Isolated Medullary Tumefactive Demyelination Masquerading as Tumor

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A 29-year-old lady presented to our department with complaints of vertigo for the past 1.5 months, ataxia for 1 month with right-sided sensorimotor weakness. On examination, the left-sided ataxia with the left 6th and 7th nerve palsy were identified.

The first magnetic resonance imaging (MRI) was done outside after 15 days of onset of symptoms and showed T2 and Fluid-attenuated inversion recovery (FLAIR) hyperintense expansile lesion in the left half of the medulla, inferior aspect of the pons, and cervicomedullary junction, showing incomplete horseshoe (incomplete ring?) enhancement on the post-contrast study [Figure 1]. Restricted diffusion was seen in the medial aspect of the lesion and no blooming on Gradient echo (GRE). There was no other site of abnormal signal intensity on FLAIR. The outside MRI was reported as brainstem glioma. Repeat imaging after 1 month in our department showed an increase in the extent of signal intensity and contrast enhancement [Figure 2a–d]. Considering the pattern of FLAIR hyperintensity with an incomplete horseshoe pattern of enhancement, demyelination possibility was kept ahead of brainstem glioma. With these symptoms and imaging findings, the patient was started on steroids in view of demyelination. A follow-up scan performed after 30 days showed a significant decrease in the size and extent of the hyperintensity and contrast enhancement [Figure 2e and f]. A new lesion was seen in the dorsal cord. The patient was completely asymptomatic during the second follow-up scan.

Discussion

Isolated tumefactive demyelination lesion (TDL) of the medulla is a rare entity.[1] TDLs are seen more frequently in the second and third decades of life[2] with the female to male ratio being 4:1. The location of TDLs can vary, but studies report them to be more frequent in the supratentorial

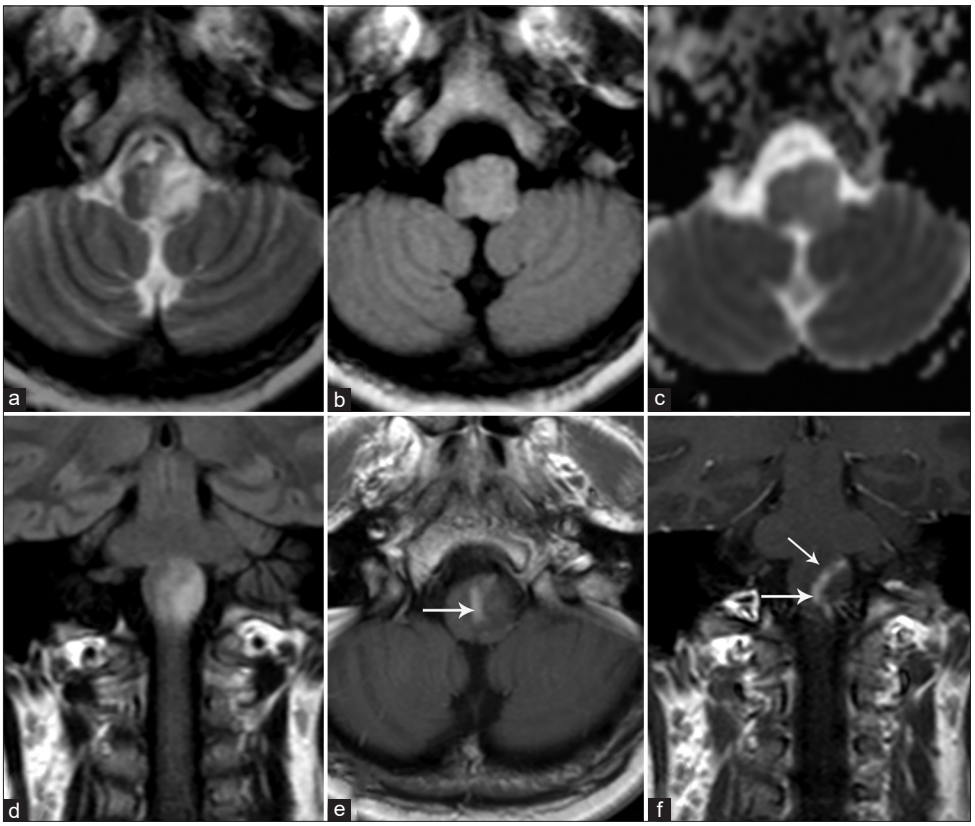

Figure 1: A well-defined T2 (a) and FLAIR (b and d) hyperintense expansile lesion in the left medulla without diffusion restriction (c) Inner margin enhances in the post-gad axial (e) and coronal (f) images

