentation as well as a more aggressive course compared with childhood SSPE. Herein we describe an adult SSPE case which presented with psychiatric symptoms and had an aggressive course.

2. Case

A 21-year-old male patient was admitted to the emergency room (ER) with rhythmic, frequent jerky movements of the trunk which involved bilateral shoulders and the neck. He experienced decreased nighttime sleep and a sentimental mood for the prior month. Initial neurological examination revealed severe apathy, abulia, very frequent myoclonic jerks of the neck and the trunk. Fever and meningeal irritation signs were absent.

Blood biochemistry was normal. Cerebrospinal fluid (CSF) analysis showed normal protein level (43,4 mg/dL [15–45 mg/dL]) with no cells. CSF FilmArray including meningitis/encephalitis panel (FA ME, BioFire Diagnostics, LLC) was negative. Brain magnetic resonance imaging (MRI) showed no structural abnormalities. Electroencephalographic (EEG) recording demonstrated periodic bilateral generalized synchronous spike-and-slow waves in couplets every 5 to 9 s (Fig. 1). Both CSF (230 U/mL [<25 U/mL]) and serum measles IgG levels (4,2 IU/L [0–0,99 IU/L]) were detected in very high titres. However, a past history for measles infection was not confirmed. With the clinical presentation, periodic discharges in the EEG recording, and high measles antibody titer in CSF he was diagnosed with SSPE. Isoprinosine 1500 mg per day was started. A total of 5 plasma exchanges was performed on alternate days. Intravenous levetiracetam was started for symptomatic treatment of myoclonus. Afterward, carbamazepine was added in a dose of 400 mg twice a day due to incomplete control of myoclonic jerks. Apathy and the abulic state resolved and the frequency and intensity of the myoclonic jerks decreased. As a result, he started to ambulate with assistance. The patient was discharged from the hospital with symptomatic anti-seizure treatment and oral isoprinosine.

Three weeks after discharge he was readmitted from the ER with impaired alertness, severe abulia, and apathy. It was reported that one week after initial discharge the patients had decreased oral intake, heavy sweating, declining ambulation and increased jerks. In the ER, his eyes would open spontaneously but; he had no verbal response or eye contact. Frequent axial myoclonic jerks were present. He was transferred to another hospital ICU and there underwent a tracheotomy and received supportive treatment.

3. Discussion

Measles is one of the world's most contagious diseases, and it may cause death. The incidence is still high in the world and has continued to rise in 2019. According to the World Health Organizaton (WHO), reported cases of SSPE rose by 300%. The WHO African region has recorded a 700% increase, the Region of the Americas 60%, the European region 300%, the Eastern Mediterranean region 100%. The reason behind this differs in different parts of the globe. In underdeveloped countries vaccination mainly tends to fail because of challenges in acquiring and administering measles vaccine. On the contrary, increasing number of families refuse measles vaccine in developed countries. Moreover, a large group of unvaccinated people have emigrated from Middle Eastern region to Eastern Mediterranean and European region. The

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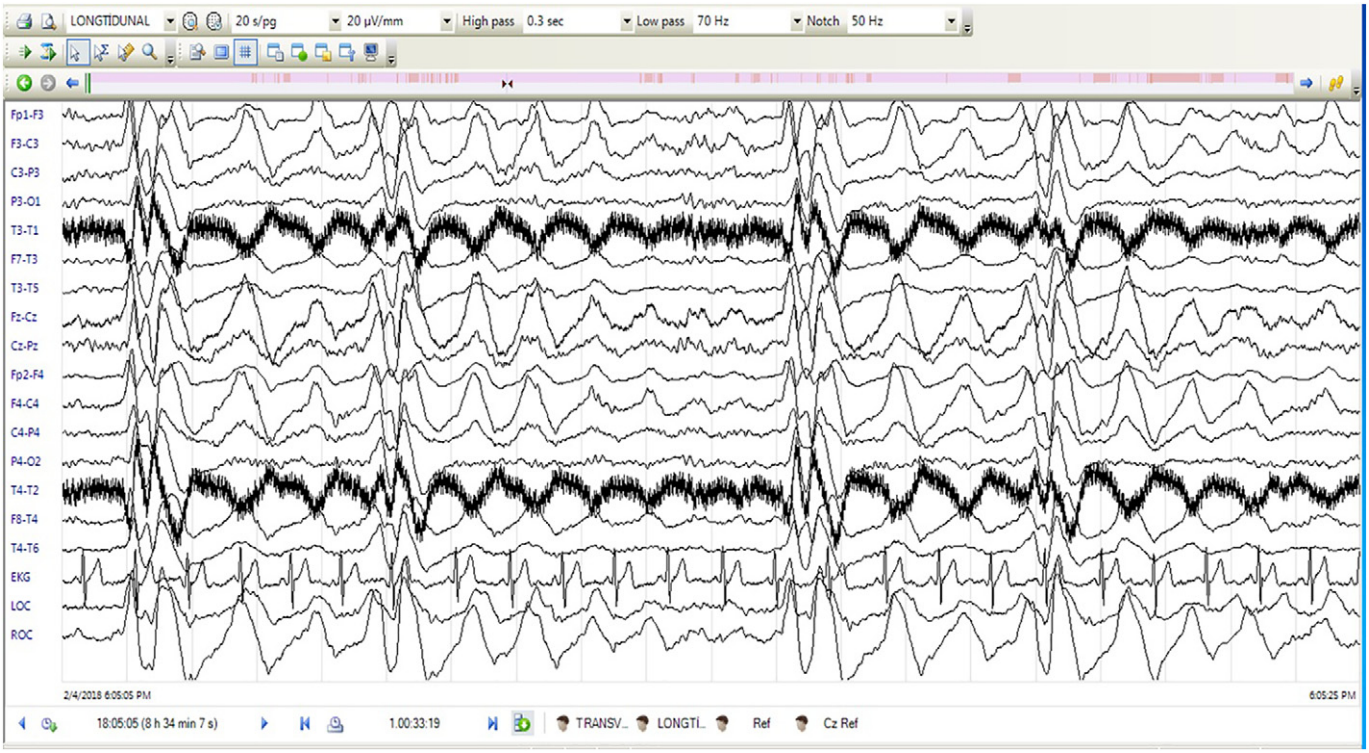

