International Journal of Research in Medical Sciences
Suthar PP et al. Int J Res Med Sci. 2014 May;2(2):772-774
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: 10.5455/2320-6012.ijrms20140579

Case Report

Rasmussen's encephalitis in an Indian child

Pokhraj P. Suthar[1]*, Shreyans M. Doshi[2], Prakash J. Rana[1]

[1]Department of Radiology, Government Medical College and S.S.G. Hospital, Vadodara, Gujarat India
[2]M.B.B.S, Government Medical College, Surat, Gujarat, India

Received: 3 March 2014
Accepted: 25 March 2014

***Correspondence:**
Dr. Pokhraj Prakashchandra Suthar,
E-mail: pokhraj_suthar@yahoo.co.in

ABSTRACT

A 5 year old boy with history of recurrent seizures and progressive weakness in left upper & lower limb. On MRI scan atrophy of right cerebral hemisphere with prominent right sided sulci & sylvian fissure. Temporal horn of right lateral ventricle appears prominent. Body and genu of corpus callosum appears hypo plastic. Electroencephalography revealed moderate voltage record showing mixed activity comprising ill-defined 8-10 Hz alpha rhythm plus 4-7Hz waves and random background delta components at 3Hz frequency. A clinical diagnosis of focal tonic-clonic epilepsy was made secondary to Rasmussen's encephalitis, based on the features MRI scan findings of the brain.

Keywords: Paediatrics, Central nervous system, MRI

INTRODUCTION

Rasmussen's encephalitis is a disease of childhood with mean age of presentation is between 6 and 8 years.[1] There is chronic inflammation of the brain, with infiltration of T lymphocytes into the brain tissue. In most cases, affects only one cerebral hemisphere, either the left or the right leading to atrophy of the hemisphere.[2] Focal motor seizures with progressive hemiplegia and progressive motor impairment is most common clinical presentation. The diagnosis of Rasmussen's Encephalitis is made by finding of cerebral hemiatrophy on CT and MRI scans. Patients with adult-onset Rasmussen's encephalitis usually experience a rather mild course and a relatively good long-term outcome.[3]

CASE REPORT

A 5 year old boy presented with two week history of recurrent seizures and progressive weakness in left upper & lower limb. Seizure was focal tonic-clonic type, each of which lasted around 2 minutes. Seizure was sudden in onset and frequency was about 2-3 for the day, occurs on alternate days and with consistency of nature. There was no preceding aura, automatic behaviour, or associated loss of consciousness. Post-ictal sleep does not occur, but there was weakness after seizures which last for some minutes. There was no loss of continence. Antenatal, perinatal and postnatal histories were normal. No history of fever or previous tuberculosis. Patient had past history of convulsion and taking medicine for it. There was a delayed progression of developmental mile stone. On examination no dysmorphic facial features. Vision and hearing were intact but speech is incoherent and his words were not clear. Other cranial nerves were normal. Power was grade 3 on the left upper limb and 4 on the left lower limb, but a full 5 on the right upper and lower limbs. The muscle tone and tendon reflexes were increased on the left side. The cardiovascular, respiratory and gastro-intestinal systems were normal. On routine investigation haemoglobin 10.1 gm%, total WBC count 8300 cell/cu.mm, ESR was within normal limit. Chest X-ray is normal. CSF examination was normal.

On Magnetic Resonance Imaging (MRI) scan atrophy of right cerebral hemisphere with prominent right sided

sulci & sylvian fissure. Temporal horn of right lateral ventricle appears prominent as compare to left side (Figure 1, 2, and 3). Body and genu of corpus callosum appears hypo plastic (Figure 4). However splenium and rostrum of corpus callosum appears normal. No evidence of any abnormal signal intensity lesion or restricted diffusion in cerebral white matter.

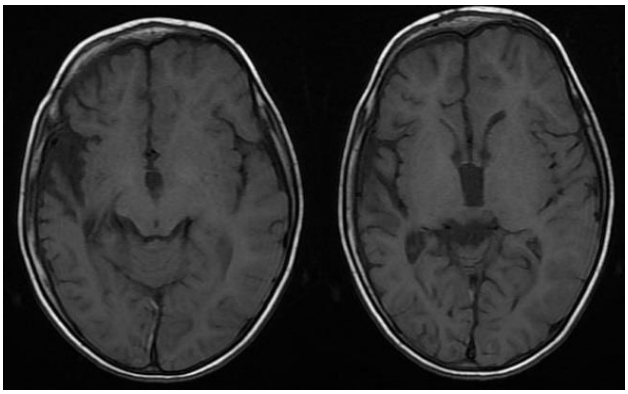

Figure 1: Rasmussen's encephalitis. Axial T1 MRI scan of the brain showing dilated right sylvian fissure, right cerebral sulci and right sided cerebral atrophy.