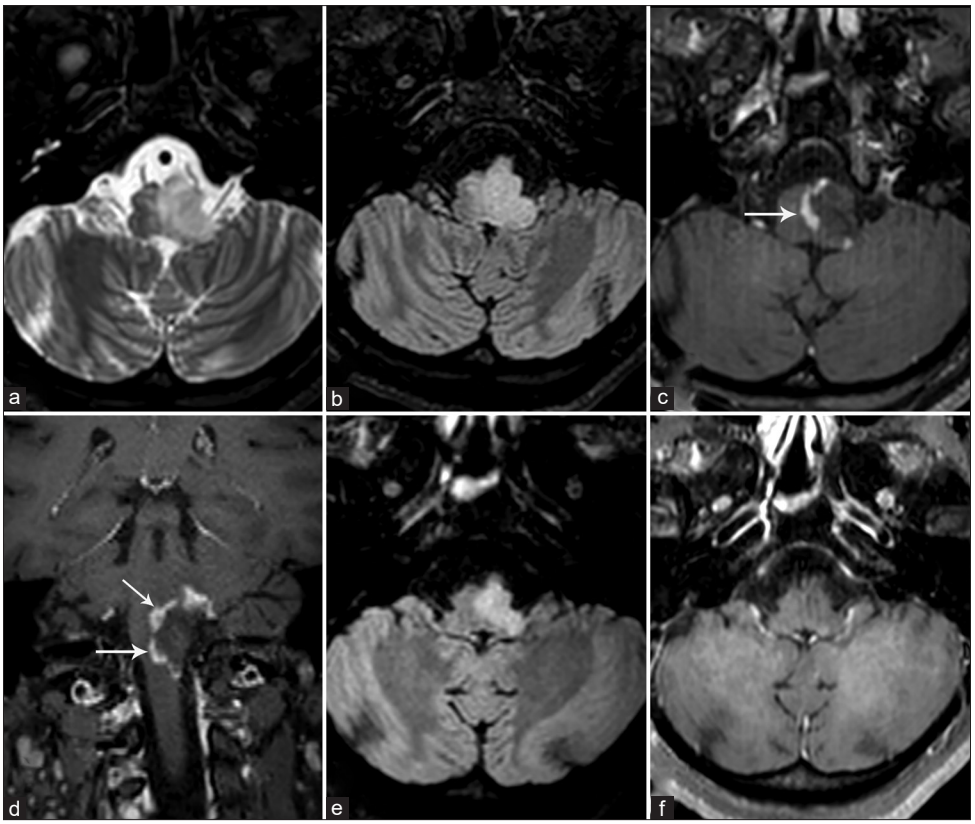

Figure 2: Interval MRI shows increase in size and enhancement of lesion (a–d) Post-treatment MRI reveals regression in size and enhancement (e–f)

region, mostly in the frontal and parietal lobes[3] with rare involvement of the corpus callosum, the brainstem, or the spinal cord.

The most specific enhancement pattern is the incomplete ring, typically open toward the gray matter, but almost any enhancement pattern can be found (complete-ring, heterogeneous, homogeneous, nodular, punctate, patchy) and up to 5% might not show any contrast uptake.[3] Incomplete ring-enhancement gives clue to the diagnosis in the present case.[4]

Biopsy is considered to be the gold standard diagnostic test for these lesions. The procedure might not be feasible in all of them due to technical or anatomical issues like brainstem lesions. With this typical pattern of enhancement, a life-risking biopsy can be avoided, especially in the brainstem and a close follow-up scan is needed with the treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Rovira À, Sastre-Garriga J, Auger C, Rovira A. Idiopathic inflammatory demyelinating diseases of the brainstem. Semin Ultrasound CT MR 2013;34:123-30.
2. Lucchinetti CF, Gavrilova RH, Metz I, Parisi JE, Scheithauer BW, Weigand S, *et al*. Clinical and radiographic spectrum of pathologically confirmed tumefactive multiple sclerosis. Brain 2008;131:1759-75.
3. Sanchez P, Meca-Lallana V, Barbosa A, Manzanares R, Palmí I, Vivancos J. Tumefactive demyelinating lesions of 15 patients: Clinico-radiological features, management and review of the literature. J Neurol Sci 2017;381:32-8.
4. Suh CH, Kim HS, Jung SC, Choi CG, Kim SJ. MRI findings in tumefactive demyelinating lesions: A systematic review and meta-analysis. AJNR Am J Neuroradiol 2018;39:1643-9.

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Received: 24-Mar-2021 **Revised:** 15-May-2021 **Accepted:** 11-Jun-2021
Published: 20-Apr-2022

DOI: 10.4103/aian.AIAN_250_21