Adrenoleukodystrophy-Like Presentation of MOG-Antibody -Associated Demyelination

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Case Description

A nine-year-old boy presented with sub-acute onset of visual symptoms, inability to recognize objects, and walking difficulty that evolved over 2-3 weeks. At the onset of the illness, the patient's mother noticed he was not making eye contact while speaking and could not see distant objects. He sometimes was caught bumping into furniture at home when walking. He did not have headaches, fever, neck pain, seizures, or alteration in consciousness. His best corrected visual acuity was 6/18, pupils were reactive, and fundi were normal. No other neurological deficits were observed on clinical examination. MRI images at the onset of visual symptoms have been depicted in Figure 1a-c. A possibility of adrenoleukodystrophy was considered at another center and was subjected to genetic testing and referred. Exome sequencing was negative. Further evaluation revealed strong seropositivity for anti-myelin oligodendrocyte glycoprotein antibodies. Anti-nuclear antibodies, ESR, CRP, and HIV serology were negative. CSF studies were within normal limits. Visual evoked potentials showed prolonged P100 latencies (150 ms in both eyes). He was treated with steroids, and follow-up MRI imaging showed near complete resolution [Figure 1d-f]. Myelin oligodendrocyte glycoprotein-IgG associated disease (MOGAD) may have atypical presentations and neuroimaging findings.[1]

Discussion

This case highlights an unusual and novel presentation of symmetrical parieto-occipital demyelination in MOGAD.

In addition to common phenotypes of MOGAD, such as optic neuritis, acute disseminated encephalomyelitis (ADEM), and myelitis, rarer manifestations include aseptic meningitis[2] and

