Childhood disintegrative disorder: A case report

Sadia Sultan, T. Ravi Kanth

ABSTRACT

Introduction: Childhood disintegrative disorder (CDD), is a very rare disorder that is characterized by development of autistic like picture and marked regression following some period (at least two years) of normal development. The condition was first described by Thomas Heller, in 1908, also called as Heller’s syndrome. Typically, the age of onset is 3–4 years, but it can happen any time up to age 10. Case Report: A 6-year-old child brought to the OPD with aggressive behavior, communication problem, repetitive body movements and overactivity since past 18 months. Onset is insidious and progressive. History suggestive of measles four years back and GTCS for two years of age. Since last one and half years there is insidious and progressive regression in developmental milestones which were previously attained at appropriate age. Conclusion: To conclude, the study states the importance to the clinicians about the possibility of the diagnosis of CDD following measles, especially when there is a regression in developmental milestones following normal development.

Keywords: Autistic spectrum disorders, Childhood disintegrative disorder, Disintegrative psychosis, Heller’s syndrome

INTRODUCTION

Childhood disintegrative disorder (CDD), is a very rare disorder that is characterized by development of autistic like picture and marked regression following some period (at least 2 years) of normal development [1]. The condition was first described by Thomas Heller, in 1908 who reported a series of six cases that displayed a marked and persisting developmental regression after 3 or 4 years of normal development. It is also called as Heller’s syndrome, dementia infantilis, disintegrative psychosis [2].

It is presently grouped with the autism spectrum disorders according to the recently published Diagnostic and Statistical Manual of Mental Disorders, 5th edition [3]. Typically, the age of onset is 3–4 years, but it can happen any time up to age 10 [4]. While a very few cases of childhood disintegrative disorder are caused by cerebral lipidosis or leukodystrophy, in most cases a cause cannot be established [5]. It remains quite unknown whether it constitutes an atypical variant of ASD or some meaningfully different condition. The following case is a child diagnosed with CDD.

CASE REPORT

A male child aged 6 years was brought forcibly by parents with complaint of irritable and aggressive behavior, over activity, communication problems and repetitive body movements since past 18 months. The onset was insidious and gradually progressive.
The child was born as one of the twins, by full term normal vaginal delivery in hospital setting, no history intranatal maternal infections, no complications after birth. The child attained age appropriate motor and language milestones and acquired social skills until 4 1/2 years of age. He had history of epilepsy (GTCS type) since the age of two years and was on irregular medication since then. History of rash four years ago suggestive of measles. Informant is unable to recall whether the seizure started before or after this episode of rash and fever.

At the age of 4 1/2 years the parents noticed that the child was increasingly irritable, restless, became withdrawn, developed gaze avoidance and started having nightmares. Over the next few weeks he became overactive and started screaming and running haphazardly, and also developed peculiar repetitive movements like flapping hands, jumping and wrist biting. He lost interest in studies and stopped going to school and his speech also deteriorated to a few unintelligible words, and finally he became totally mute. He gradually stopped relating to people as well as to his parents and no longer enjoyed being touched or cuddled, showed no interest in playing with other children. He would not even indicate his toilet needs or ask for food or other things, he stopped bothering about his hygiene, started eating mud and other waste materials later at one point ate his own feces and subsequently he passed worms in his stools for which he was treated by a local physician. He even lost the toilet training he acquired previously and started to pass stools and urine in clothes. This regressive course continued for another 6 to 8 months after which he developed aggressive behaviour like hitting, throwing and breaking objects.

With the above complaints the patient was brought to SVS General Hospital to the Psychiatric OP. He was admitted in psychiatry ward all the routine investigations (CBP, LFT, RFT, TFT) were done and were normal. EEG showed hyper synchronous discharge in temporal region with secondary generalization. MRI of brain showed mild atrophy of right hemisphere and white matter changes in the right fronto-temporo-parietal regions. On examination, he was observed to have hyperactivity, flapping hands, wrist biting, fleeting eye contact with examiner, and was unable to communicate verbally or non-verbally, hence IQ testing could not be done.

The patient was started on risperidone 1 mg once a day, atomoxetine 10 mg per day, along with divalproate sodium 400 mg per day in divided doses. Over the next 4 week he showed some improvement, the symptoms of over activity like running haphazardly has reduced and fine motor skills improved as is evident in his holding the glass and drinking by himself, less spilling of food while eating by himself. There is also significant improvement in aggressive behavior that he was no more hitting and throwing objects. He also stopped eating waste materials and started to indicate that he want to pass urine or stools by stripping his innerwear. He started speaking bi-syllables to call his family members and showing water and food if he is thirsty or hungry.

