A conundrum of West syndrome, behavioral problems and parental expressed emotions: a case report

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ABSTRACT

West syndrome (WS) is the most common symptomatic syndrome in infancy characterised by epileptic spasms, hypsarrhythmia and neurodevelopmental problems. Epileptic spasms remain in many ways a conundrum, and the ideal intervention, as well as how to screen patients to provide optimal care and certainly its genetic cause, remains puzzling. It is important to screen infants for early recognition and intervention to achieve the optimal outcome. We hereby discuss the approach to management of a boy aged 4½ years old with WS and behavioural problems and of parental expressed emotions.

INTRODUCTION

West syndrome (WS), a form of infantile epilepsy characterised by the triad of infantile spasms, intellectual disability and hypsarrhythmia (typical electroencephalographic findings of chaotic and disorganised background activity with asynchronised, large-amplitude slow waves mixed with single focal and multifocal spikes and slow waves followed by attenuation), was first described by Dr West in 1841.1 The burden of neurodisability, that is, neurological impairments, neurodevelopmental disorders (intellectual disability, autism spectrum disorders and attention deficit hyperactivity disorder (ADHD)) and learning difficulties, is high in children and adults with epilepsy, and more so in WS as it affects children at a very young age, that is, infancy.2 In this paper, we discuss the approach to the management of a boy aged 4½ years old with WS and behavioural problems and of parental expressed emotions.

CASE REPORT

A boy aged 4½ years old was brought to the outpatient services of the Department of Psychiatry with complaints of not being able to sit in one place for more than a few seconds and excessive behavioural problems. On exploration, it was found that the boy was born via preterm vaginal delivery and cried immediately after birth. There were no intranatal/postnatal complications. He had adequate birth weight and was growing normally until approximately 5 months of age when he started to have sudden, jerky spasms of the neck and upper limbs occurring in clusters (5–6 spasms/cluster) for approximately 3–4 min, especially when waking up from sleep. A private physician started him on valproate syrup within a month of onset of these symptoms but without any response. Within 3–4 months of symptom onset, he was brought to a tertiary care hospital where he was diagnosed with WS. Investigation was conducted that includes MRI brain, metabolic work-up and spectroscopy with no abnormalities detected, and electroencephalogram (EEG) showed characteristic findings of hypsarrhythmia. He was treated with adrenocorticotropic hormone injection (150 IU/m² for 10 weeks) and valproate syrup 35 mg/kg/day (continued for 3 years). He became spasm-free for the next 3 months after initiation of treatment and his parents also became overcautious and overprotective of him. By this time, his mother noticed that he had lost the milestones (social smile) that he earlier had achieved and also had delayed fine motor and gross motor development (eg, pincer grasp and sitting with support at 1 year). He started to speak bisyllables at 1½ years of age, played with toys and with children of his age, but had started to throw temper tantrums (screaming, shouting, trying to pull his hair, rolling down on the ground, hitting his parents and pulling their hair) if his demands were not fulfilled. His parents ultimately would give in to his demands after initial reluctance to calm him down. Around the same time, it was also observed that he did not sit in one place for more than a few seconds, and on many occasions he had gone out of the house (whenever he found the gates were open) and was not able to find his way back home. Also, he readily approached strangers, mingled with them, and took sweets
and toys offered to him, which worried his parents. By 2½
years of age, he was walking independently, spoke a few
meaningful words, scribbled and was put in play school.
However, his parents received frequent complaints from
his teachers that he disturbed other children, took away
their pencils and erasers, did not sit in class, and cried,
screamed, bit and pulled his teachers’ hair when he was
being disciplined. At times, his teachers would hit him
in an attempt to control his behaviour but to no avail,
and they finally stopped attending to him and allowed
him to do whatever he wanted. Due to these complaints,
the mother became very distressed and would beat
him up on many occasions. His behavioural problems
gradually increased both at home and school, and he
would now snatch other children’s food and screams if
they protested, keeps on running in circles on the play-
ground, and bangs his head and bites if he is prevented
from doing so. He would be aware and conscious while
indulging in these behaviours. His mother has become
extremely distressed because of these and also because
she found him to be lagging in all domains of growth and
development compared with age-related peers and would
often be critical of him. However, it was important to note
that he would not indulge in such behaviours in front of
a strict teacher or with children apparently stronger than
him, or for 4–5 hours whenever he was reprimanded by
his mother. For these behaviours, he was given risper-
done tablet 0.5 mg/day for 1–2 months by the treating
physician, with initial improvement but not sustained
despite good compliance. Thereafter, he was given a trial
of methylphenidate tablet 10 mg/day for 2 weeks and
aripiprazole tablet 2.5 mg/day for 4–6 weeks without any
improvement. Valproate syrup was stopped after 3 years
of no seizure recurrence and he was then referred for
psychiatric consultation for behavioural problems. He
was admitted to the psychiatric unit for management.
Ward observation revealed adequate social communica-
tion, unclear speech, no tics/abnormal movements and
no hearing/visual impairment. Temperamentally (as
per the Temperament Measurement Schedule),4 he is
an active child, readily approaches others, is less adapt-
able to any change, reacts instantaneously with high-
intensity reactions for unfulfilled wishes and becomes
irritable if demands were not met, but would persist
with a task until he is faced with difficulty. In addition
to being a temperamentally difficult child, inconsistent
parenting led to an increase in behavioural problems.
His mother was very rigid, punitive and non-tolerant to
deviance and is inconsistent, comparing him with other
peers and pointing out his deficits in development; the
father’s behaviour was opposite that of the mother. No
abnormality was detected on general physical examina-
tion, and mental status examination revealed a tidy and
well-groomed boy of stated age. Eye-to-eye contact was
limited initially but improved as the interview progressed.
He followed instructions, drew circles on a paper for 5–7
min at a stretch, and started roaming around the room
after about 10 min but complied with the instructions to
sit down, did not initiate conversation, took a long time
to reply to questions, and his speech was unclear. He did
not show any signs of separation anxiety when his parents
were asked to leave the room; however, he started to cry
and scream when he was being forcefully taken out of
the room, but calmed down when left alone. His intellec-
tual function was assessed on the basis of history of social
maturity (Vineland Social Maturity Scale applied) given
by the parents and the Gesell Drawing Test Score. He
had an IQ score of 67 (mild intellectual disability; mental
age 3 years) on assessment. High negative expressed
emotions, high expectations from the child and distress
from the parents especially the mother were evident on
the family front. Additionally, he was refused admission
due to behavioural problems, which increased parental
burn-out. During his ward stay, it was observed that the
patient would have five to six episodes per day of tempo-
rary cessation of activity and vacant stare lasting for 1–2
s and was non-responsive, but with no other abnormal
activity. These events, although seen by the mother,
were not reported. Paediatric neurology consultation
and video EEG confirmed ongoing seizure activity, for
which valproate syrup up to 600 mg/day was prescribed.
The mother was evaluated and managed for moderate
depressive disorder. Parental psycheducation for intel-
lectual disability and associated behavioural problems,
epilepsy, and for inconsistent parental handling and the
role of expressed emotions was also provided. Now, the
child is interacting well and attends to a task at hand for
10–12 min without getting distracted. Positive reinforce-
ment, structured routine and realistic expectations with
no comparisons, as well as parent management training,
were employed, which brought about positive results to
the child.

