Role of Early Multimodal Interventions in a Case with Autistic Regression

Alka Nizamie, Uma Sengupta, Biswa Ranjan Mishra, Samir Kumar Praharaj, and S. Haque Nizamie

Abstract- Autistic regression consists of paucity of social and emotional reciprocity, disorders of language and communication, and stereotyped behaviors that are noted after a period of normal or near-normal development for one or two years. We report a case who presented autistic regression symptoms but was improved with comprehensive multi-modal treatment approach in motor, language, and social domains, and also in the activities and skills of daily living.

Key Words: Autistic regression, Autistic epileptiform regression, Language regression, Landau-Kleffner syndrome, Multi-modal treatment

Acta Neurol Taiwan 2010;19:51-56

INTRODUCTION

Language regression in childhood following an apparently normal language development can lead to disabling consequences. This may be seen in deafness, autistic regression, elective mutism, Landau-Kleffner syndrome, mixed receptive/expressive disorder, or conditions involving organic insult to any language areas. Autistic regression consists of paucity of social and

emotional reciprocity, language and communication disorders, and stereotyped behaviors that are noted after a period of normal or near-normal development for one or two years. The loss of previously acquired language and social as well as cognitive skills is usually noted during the second year of life, most likely between 15 and 30 months of age⁽¹⁾. The prevalence of regression varies from 15% to 47% of children diagnosed with an autism spectrum disorder⁽²⁾. Changes are often gradual, taking

From the Associate Professor of Clinical Psychology & Director (Academics), Deepshikha Institute for Child Development and Mental Health, Arya Samaj Mandir, Shraddhanand Road, Ranchi, Jharkhand, India; Physiotherapist, Deepshikha Institute for Child Development and Mental Health, Arya Samaj Mandir, Shraddhanand Road, Ranchi, Jharkhand, India; Junior Resident in Psychiatry, Central Institute of Psychiatry, Kanke, Ranchi, Jharkhand, India-834006; Senior Resident in Psychiatry, Central Institute of Psychiatry, Kanke, Ranchi, Jharkhand, India-834006; Director & Head of Center for Cognitive Neurosciences, Central Institute of Psychiatry, Kanke, Ranchi, Jharkhand, India-834006.

Received June 2, 2009. Revised July 29, 2009. Accepted August 24, 2009.

Reprint requests and correspondence to: Samir Kumar Praharaj, MD., D.P.M. Senior Resident in Psychiatry, Central Institute of Psychiatry, Kanke, Ranchi, Jharkhand, India-834006.

E-mail: samirpsyche@yahoo.co.in

place over the course of weeks to months, although in some patients fairly abrupt changes become evident over the course of days⁽²⁾. In most of the cases the etiology of the deterioration usually remains unknown⁽³⁾. The course of regression is characterized by a prolonged period of stagnation lasting for weeks or months, usually followed by a slow course of incomplete recovery of language and other skills⁽²⁾. We present a case who showed autistic regression, but had good response to combined treatment strategies of psychopharmacological and behavioral interventions.

CASE STUDY

Ms. A, a 4-year-old girl child, was born out of nonconsanguineous marriage through caesarian section because of polyhydramnios and cord tie during the antenatal period. The baby had normal birth cry, showed no obvious complications, and received immunization as per schedule. She had a delay of 2-3 months in attaining head control, sitting, and standing without support, and was able to walk just a few steps by $1^1/_2$ years. On the other hand, she attained social smile timely with adequate emotional responsiveness, and would respond to her name by 9-10 months. She was able to speak first words clearly by one year.

From the age of $1\frac{1}{2}$ to 2 years it was noticed that she gradually lost her previously acquired speech and was left with the ability to produce only few sounds. She became socially withdrawn with poor eye contact and was less responsive to environmental changes. She also lost the capability of walking and gaze fixation. There was generalized hypotonicity of the limbs, with the appearance of hyperactive behaviors including stereotypic as well as rocking movement and self-injurious actions.