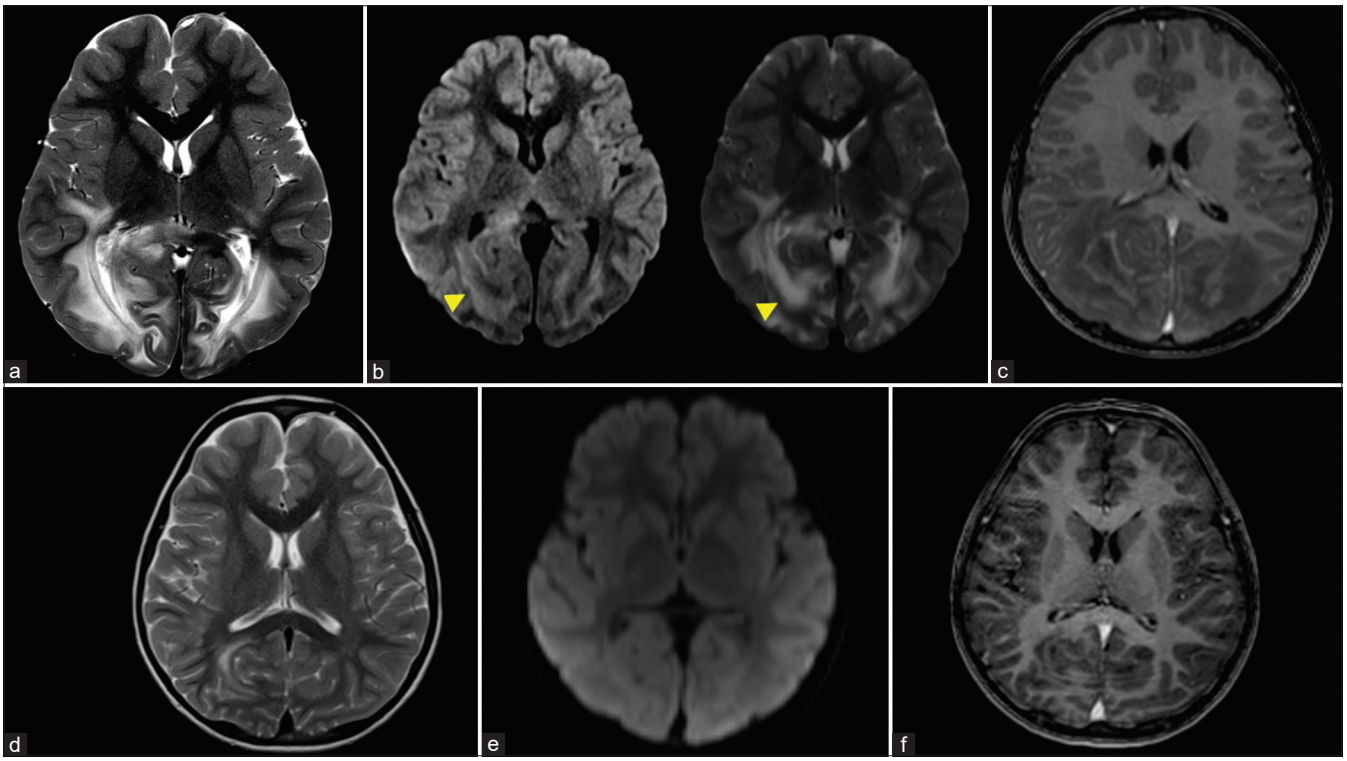

Figure 1: (a) T2-axial image showing bilateral symmetrical bilateral confluent involvement of parieto-occipital periventricular and subcortical white matter with extension into the splenium of the corpus callosum. (b) Diffusion restriction brightness (1st panel b) and T1-contrast enhancement (c) were observed at the edge of the lesion, similar to ALD. Note that the areas of diffusion restriction are hypointense on ADC (marked by yellow arrows). (d-f) Follow-up MRI after three months showing near complete resolution of the lesion. ALD: Adrenoleukodystrophy, ADC: Apparent diffusion coefficient

focal cortical lesions.[3] Confluent and bilateral involvement of the white matter in MOGAD has been referred to as the leukodystrophy-like (LD) variant.[4] In a series of 7 children who presented with the LD variant, subacute encephalopathy was present in nearly all the cases.[4] Two children had a leukodystrophy-like MRI at presentation, and the remaining developed such features in later relapses. Nodular contrast enhancement was observed in a few in that case series. Interestingly, MRI features may not always correlate with the clinical condition. It has also been observed that this variant tends to respond to immunotherapy poorly. In our case, the patient's initial MRI findings showed diffuse white matter abnormalities, particularly in the parieto-occipital regions, with contrast enhancement and diffusion restriction resembling typical Adrenoleukodystrophy (ALD). Such imaging abnormalities in MOGAD are hitherto undescribed. Conspicuously, our patient did not present with features of encephalopathy as described in the earlier study by Hacohen *et al*.[4]

By analyzing the structures involved, patterns of restriction, presence of enhancement, and peaks on spectroscopy, one could differentiate the various leukoencephalopathies that involve the posterior cerebral region. Krabbe's disease is a well-known leukodystrophy affecting the parieto-occipital region. T2 hyperintensities are usually seen in the periventricular and subcortical areas, sparing the U-fibers. Internal capsule, splenium, and pyramidal tracts in the midbrain and pons are typically involved, and diffusion restriction/contrast enhancement of affected structures is absent. Late-infantile forms of metachromatic leukodystrophy, AARS2-related leukoencephalopathy, and adult-onset polyglucosan bodies are other rare genetically mediated causes of posterior leukoencephalopathies. Leukoencephalopathy with brainstem involvement and elevated lactate (LBSL) is another rare mitochondrial disorder that can involve the posterior structures. The patient was treated with monthly intravenous methylprednisolone (30 mg/kg × 5 days). His visual symptoms resolved entirely, and his visual acuity was normal after steroid therapy. Follow-up MRI imaging after three months showed near complete resolution of the white matter abnormalities, reflecting the patient's response to steroid treatment. Resolution of MRI abnormalities and improvement in the clinical condition further support the possibility of an immune-mediated disease.

This case demonstrates the importance of considering MOGAD in the differential diagnosis of patients with atypical neurological presentations. Early diagnosis and treatment of MOGAD can significantly improve clinical outcomes.

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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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