

An Unusual Findings of Elevated Urinary Copper Excretion in a Patient with Subacute Sclerosing Panencephalitis (SSPE): A Case Report

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Abstract

Subacute sclerosing panencephalitis is a progressive neurological disorder of childhood and early adolescence. It is caused by persistent defective measles virus. We report a ten years old normally developed female patient who came with a history of drop attacks while walking, declining scholastic performance, progressively increasing myoclonic jerks. She had history of measles at five years of age though she was vaccinated as per EPI schedule. Physical examination and cerebrospinal fluid findings along with EEG changes in addition supported its diagnosis as a case of SSPE. The presence of increased urinary excretion of copper in SSPE is so far not yet reported in any published literature. [Journal of National Institute of Neurosciences Bangladesh, 2017;3(2): 113-115]

Keywords: Subacute sclerosing panencephalitis; urinary copper; myoclonic jerks

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Introduction

Subacute sclerosing panencephalitis is a chronic complication of measles with a delayed onset, the outcome of which is nearly always fatal. It appears to result from a persistent infection with an altered measles virus that is harbored intracellularly in the central nervous system for several years. After 7-10 years the virus apparently regains virulence and attacks the cells in the central nervous system that offered the virus protection. This slow virus infection results in inflammation and cell death, leading to an inexorable neurodegenerative process. Virtually all patients with SSPE eventually succumb². This paper is to present a case of SSPE who has elevated urinary copper excretion.

Case Presentation

A previously healthy and developmentally normal ten years old girl born to non-consanguineous parents presented with a history of unsteady gait, drop attacks while walking and declining scholastic performance for last eleven months. This was followed by progressively increasing myoclonic jerks and inability to sit or stand for last three months. She has no history of jaundice, behavioral abnormality, headache, visual disturbance, and bowel or bladder incontinence. She has a history of measles at five years of age and she was immunized as per EPI schedule with no significant family history. On physical examination she was conscious, oriented in time, place and person. Increased muscle tone with

decreased strength (3/5) in all four limbs and ankle clonus on left side was found. Meningeal signs were absent, cranial nerves and sensory examination were normal. Slit lamp examination of eye excluded the presence of Kayser-Fleischer(K-F) ring. Full blood count, urea, creatinine, electrolytes, calcium, liver function tests were normal. Cerebrospinal fluid analysis showed absence of pleocytosis (2 cells/mm³) with normal protein (18.5mg/dl), glucose (75mg/dl) and lactate (12mg/dl) concentration. Cerebrospinal fluid was also assayed for measles IgG antibody which was positive (AI: 2.9).

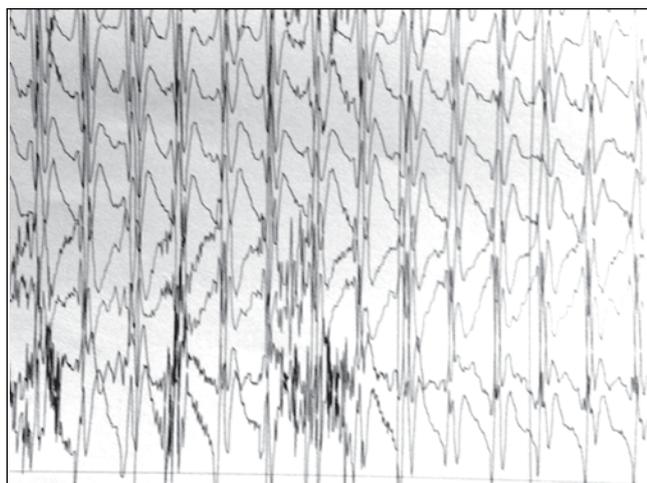


Figure I: EEG showing Periodic Complexes containing spikes and slow waves (A Standard 10 Sec. Epoch)

Serum ceruloplasmin was normal (260mg/l) but twenty four hours urinary copper was raised on two occasions measured two months apart (2363 μ gm/24 hours and 1817 μ gm/24 hours respectively) following Penicillamine challenge. Her other three sib's serum ceruloplasmin levels were also measured and they were within normal limits and slit lamp examination showed no K-F rings in their eyes. Electroencephalogram showed frequent periodic bursts of high voltage complexes containing spikes and slow waves from the background at regular interval followed by burst suppression (periodic complexes). Magnetic resonance imaging of brain revealed periventricular subtle hyper intensities in T2/FLAIR sequences which were isointense in T1-weighted sequence. There was also generalized cortical atrophy with widening of cortical sulci, fissures, ventricles and thinning of corpus callosum. Patient was treated with valproic acid and clonazepam.

Discussion

Subacute sclerosing panencephalitis is a rare progressive neurological disorder of childhood and

early adolescence that is more frequent in childhood. Subacute sclerosing panencephalitis remains an important public health issue in parts of the developing world due to limited measles immunization policies^{3,4}. Children infected in the first two years of life are at greater risk, and case series consistently show SSPE to be more frequent in boys. The median interval between acute measles infection and SSPE is eight years, with a range from 2-12 years¹.



Figure II: Same Epoch of 5 Sec.

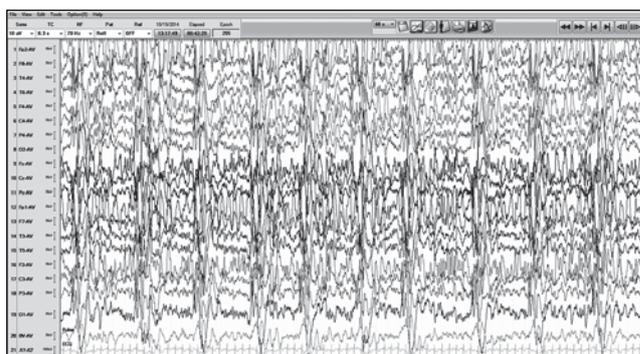


Figure III: Same Epoch (30 Sec.) Showing the Periodicity

There are four stages of clinical impairment described in SSPE. In stage I, mild intellectual deterioration and behavioral changes without any apparent neurological signs or findings. The second stage is characterized by typical periodic or quasi periodic axial myoclonic jerks whose manifestation causes recurrent falls. Generalized rigidity with extra-pyramidal features and unresponsiveness appear in stage 3. Stage 4 is the terminal stage of the disease and is characterized by minimal conscious state and later akinetic mutism associated with persistent high fevers and bouts of generalized sweating; both being due to autonomic failure³. The prognosis of SSPE is poor with death typically occurring within two to four years of onset as no curative treatment is available³.



Figure IV: MRI FLAIR image showing periventricular subtle hyperintensities with generalized cortical atrophy

The diagnosis of SSPE is based on typical clinical features, periodic EEG complexes and elevated measles antibody titre in CSF. After appearance of myoclonus the diagnosis of SSPE can reliably be established with the help of Dyken's diagnostic

criteria⁵. Our patient fulfilled three out of five Dyken's diagnostic criteria for SSPE (criteria 1, 2 and 4). Agarose gel electrophoresis of CSF to identify oligoclonal band of immunoglobulin and brain biopsy were not performed. To rule out Wilson's disease investigations were done and patient had high urinary copper excretion with normal serum ceruloplasmin and absence of K-F ring in the eyes. Her three sibs also had normal serum ceruloplasmin level with absence of K-F ring.

Conclusion

Low level of serum ceruloplasmin can be found in many neurological diseases including SSPE⁶. But till date no published literature reported the excretion of high urinary copper in a case of SSPE. This observation remains to be explained in future.

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