At 3 years of age she had repeated seizures, which were mostly nocturnal occurring in the first half of the

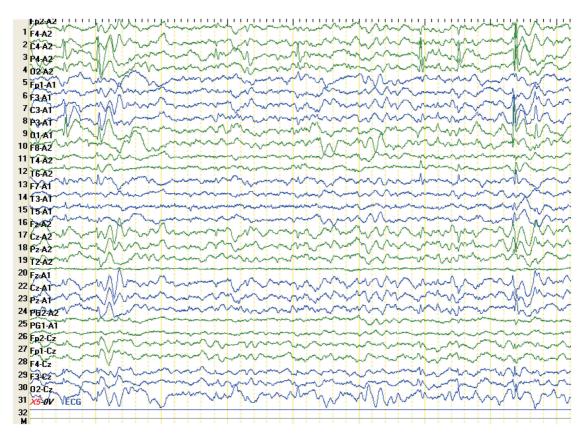


Figure 1. EEG showing spike and slow waves over bilateral fronto-centro-temporal regions.

Developmental Levels in Months	Perceptual Fine Motor	Cognition	Language	Social/Emotional	Self-care			Gross Motor
					Reading	Toileting	Dressing	51000 MOTO
32-35								
28-31								
24-27								
20-23								
16-19								
12-15							Í	
9-11					-			1
6-8				/				
3-5								
0-2	•	•						

Figure 2. Schaffer's Developmental Scale before intervention at the age of 4 years.

Developmental	Perceptual Fine Motor	Cognition	Language	Social/Emotional	Self-care			Gross Motor
Levels in Months					Reading	Toileting	Dressing	STOCK MOTOR
32-35								
28-31								
24-27								
20-23						~		
16-19					/		1	
12-15					1			-
9-11				_ /				
6-8	_							
3-5	-							
0-2								

Figure 3. Schaffer's Developmental Scale after intervention at the age of 5 years.

nighttime sleep. The semiology consisted of getting up from sleep with blank stare, followed by deviation of mouth angle and head towards the left side. Generalized tonic-clonic movements then developed and usually lasted for 1-2 minutes. Thereafter she would become lethargic and go off to sleep for another half an hour or so. The clinical pictures were suggestive of complex partial seizure with secondary generalization. The awake electroencephalogram (EEG) showed frequent occurrence of

independent or bisynchronous high voltage spike and slow waves over fronto-centro-temporal region (fig 1). The magnetic resonance imaging (MRI) scan of the brain with contrast suggested generalized cerebral atrophy disproportionate to the age, more marked in the frontal lobe.

In view of her apparent normal development except for the slight delay in motor milestones in the first $1^{1}/_{2}$ years of life, followed by loss of previously acquired lan-

guage ability as well as social and motor skills. Also, there are clinical seizures with epileptiform abnormalities in EEG. A diagnosis of autistic epileptiform regression was considered. She was treated with oxcarbazepine up to 900 mg per day, and her seizures were adequately controlled. Risperidone 1 mg per day was given for stereotypic and self-injurious behaviors. Specific psychosocial interventions and physiotherapy were done by trained personnel for one year. For the assessment of developmental level, Schaffer's Developmental Scale was administered at baseline and after intervention to evaluate the improvement. Visual tracking exercises were instituted, resulting in significant improvement in her eye contact. Speech therapy techniques such as tongue exercises, blowing, sucking, biting, vocalizing, masking the sound and lip reading were applied. She was able to produce 2-3 words clearly after the therapy. She could also express her needs non-verbally. In order to improve her attention span, she was engaged in activities such as matching, sorting small objects, and picture identification with a mirror. Over-corrective shoes and weight bearing exercises markedly improved her capability to balance herself and walk independently. Her rocking movements and stereotype behaviors were significantly reduced after play therapy and arrangement of proper height for sitting. In summary, this comprehensive multi-modal treatment approach resulted in remarkable improvements in motor, language, social skill, and activities of daily living (see figs. 2 and 3).