Fig. 1. Electroencephalographic (EEG) recording with periodic bilateral generalized synchronous spike-and-slow waves coupletes every 5 to 9 s.

worldwide increase in the incidence in 2017 was also observed in Turkey. 572 measles cases were reported in 2014, 342 in 2015, 9 cases in 2016, 69 cases in 2017 and 568 case in 2018 [4].

The main complication of measles is SSPE and it presents primarily during childhood and adolescence, and is rarely seen during adulthood. In a case with atypical clinical presentation or atypical age of onset, like an adult, SSPE diagnosis needs special consideration. A patient with progressive cognitive decline should cause clinicians to suspect neurodegenerative conditions including progressive myoclonus epilepsies (PME) and autoimmune epilepsies.

In the case of PME, myoclonic jerks have complex features and generalized tonic–clonic seizures may occur. The clinical course shows progression manifest as progressive cognitive deterioration and ataxia which usually coexist. EEG shows generalized spike- or polyspike-and-waves with generalized background slowing and a photoparoxysmal response. Nuclear gene defects may be inherited in an autosomal recessive or an autosomal dominant manner [5].

As for autoimmune encephalitis, the clinical presentation includes seizures, neuropsychiatric features, movement disorders and cognitive decline similar to SSPE. An autoimmune epileptic encephalopathy is a potentially treatable neurological syndrome. It is characterized by the coexistence of a neuronal antibody in the CSF and serum. Brain MRI studies show T2- and T1-hyperintensities in the basal ganglia and medial temporal lobe. N-methyl-D-aspartate receptor-antibody encephalitis has a specific interictal EEG pattern, the “extreme delta brush”, that is found in up to 30% of patients during the course of the illness. It is a pattern of rhythmic delta activity with superimposed beta frequency “riding” on each delta wave unique for adults with autoimmune encephalitis [6,7].

Autoimmune encephalopathy panels are not done in our hospital's laboratory and therefore, patients must pay for the test in private laboratories. The present case report did not have an autoimmune encephalopathy panel in serum or CSF limiting definitive exclusion. However, the diagnosis of SSPE was established by clinical features supported by elevated CSF measles antibody titers and periodic, stereotyped, high voltage epileptiform discharges on EEG [1].

Adult-onset SSPE has a longer prodromal stage and shows a higher proportion of negative or history of undocumented measles infection than pediatric cases and may present with atypical clinical features such as psychotic features, viral retinitis, complex seizures, ataxia, extrapyramidal signs and speech abnormalities [8]. However, Prashanth et al. found no difference between the clinical profile of adult-onset and juvenile SSPE [3]. In the published literature the mean delay between disease onset and diagnosis is 6 to 10 months; however, our case was diagnosed after a month of symptom presentation [3,9]. Unfortunately, the disease progression was very aggressive, so that when diagnosed he was almost bedridden. Adult-onset SSPE is generally known to have a more aggressive course [9]. Some authors reported shorter survival and most patients die within 1–2 years [9], but others reported longer survival with a higher rate of spontaneous remission [1].

Thus far, antiviral drugs (amantadine, isoprinosine, ribavirin) and immunomodulatory modalities (methylprednisolone, interferon alpha, intravenous immunoglobulin, plasmapheresis) have been tried alone or in combination [3]. However, there is no currently proven curative treatment for SSPE. The present case was treated with isoprinosine, plasmapheresis and symptomatic anti-seizure drugs. Even though he underwent a minor improvement after therapy induction, his clinical status worsened rapidly. Because we started isoprinosine and plasma exchange together, we could not tell for sure which one caused the temporary improvement or whether minor remission was actually a part of the natural course of the disease.

4. Conclusion

As a disease mainly involving pediatric patients, SSPE may elude diagnosis in adult cases if not included in the differential diagnosis. At this time, SSPE is a fatal disease without any curative treatment. Measles vaccination is the only measure that can reduce the risk of SSPE, thus, vaccination programs should be taken seriously for public health concern.

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