Adult Onset Rasmussen's Syndrome Presenting with Psychosis

Theodore Rasmussen and his colleagues first described his eponymous disease in 1958, in three children with a longstanding illness causing focal seizures and worsening damage to one cerebral hemisphere. Rasmussen's encephalitis, also called Rasmussen's syndrome, is a progressive disease characterized by drug-resistant focal epilepsy, progressive hemiplegia, and cognitive decline, with unihemispheric brain atrophy. The syndrome has been divided into three clinical stages. The first is the prodromal stage, with a median duration of seven months (range: 0 months to 8.1 years), a low frequency of seizures and mild hemiparesis. The acute stage, which comes next, has a median duration of eight months and is characterized by frequent seizures. It is accompanied by prominent neurological signs like worsening symptoms of hemiparesis, hemianopia, and cognitive deterioration. The involvement of the dominant hemisphere may lead to aphasia. The last stage is the residual stage with irreversible damage and less frequent seizures than in the acute stage.3

Though considered as an illness of childhood, adult and adolescent patients account for 10% of all cases.4 This report describes the complex case of an adult patient who presented with psychosis as a sequela of Rasmussen's encephalitis.

Case Details
A 32-year-old man came to the outpatient department accompanied by his father, with complaints of withdrawn behavior, unprovoked anger outbursts, occasional smiling to self, poor comprehension, and delayed response for 13 years. According to the informant, the patient had achieved developmental milestones at the appropriate ages. He was reported to have a well-adjusted premorbid personality, has been able to handle all his responsibilities, and was pursuing graduation when the symptoms started. One month prior to the onset of symptoms, the person had a high-grade fever with multiple episodes of sudden onset of abnormal movements of all the four limbs, loss of consciousness, frothing at the mouth and incontinence, for which he had received inpatient treatment. Following discharge, he was prescribed T. Sodium valproate 1500 mg in two divided doses, with which the frequency and severity of the seizures decreased.

The last episode of seizure was eight years back. After five years of remaining seizure free, the antiepileptic medication was tapered and stopped three years back. He also had left sided weakness, which improved gradually but never recovered completely. However, throughout these last 13 years, the patient had been withdrawn, made no verbal communication, and sometimes used gestures to communicate. He was often noticed to be smiling or muttering incomprehensively by himself. He seemed to have poor comprehension of instructions and had a slowing of response. He also had episodes of unprovoked aggression toward family members and outsiders. He had received trials of risperidone and olanzapine in adequate doses in the past, with unsatisfactory response.

He had resting tremors bilaterally. The left upper and lower limbs had hypertonia and hyperreflexia. An equivocal plantar response and ankle clonus were observed on the left side. Romberg's test was positive. There was no aniso-
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senting with psychotic symptoms was made. His score on the Brief Psychiatric Rating Scale was 46.

He was started on T. amisulpride 100 mg twice daily, in view of the predominantly negative symptoms and the inadequate past response to other antipsychotics. During the follow-up after one month of continuing the medication, the family reported some improvement over telephone, including the patient starting to articulate some phrases to communicate with his family members.

The subject of this case study most probably presented in the residual stage of Rasmussen's encephalitis as he exhibited hemiparesis, aphasia, and other neurological symptoms along with certain psychotic symptoms. According to etiopathogenesis, Rasmussen's syndrome may be of three types—antibody-mediated, T-cell cytotoxicity mediated, and microglia-induced degeneration. No laboratory test can be used to confirm the diagnosis of Rasmussen's syndrome. An MRI of the brain in most patients with Rasmussen's syndrome shows a unilateral enlargement of the CSF compartment with maximum accentuation in the insular and peri-insular region, and increased signal intensity in the cortical, subcortical, or both regions. MRI findings in our patient conformed to these usual findings. However, cerebellar involvement was also seen, which is much more unusual in patients with Rasmussen's syndrome. Other investigations like positron emission tomography, single photon emission computed tomography, and magnetic resonance spectroscopy can be used to confirm the unilateral nature of brain abnormalities in suspected Rasmussen's syndrome.

The goals of therapy in acute stage are to decrease the inflammation, restore the functional capacity, and control seizures, and in residual stage, the goal is to restore some degree of functionality. The various modalities of treatment available to achieve these goals include pharmacological, immunotherapeutic, surgical, and rehabilitative techniques. Based on clinical and MRI brain findings, the case reported here fulfilled the European consensus diagnostic criteria for the diagnosis of Rasmussen's syndrome.

coria, abnormality of pupillary reflexes, abnormal extraocular eye movements, papilledema, or neck stiffness. The rest of the central nervous system and systemic examination were within normal limits. On mental status examination, the patient was conscious but remained withdrawn throughout the interview. His attention was aroused with difficulty but not sustained. During the course of the interview, his responses to instructions were inconsistent and there were no verbal responses. His affect was blunted; mood, thought, perception, higher mental functions and judgment could not be assessed. The clinical differential diagnoses considered were intracranial space-occupying lesion like tuberculoma or neurocysticercosis and encephalitis, tuberculous or viral in origin.

An MRI of the brain showed diffuse reduction of volume, with thinning of the cortex and subcortical gliosis in right cerebral parenchyma and prominent sulci and sylvian fissure; gross dilation of right lateral ventricle, and mild shift of midline toward the right side. Volume loss was noted in the right cerebral peduncle (Figure 1), thalamus, and left cerebellar hemisphere, with thinning of folia and left cerebellar peduncle (Figure 2).

With a history of fever and seizures, the aforementioned clinical findings, and findings on the contrast-enhanced MRI of the brain, a diagnosis of Rasmussen's encephalitis in residual stage pre-

FIGURE 1. MRI of the Brain Shows Atrophy of Right Cerebral Hemisphere (Horizontal Arrow) with Dilatation of the Right Lateral Ventricle (Upwards Pointing Arrow)

FIGURE 2. MRI of the Brain Shows Atrophy of Cerebellar Peduncle (Left) As Well As Atrophy of Cerebral Hemisphere (Right)
Conclusion

Rasmussen’s syndrome with adult onset, presenting with psychosis, is a rare entity. As the neurological symptoms were largely absent, management with antipsychotic for the psychiatric symptoms provided some positive results. However, the response to antipsychotic amisulpride observed in this case needs to be further explored.

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Anweshan Ghosh, Prosenjit Ghosh, Madhurima Khasnobish
Silchar Medical College, Silchar, Assam, India

Address for correspondence:
Prosenjit Ghosh, Parijat Apartment, College Road, Ambicapatty, Silchar, Assam 788004, India. E-mail: p_ghosh72@yahoo.com

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