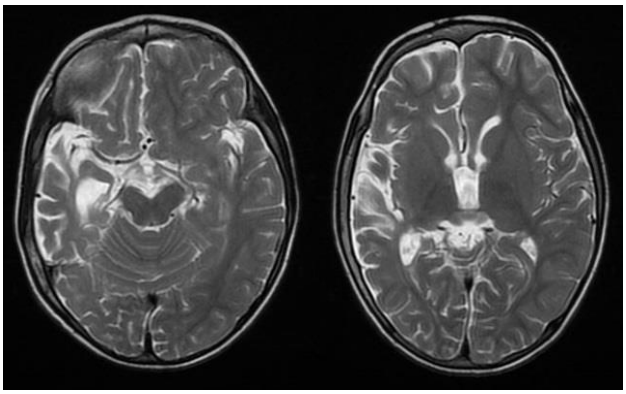

Figure 2: Rasmussen's encephalitis. Axial FSE T2 MRI scan of the brain showing dilated right temporal horn of lateral ventricle, right cerebral sulci and right sided cerebral atrophy.

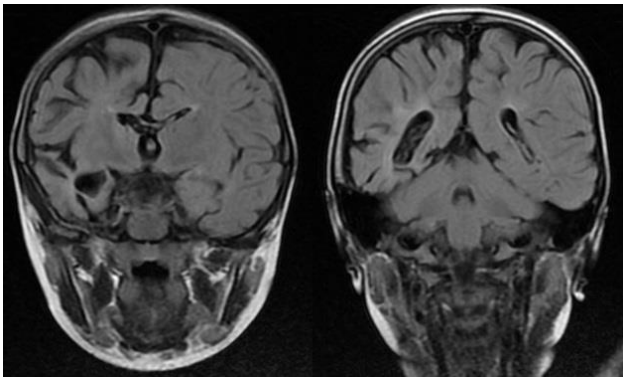

Figure 3: Rasmussen's encephalitis. Coronal T1 FLAIR MRI scan of the brain showing dilated sulci, right lateral ventricle and cerebral hemiatrophy.

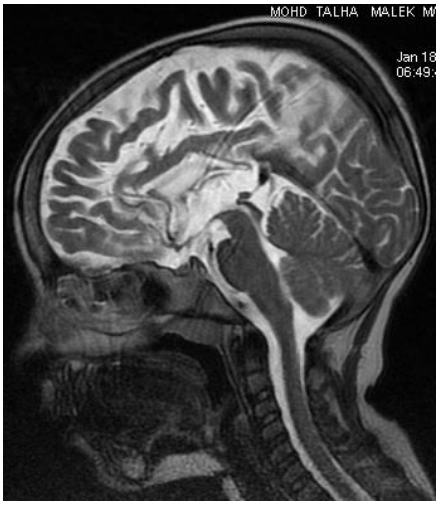

Figure 4: Rasmussen's encephalitis. Sagittal FSE T2 MRI scan of the brain showing hypo plastic genu and body of corpus callosum.

Electroencephalography revealed moderate voltage record showing mixed activity comprising ill-defined 8-10Hz alpha rhythm plus 4-7Hz waves and random background delta components at 3Hz frequency. The alpha waves were symmetrical in distribution, better formed and of higher amplitude on the right than the left hemisphere.

A clinical diagnosis of focal tonic-clonic epilepsy was made secondary to Rasmussen's encephalitis, based on the features MRI scan findings of the brain. Patient is currently on outpatient with anticonvulsant drug therapy.

DISCUSSION

Rasmussen's encephalitis is a disease of childhood. The mean age at presentation is between 6 and 8 years, and these children typically have had a normal course of neurologic development.[1] Rasmussen's encephalitis is a chronic, progressive inflammation of the brain of unknown origin. Recent research suggests a possible viral origin or a viral-induced autoimmune mechanism.[4] There is chronic inflammation of the brain, with infiltration of T lymphocytes into the brain tissue. In most cases, this affects only one cerebral hemisphere, either the left or the right. This inflammation causes permanent damage to the cells of the brain, leading to atrophy of the hemisphere; the epilepsy that causes may itself contribute to the brain damage. The epilepsy might derive from a disturbed GABA release.[2] Although viral etiology is suspected, it is suggested that it might be autoimmune disease arising from autoantibodies to glutamate receptors.[4] They present with focal motor seizures although generalised also have been noted. It can be progress to hemiplegia, cognitive impairment and progressive motor impairment. CSF exam in most cases are normal but sometimes show increased total protein, Ig G index and oligoclonal band.[5]

