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A RARE CASE OF PAEDIATRIC DEVIC SYNDROME

Radio-Diagnosis

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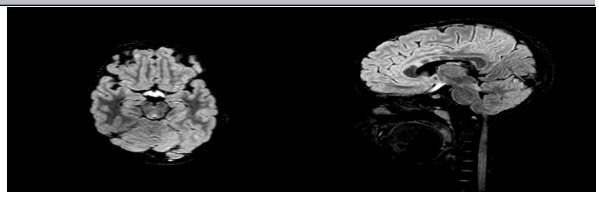
ABSTRACT

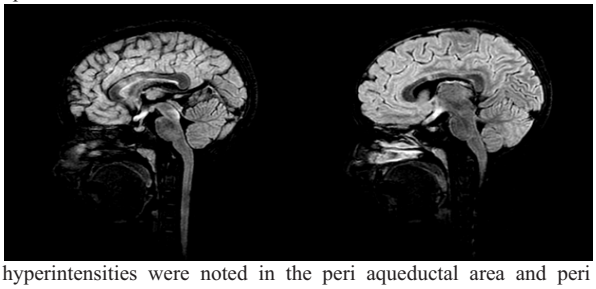
Aquaporin-4 (AQP4) water channels are the main target antigens in neuromyelitis optica (NMO), an autoimmune demyelinating disorder. New insights into the pathogenesis, clinical presentation, and imaging patterns of NMO have been gained. The breakthrough came with identifying the AQP4 antibody, which recognises only this condition. The spectrum of NMO has expanded as new clinical and radiologic features in seropositive patients have been described, and the term NMO spectrum disorder (NMOSD) has been adopted to describe this phenomenon. The list of possible diagnoses that now includes NMOSD continues to grow. Early NMOSD diagnosis, especially for unusual clinical manifestations of this demyelinating disease, has been greatly aided by the recognition of NMOSD imaging patterns and their mimicry of disorders. This image-based review compiles key information about NMOSD, including its history, pathophysiology, clinical manifestations, and differential diagnosis across a wide range of imaging modalities.

Learning And Objectives :
Optic neuritis is a possible differential diagnosis for patients who have experienced optic nerve involvement. Individuals with aquaporin-4-IgG-positive NMOSD typically have longer-segment, less prominent involvement of the optic nerves, and more frequently involve the chiasm and optic tracts, in contrast to patients with myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD), which can present very similarly. The differential diagnosis for patients with cerebral involvement is extensive and includes multiple sclerosis, Susac, and ADEM. A longitudinally extensive spinal cord lesion is a differential diagnosis for patients with spinal cord involvement. An acute spinal cord ischemic lesion is a differential from the typical bright spotty lesion.

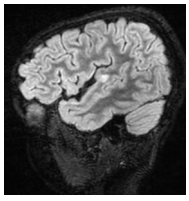
Materials And Methods :
A female child aged 13 years came with bilateral transient loss of vision for 5 hours MRI was done using a SIGNA™ 3.0T MRI Scanner



T2 FLAIR hyperintensities in bilateral optic chiasma and proximal optic tracts.



hyperintensities were noted in the peri aqueductal area and peri ependymal surface of the corpus callosum along the callosum-septal interface.



An ill-defined T2 FLAIR hyperintense lesion in the posterior aspect of the superior temporal gyrus in the juxta cortical location.

KEYWORDS

INTRODUCTION:
Neuromyelitis Optica Spectrum Disorder (NMO), also known as Devic Syndrome, is an extremely debilitating form of acute demyelinating disease that primarily affects the spinal cord and optic nerves, with only mild involvement of the brain's white matter.

T2/FLAIR hyperintensity in the optic chiasma, periaqueductal grey matter, periventricular (hemispheric) confluent smooth sessile white matter involvement, and spinal cord involvement extending more than three vertebral body lengths are the most significant imaging findings.

The autoimmune inflammatory demyelinating disease neuromyelitis optica (NMO) affects the central nervous system (CNS) and can be debilitating. In 2004, aquaporin-4 (AQP4)-immunoglobulin G (IgG), an antibody against the astrocyte water channel, was discovered, shedding light on the role of autoimmunity in the etiopathogenesis of NMO. This led to the identification of NMO as an autoimmune condition associated with water channels.

In light of AQP4-IgG's high specificity, a wider range of NMO-related clinical and radiologic features have been identified. Patients with AQP4-IgG positivity have also been reported to have lesions in the diencephalon, brainstem, and cerebral hemispheres. This suggests that CNS involvement is not limited to the optic nerves and spinal cord. The term NMO spectrum disorder (NMOSD) has been adopted to describe the collection of overlapping autoimmune diseases found in the seropositive population.

RESULTS :
- T2 FLAIR hyperintensities in bilateral optic chiasma and proximal optic tracts.
- Similar hyperintensities were noted in the peri aqueductal area and peri ependymal surface of the corpus callosum along the callosum-septal interface.
- There is, in addition, an ill-defined T2 FLAIR hyperintense lesion in the posterior aspect of the superior temporal gyrus in the juxta cortical location.

CONCLUSION:
The average age of onset for neuromyelitis optica patients is 41 years old, making them slightly older than MS patients, and there is an even stronger female predilection (F: M 6.5:1) than there is for MS. Patients

of Asian, Indian, and African descent have higher rates of this condition.

It's important to note that none of the characteristics that set NMO apart from MS can be used to diagnose the former disease on their own. It's also worth noting that there may be a higher rate of overlap between the imaging findings of these two entities in Asian populations.

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