**DISCUSSION**

The diagnosis of WS in the index case was not difficult
given the characteristic presentation and EEG findings.
Psychomotor development before convulsions is essen-
tially normal in most cases, as was the finding in the index
case.4 Multiple cognitive, behavioural and emotional
problems are experienced by children with epilepsy, in
addition to the burden of epilepsy itself, the most common
problem is impaired attention and hyperactivity.5 The
index case also displayed a variety of behavioural prob-
lems, which were also aggravated by the mother’s negative
expressed emotions and inconsistent parental handling.
Behavioural problems improved following the environ-
mental modification, employing behavioural techniques,
and improving parental handling skills.

This case also highlights the fact that ‘Every child
presenting with hyperactivity is not necessarily a case of
ADHD (attention deficit hyperactivity disorder)’. The
hyperactivity in the index case was accounted for the
seizure activity, manifested as absence seizures clinically
but was missed by the family members. Hyperactivity
improved after valproate syrup was added. Literature
supports worsening neurocognitive and behavioural dysfunction due to ongoing epilepsy and interictal discharges, a concept referred to as ‘epileptic encephalopathy’, and children presenting with such symptoms may not fit neatly into nosological diagnostic systems of ICD-10 (International Classification of Diseases-10) or DSM-5 (Diagnostic and Statistical Manual of Mental Disorders-5). In addition to ongoing seizure activity causing hyperactivity, this boy was temperamentally a difficult child. Temperamental difficulties were further worsened by inconsistent parental handling and punitive mother. Although he was an active child, he did not meet the diagnostic criteria for ADHD as his hyperactivity improved on initiation of valproate and also because he was able to attend to a task for 15–20 min at a stretch with an attention span more than adequate for preschool children.

Cognitive and behavioural comorbidities in association with epilepsy are underdiagnosed and consequently undertreated. In a population-based study, 80% of school children with active epilepsy had behavioural and/or cognitive comorbidity and only a third had been diagnosed before the study was undertaken. More importantly, it adversely affects psychosocial outcomes.

The approach to evaluating preschoolers with hyperactivity disorder should include assessment for neurodevelopmental disorders such as ADHD, autism spectrum disorder and intellectual disability with problematic hyperactivity (not appropriate to mental age), physical problems, sensory impairments, environmental issues (neglect, abuse, social isolation/deprivation), and temperament of the child. Thus, it becomes important to examine preschoolers presenting with such a plethora of symptoms in a holistic way and not be in a rush to put a diagnostic label. Rather, efforts should be made to understand the genesis of the problems in its entirety.

Correction notice This article has been corrected since it was published. Author bio has been updated.