DISCUSSION

Epilepsy is the most common neurological comorbidity in autism with a median incidence rate of 16.7%. The figure may vary among studies because of the age distribution of the sample, the degree of mental retardation, and the type of language disorder⁽⁴⁾. Studies suggest that there are two peaks of seizure onset in autism; one in early childhood and the other in adolescence⁽⁵⁾. In studies that include younger preadolescent children with autism, the reported rate of epilepsy is less than 10%. Epilepsy and epileptiform EEGs are more common in autistic children who experience regression^(6,7). The sub-

group of patients with autistic regression along with epileptiform EEGs or seizures have been classified as autistic epileptiform regression^(5,8). It has been suggested that there is an association between epilepsy and autistic regression^(7,9).

Typical Landau-Kleffner syndrome is characterized by an age of onset from 3 to 10 years in patients with insidiously or abruptly acquired aphasia but previously normal language development, verbal auditory agnosia, behaviour disturbances (attention deficit, hyperactivity), and rare nocturnal focal motor or secondarily generalized seizures or atypical absences. Awake EEG shows focal or multifocal spikes-and-waves predominantly over temporal regions. Sleep EEG shows an activation of interictal EEG abnormalities, and during the course of the syndrome in the acute phase, there may be sub-continuous or continuous spike-wave complexes in NREM(10). However, there is usually a favorable outcome for the seizures and EEG abnormalities. In contrast, the language dysfunction can have a more variable course. Although the language deficits tend to improve with age, a substantial portion of children are left with significant language deficits(11). There is global language deterioration in autistic language regression, but it is usually not very severe, and the comprehension domain tends to be relatively preserved when compared to the expressive domain. On the other hand, stereotypic behaviors and impaired social interaction including pragmatics (the communicative use of language) are more prominent in autistic regression(12,13). In our case, the patient developed clinical seizures one year after the language and social deterioration had started. Autistic epileptiform regression generally begins before 3 years of age, whereas Landau-Kleffner syndrome tends to occur after 3 years of age^(7,8). Abnormal EEG findings such as increased focal and multifocal epileptiform activity during sleep may be seen in both autistic epileptiform regression and Landau-Kleffner syndrome(14). It has been argued that Landau-Kleffner syndrome should be broadened to include children with autistic regression as long as they have epileptiform EEGs during sleep(15,16). Whether such children represent a variant of autistic spectrum disorder or Landau-Kleffner syndrome or a biologically distinct

syndrome remains controversial.

A subgroup of autistic children are diagnosed as childhood disintegrative disorder. These patients would lose previously acquired skills after the age of 2 years, including motor regression and loss of bowel as well as bladder control⁽¹⁷⁾. The prevalence of epilepsy in childhood disintegrative disorder is reported to be as high as 77%⁽¹⁸⁾, and EEG abnormalities are significantly more common in children with childhood disintegrative disorder than those with infantile autism.(19) Our case bears resemblance to childhood disintegrative disorder, except for the discrepancy in the age of regression.

For the patients of autistic regression, improvement in language and social skills have been seen with antiepileptic agents(20,21) and corticosteroids(22). Speech therapy has been used for the management of language regression⁽²³⁾. The use of the other behavioral modes of interventions for such cases, however, has not yet been highlighted adequately in the literature. In addition to pharmacological treatment for epilepsy, a combination of different methods including visual tracking exercises, speech therapy, attention enhancing techniques, and physiotherapy was helpful in our patient. There was significant improvement in all domains, especially in speech which is one of the most disabling features in such cases. A multimodal treatment approach targeting individual deficits appears to be advisable in the management of cases with autistic regression involving several domains in addition to language deterioration.

