MRI brain showed hyperintense lesions in T2 and FLAIR images, involving the right half of the brainstem, the right cerebellum, and the posterior limb of the right internal capsule. These lesions were isointense on T1 and did not show any diffusion restriction. The right half of the brainstem was swollen. No significant contrast enhancement was noted [Figures 1-4].

A 30-year-old male presented with acute-onset headache, vomiting, and facial deviation to the left. He had had an upper respiratory infection 1 week earlier that had subsided within 2 days. Clinical examination showed right gaze palsy, right lower motor neuron (LMN) facial palsy, and minimal right cerebellar signs. Fundus examination was normal and meningeal signs were absent.

Images in Neurology

Hemi-capsulo-rhombencephalic demyelination

C. J. Suresh Chandran, V. Maheshwaran1, Madhavan Unni1
Departments of Neurology and 1Radiology, Kerala Institute of Medical Sciences, Trivandrum, Kerala, India

For correspondence:
Dr. C. J. Suresh Chandran, Kerala Institute of Medical Sciences, Trivandrum, Kerala, India. E-mail: drceejay@rediffmail.com

Ann Indian Acad Neurol 2010;13:150-1 [DOI: 10.4103/0972-2327.64631]

Figure 1: Axial T2 image showing hyperintensity in the right pons and cerebellum

Figure 2: Axial T2 image showing hyperintensity in the right half of the midbrain. Note that the right side of the midbrain is swollen

Figure 3: Axial FLAIR image showing hyperintensity of the posterior limb of the right internal capsule

Figure 4: Axial T1 contrast image showing a swollen right half of pons. The lesions are isointense on T1, without any enhancement. Note the compression of the fourth ventricle from the right side
MR angiography and MRI spine were normal. Cerebrospinal fluid (CSF) study showed elevated protein (90 mg/dl), normal sugar, and 5 lymphocytes/mm³ CSF oligoclonal band, Indian ink staining, and HSV PCR were negative. Gram's stain did not yield any organisms. Vasculitic workup, HIV test, and serum and CSF VDRL were negative. We diagnosed acute disseminated encephalomyelitis (ADEM) – right hemi-capsulo-rhombencephalic demyelination. The patient was treated with intravenous methyl prednisolone 1 gm once daily for 3 days, followed by oral prednisolone 1 mg/kg for 2 weeks. The neurological deficits resolved in 1 week. Repeat MRI done 4 weeks later showed resolution of the lesions [Figure 5].

Figure 5: MRI (T2 axial) done 4 weeks after presentation showing resolution of the lesions

To the best of our knowledge, no cases of unilateral demyelination involving the brainstem and internal capsule have been reported earlier. Our case of hemi-capsulo-rhombencephalic demyelination is thus a unique case of site-restricted ADEM.

References

1. Murthy JM. Acute disseminated encephalomyelitis. Neurol India 2002;50:238-43.
2. Armstrong RW, Fung PC. Brainstem encephalitis (rhombencephalitis) due to Listeria monocytogenes: Case report and review. Clin Infect Dis 1993;16:689-702.
3. Garcia-Cazorla A, Olivan JA, Pancho C, Sans A, Boix C, Campistol J. Infectious acute hemicerebellitis. J Child Neurol 2004;19:390-2.
4. Jabbour P, Samaha E, Abi Lahoud G, Koussa S, Abadjian G, Nohra G, et al. Hemicerebellitis mimicking a tumor on MRI. Childs Nerv Syst 2003;19:122-5.
5. Singh S, Alexander M, Sase N, Korah IP. Solitary hemispheric demyelination in acute disseminated encephalomyelitis: Clinicoradiological correlation. Australas Radiol 2003;47:29-36.
6. Sackey AH, Brodhead RL. Hemi-paresis after measles, mumps, and rubella vaccination. BMJ 1993;306:1169.
7. Pradhan S, Pandey N. Acute disseminated encephalomyelitis presenting as ataxic hemiparesis. Neurol India 1998;46:454-7.
8. Bien CG, Granata T, Antozzi C, Cross JH, Dulec O, Kurthen M, et al. Pathogenesis, diagnosis and treatment of Rasmussen encephalitis: A European consensus statement. Brain 2005;128:454-71.
9. Aviv RI, Benseler SM, Silverman ED, Tyrell PN, Deveber G, Tsang LM, et al. MR imaging and angiography of primary CNS vasculitis of childhood. AJNR Am J Neuroradiol 2006;27:192-9.
10. Eidelberg D, Sotrel A, Horoupian DS, Neumann PE, Pumarola-Sune T, Price RW. Thrombotic cerebral vasculopathy associated with herpes zoster. Ann Neurol 1986;19:7-14.

Unilateral involvement is rare in ADEM. There have been rare reports of hemicerebellitis in children and solitary hemispheric tumefactive demyelinating lesions have been reported.[3–5] ADEM presenting as hemiplegia and ataxic hemiparesis have been reported, but MRI shows bilateral involvement in these cases.[6,7] Inflammatory disorders with unilateral brain involvement include Rasmussen's encephalitis, primary angiitis of the central nervous system, and herpes zoster–related vasculopathy. Rasmussen's encephalitis is an inflammatory immune-mediated brain disorder characterized by unilateral hemispheric atrophy, intractable seizures, and progressive neurological dysfunction. MRI features of Rasmussen's encephalitis include unilateral enlargement of CSF compartments (most accentuated in the insular and perinsular regions), with increased cortical and/or subcortical T2 and FLAIR signals and caudate head atrophy.[8] In primary angiitis of the central nervous system the common pattern of parenchymal involvement is multifocal, unilateral, proximal lesions in the anterior circulation.[9] Herpes zoster–related vasculopathy also shows unilateral lesions in the anterior or middle cerebral artery territory.[10]