He was referred to a higher center wherein he is being given special training to improve his cognitive abilities. The follow up has to be done.

DISCUSSION

Childhood Disintegrative Disorder (CDD) is an extremely rare condition, only 173 cases have been reported until now since 1908 as reviewed by Volmar and colleagues [1]. The condition can be diagnosed if the symptoms are preceded by at least two years of normal development and the onset is prior to age 10 years though the mean age of onset is 3–4 years and may be insidious or abrupt [6]. The patient was diagnosed as Childhood Disintegrative Disorder (CDD) as per DSM-IV-TR diagnostic criteria shown in the following Table 1 [6].

The condition was not included in either DSM-III or DSM-III R on the assumption that the condition was almost always the function of some identifiable medical condition, but review of cases has not supported this [7, 8]. For that reason, CDD was included as a separate disorder in the fourth edition of the Diagnostic and Statistical Manual of Mental Disorders, or DSM-IV, in 1994 [9]. But in the recent review it lost its separate identity and was included under a larger group of autism spectrum disorders that was published in the fifth edition of the Diagnostic and Statistical Manual of Mental Disorders, or DSM-V, 2013 [3].

Table 1: DSM IV-TR Criteria for CDD

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<th>DSM-IV-TR Diagnostic Criteria for Childhood Disintegrative Disorder</th>
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<td><strong>A.</strong> Apparently normal development for at least the first 2 years after birth as manifested by the presence of age-appropriate verbal and nonverbal communication, social relationships, play, and adaptive behavior.</td>
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<td><strong>B.</strong> Clinically significant loss of previously acquired skills (before age 10 years) in at least two of the following areas:</td>
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<td>a. expressive or receptive language</td>
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<td>b. social skills or adaptive behavior</td>
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<tr>
<td>c. bowel or bladder control</td>
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<tr>
<td>d. play</td>
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<td>e. motor skills</td>
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<tr>
<td><strong>C.</strong> Abnormalities of functioning in at least two of the following areas:</td>
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<td>a. qualitative impairment in social interaction (e.g., impairment in nonverbal behaviors, failure to develop peer relationships, lack of social or emotional reciprocity)</td>
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<tr>
<td>b. qualitative impairments in communication (e.g., delay or lack of spoken language, inability to initiate or sustain a conversation, stereotyped and repetitive use of language, lack of varied make-believe play)</td>
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<td>c. restricted, repetitive, and stereotyped patterns of behavior, interests, and activities, including motor stereotypes and mannerisms</td>
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<td><strong>D.</strong> The disturbance is not better accounted for by another specific pervasive developmental disorder or by schizophrenia.</td>
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Fombonne reviewed 32 epidemiological surveys of autism and PDD, CDD was mentioned only in four studies. The pooled prevalence estimate across these studies was 1.7 per 1,00,000 [10]. There are high rates of EEG abnormalities and seizure disorders observed as evident in our case [1]. The condition has been associated with various general medical conditions and while intensive search for such conditions is always indicated, they are not usually found [1]. Given the highly distinctive pattern of onset, a search for potential genetic factors is clearly indicated. One case report points at a possible genetic link with autism in a case where autism and CDD occurred in two half-brothers [11].

Speech and language therapy, occupational therapy, social skills development, and sensory integration therapy may all be used according to the needs of the individual child. Currently, there are no pharmacological interventions that specifically target the core symptoms of PDD. However, studies have demonstrated that atypical antipsychotics and selective serotonin reuptake inhibitors may be beneficial for behavioral problems associated with PDD [12]. In our case study, hyperactivity and psychotic behavior like eating waste materials improved after taking risperidone and atomoxetine. Dopaminergic dysfunction has been reported to be associated with decreased motor coordination and given that atypical antipsychotics can be thought of as a dopamine system stabilizer, this may represent the mechanism of therapeutic action [13]. The seizures were controlled by divalproate sodium. The largest follow-up study was conducted by Mouridsen on 39 cases over a period of more than 22 years. It was seen that individuals with CDD had a lower overall functioning, were more aloof and had a great incidence of co-morbid epilepsy [14]. This supports the notion that the outcome in CDD is poorer than in autism spectrum disorders in general. Considering the limited number of CDD case reports in the literature makes any case report important and useful in improving the knowledge about this rare disorder.

CONCLUSION

The case report thereby states the importance of the diagnosis of CDD in the patients presenting with regression in the normally acquired age specific developmental milestones following the viral infections such as measles and chicken pox.

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Author Contributions

Sadia Sultan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

T. Ravi Kanth – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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REFERENCES