The diagnosis of Rasmussen's encephalitis was facilitated in this case by the finding of cerebral hemiatrophy on CT and MRI scans, in the clinical setting of hemiparesis and intractable seizure disorder. MRI has been shown to demonstrate the progression of Rasmussen's encephalitis and may suggest the diagnosis in the early stages, often before the appearance of neurological deficits.[6] Apart from atrophy of the head of the caudate nucleus; MRI may also show associated secondary changes such as atrophy of the contralateral cerebellar hemisphere, the ipsilateral hippocampus, and the brainstem.[6] There are four recognized stages of Rasmussen's encephalitis based on T2 weighted MRI criteria.[7] These are swelling with hyperintense signal (stage 1); normal volume with hyperintense signal (stage 2); atrophy with hyperintense signal (stage 3); and progressive atrophy and normal signal (stage 4).[7] Our patient presented in stage 4. Ictal Single Photon Emission Computed Tomography (SPECT) is useful for the localization of the epileptogenic focus in the respective cerebral hemisphere in RE patients.[8] The use of SPECT in this case report would have assisted us in localizing the epileptogenic focus, but this facility is not available in our centre. The age at presentation in our case report is close to the median age at disease onset of 6 years as reported by Ramesha KN et al. in 2009.[9] Our patient presented with hemiparesis, focal tonic-clonic seizures, incoherent speech, and hemiatrophy of the left upper and lower limb. Radiologically, RE should be differentiated from other causes of cerebral hemiatrophy like Sturge-Weber syndrome, Dyke-Davidoff-Masson, and hemimegalencephaly. Treatment of children with Rasmussen's encephalitis has thus far been disappointing. Surgical management with hemispherectomy has been the only successful alternative as measured by seizure eradication and prevention of further deterioration in cognition. Permanent hemiparesis is an inevitable consequence, however, and it is therefore essential to document a moderate hemiparesis prior to surgery.

Patients with adult-onset Rasmussen's encephalitis usually experience a rather mild course and a relatively good long-term outcome.[3]

ACKNOWLEDGEMENTS

The authors would like to acknowledge the help of department of radiology, Shree Sayaji General Hospital, Vadodara.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Honavar M, Janota I, Polkey CE. Rasmussen's encephalitis insurgery for epilepsy. Dev Med Child Neurol. 1992;34:3-14.
2. Rassner Michael P, van Velthoven-Wurster Vera, Ramantani Georgia, Feuerstein Thomas J. (Altered transporter-mediated neocortical GABA release in Rasmussen's encephalitis. Epilepsia. 2013 Mar;54(3):e41-4.
3. Bien CG, Widman G, Urbach H et al. The natural history of Rasmussen's encephalitis. Brain. 2002;125:1751-9.
4. Rogers SW, Andrews PI, Gahring LC et al. Autoantibodies to glutamate receptor $GluR_3$ in Rasmussen's encephalitis. Sci. 1994;265:648-51.
5. Fokuda T, Oguni H, Vanagaki S et al. Chronic Localised encephalitis preceded by Ipsilateral uveitis: a case report. Epilepsia. 1994;35:1328-31.
6. Chiapparini L, Granata T, Farina L, Ciceri E, Erbetta A, Ragona F, Freri E, Fusco L, Gobbi G, Capovilla G, Tassi L, Giordano L, Viri M, Dalla Bernardina B, Spreafico R, Savoiardo M. Diagnostic imaging in 13 cases of Rasmussen's encephalitis: can early MRI suggest the diagnosis? Neuroradiol. 2003;45(3):171-83.
7. Osborne AG. Rasmussen's encephalitis. In: Osborne AG, Blaser SI, Salzman KL, Katzman GL, Provenzale J, Castillo M, Hedlund GL, Illner A, Harnsberger HR, Cooper JA, Jones BV, Hamilton BE, eds. Diagnostic Imaging Brain. 1st ed. Utah: Amirsys Inc; 2005: 42-45.
8. Burneo JG, Hamilton M, Vezina W, Parrent A. Utility of Ictal SPECT in the presurgical evaluation of Rasmussen's encephalitis. Can J Neurol Sci. 2006;33(1):107-10.
9. Ramesha KN, Rajesh B, Ashalatha R, Kesavadas C, Abraham M, Radhakrishnan VV, Sarma PS, Radhakrishnan K. Rasmussen's encephalitis: experience from a developing country based on a group of medically and surgically treated patients. Seizure. 2009;18(8):567-72.

DOI: 10.5455/2320-6012.ijrms20140579
Cite this article as: Suthar PP, Doshi SM, Rana PJ. Rasmussen's encephalitis in an Indian child. Int J Res Med Sci 2014;2:772-4.