REFERENCES

- Hoshino Y, Kaneko M, Yashima Y, et al. Clinical features of autistic children with setback course in their infancy. Jpn J Psychiatry Neurol 1987;41:237-45.
- Stefanatos GA. Regression in autistic spectrum disorders. Neuropsychol Rev 2008;18:305-19.
- Rogers SJ, DiLalla DL. Age of symptoms onset in young children with pervasive developmental disorders. J Am Acad Child Adolesc Psychiatry 1990;29:863-72.
- 4. Hrdlicka M. EEG abnormalities, epilepsy and regression in autism: a review. Neuro Endocrinol Lett 2008;29:405-9.
- Volkmar FR, Nelson DS. Seizure disorders in autism. J Am Acad Child Adolesc Psychiatry 1990;29:127-9.

- Rapin I. Autistic regression and disintegrative disorder: how important the role of epilepsy? Semin Pediatr Neurol 1995;2:278-85.
- Tuchman RF, Rapin I. Regression in pervasive developmental disorders: Seizures and epileptiform electroencephalogram correlates. Pediatrics 1997;99:560-6.
- 8. Tuchman R. Acquired epileptiform aphasia. Semin Pediatric Neurol 1997;4:93-101.
- Oslejskova H, Dusek L, Makovska Z, et al. Epilepsia, epileptiform abnormalities, non-righthandedness, hypotonia, and severe decreased IQ are associated with language impairment in autism. Epileptic Disord 2007;Suppl 1:S9-18.
- 10. Hirsch E, Valenti MP, Rudolf G, et al. Landau-Kleffner syndrome is not an eponymic badge of ignorance. Epilepsy Res 2006;70:S239-47.
- 11. Bishop DV. Age of onset and outcome in 'acquired aphasia with convulsive disorder' (Landau-Kleffner syndrome). Dev Med Child Neurol 1985;27:705-12.
- 12. Hall NE. Developmental language disorders. Semin Pediatr Neurol 1997;4:77-85.
- 13. Rapin I, Dunn M. Language disorders in children with autism. Semin Pediatr Neurol 1997;4:86-92.
- Nass R, Gross A, Devinsky O. Autism and autistic epileptiform regression with occipital spikes. Dev Med Child Neurol 1998;40:453-8.
- 15. Echenne B, Cheminal R, Rivier F, et al. Epileptic electroencephalographic abnormalities and developmental dysphasias: a study of 32 patients. Brain Dev 1992;14:216-25.
- 16. Stefanatos GA, Kinsbourne M, Wasserstein J. Acquired epileptiform aphasia: a dimensional view of Landau-Kleffner syndrome and the relation to regressive autistic spectrum disorders. Child Neuropsychol 2002;8:195-228.
- 17. Volkmar FR. Childhood disintegrative disorder: issues for DSM-IV. J Autism Dev Disord 1992;22:625-42.
- Mouridsen SE, Rich B, Isager T. A comparative study of genetic and neurobiological findings in disintegrative psychosis and infantile autism. Psychiatry Clin Neurosci 2000; 54:441-6.
- Kurita H, Kita M, Miyake Y. A comparative study of development and symptoms among disintegrative psychosis and infantile autism with and without speech loss. J Autism Dev Disord 1992;22:175-88.
- 20. Nass R, Petrucha D. Acquired aphasia with convulsive dis-

- order: a pervasive developmental disorder variant. J Child Neurol 1990;5:327-8.
- 21. Plioplys AV. Autism: electroencephalogram abnormalities and clinical improvement with valproic acid. Arch Pediatr Adolesc Med;148:220-2.
- 22. Stefanatos GA, Grover W, Geller E. Case study: corticos-
- teroid treatment of language regression in pervasive developmental disorder. J Am Acad Child Adolesc Psychiatry 1995;34:1107-11.
- 23. Canitano R, Zappella M. Autistic epileptiform regression. Funct Neurol 2006;21:97-101.