

A Rare Case of Asymptomatic Cavernous Sinus Thrombosis in Neuromyelitis Optica Spectrum Disorder

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Abstract

Neuromyelitis optica spectrum disorder (NMOSD) is an inflammatory disease of the central nervous system that includes transverse myelitis, optic neuritis, acute brainstem syndrome, area postrema syndrome, symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic lesions on magnetic resonance imaging, and symptomatic cerebral syndrome with NMOSD-typical brain lesions. NMOSD has been associated with autoimmune conditions and rare cases of venous thrombosis. A 60-year-old woman who presented with recurrent optic neuritis and transverse myelitis was diagnosed with aquaporin 4 antibody-positive NMOSD. She presented with blurring of vision and was found to have right Optic Neuritis and incidental left cavernous sinus thrombosis. She was treated with methylprednisolone and improvement in symptoms were noted. To our knowledge, this is the first documented case of cavernous sinus thrombosis in NMOSD.

Key Words: Neuromyelitis Optica Spectrum Disorder; Cavernous Sinus Syndrome; Venous Thrombosis; Autoimmune; Demyelinating

Abbreviations: Neuromyelitis Optica Syndrome Disorder (NMOSD); Magnetic Resonance Imaging (MRI) If there are any abbreviations in the article they have to be mentioned

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Introduction

Neuromyelitis optica spectrum disorder (NMOSD) is an inflammatory disease of the central nervous system. [1] The International Panel for NMO Diagnosis revised previous diagnostic criteria for neuromyelitis optica with more stringent criteria to create the unifying term NMOSD. It is classically characterized by involvement of the optic nerve, spinal cord and brain. NMOSD has been associated with various autoimmune diseases such as rheumatoid arthritis, Sjögren's syndrome, systemic lupus erythematosus, thyroid disease, myasthenia gravis, ulcerative colitis, antiphospholipid syndrome and autoantibodies without diseases. [2] The pathophysiology and mechanism of these associations have not been recognized. However, there have been no reports of cavernous sinus thrombosis to date. Cavernous sinus syndrome consists of multiple cranial nerve involvement, Horner's syndrome, ophthalmoplegia, orbital congestion, proptosis, trigeminal

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sensory loss and sympathetic impairment. [3,4]. However, patients can be asymptomatic and may not fulfil the criteria. We report a rare case of cavernous sinus thrombosis in a patient with NMOSD.

Case Report

A 60 year-old lady, a restaurant manager by profession, had recurrent episodes of optic neuritis and transverse myelitis. Investigations revealed she was aquaporin 4 antibody positive with magnetic resonance imaging (MRI) suggestive of demyelinating disease. She was given azathioprine but could not tolerate the drug due to neutropenia and liver impairment. There have been treatment delays due to an earlier misdiagnosis of multiple sclerosis and an unfortunate situation with a renal abscess that required the prolonged use of antibiotics. Her mobility has been restricted due to recurrent transverse myelitis. Interferon therapy was initiated in September 2013 and continued until May 2014. There was one episode each of urinary tract infection and right perinephric abscess in May of 2014 and August of 2014, respectively.

Azathioprine was given from March 2014 until May 2016. In March 2016 she presented with blurring of vision and double vision. Clinical examination revealed a relative afferent pupillary defect over the right eye. Vision over the right eye revealed no perception to light and the left eye acuity was 6/15. Ishihara's colour vision test revealed deficiencies in both eyes with scores of 0/15 on the right and 1/15 on the left. Colour vision could not be assessed on the right side but fundoscopy revealed pale optic discs bilaterally; eye movements were full and there was no evidence of ophthalmoplegia. There was no speech or swallowing difficulty, tone was normal, power in the upper limbs was 5/5 bilaterally and reflexes were normal. Lower examination revealed normal tone, 4/5 power bilaterally and brisk reflexes. Plantar reflex showed Babinski's sign.

She was treated with intravenous methylprednisolone for acute optic neuritis. A computerized tomography scan revealed an incidental fullness of the left cavernous sinus region. MRI confirmed bulky cavernous sinus in the left and enhancing intracanalicular right optic nerves slightly compressed by dural thickening-Figure 1 (A and B). Intraorbital parts of the optic nerve were normal. Both optic nerves showed nerve sheath enhancement more on the right side, which also indicates prechiasmatic involvement. A repeat MRI upon completion of steroid treatment revealed resolved bulkiness of cavernous sinus region. Figure 2(A and B) There were no clinical or radiological features suggestive of central nervous system tumours such as meningioma, neurofibroma, nasopharyngeal carcinoma or metastatic lesions. No evidence of carotico-cavernous fistula or cavernous sinus aneurysm was seen. The patient did not have clinical features suggestive of connective tissue disease and there was no other evidence of infective cause. She declined lumbar puncture and cerebral venous study for further evaluation

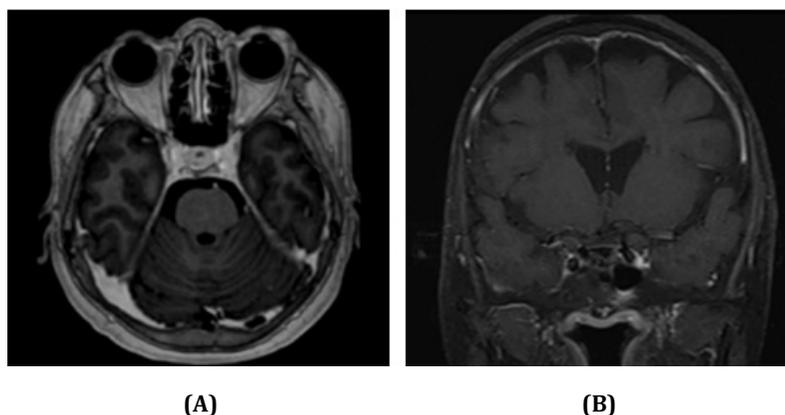


Figure 1: Initial pretreatment (25/3/2016) T1-post-gadolinium magnetic resonance imaging in axial (A) and coronal (B) planes demonstrate a bulky and thickened wall of the left cavernous sinus.

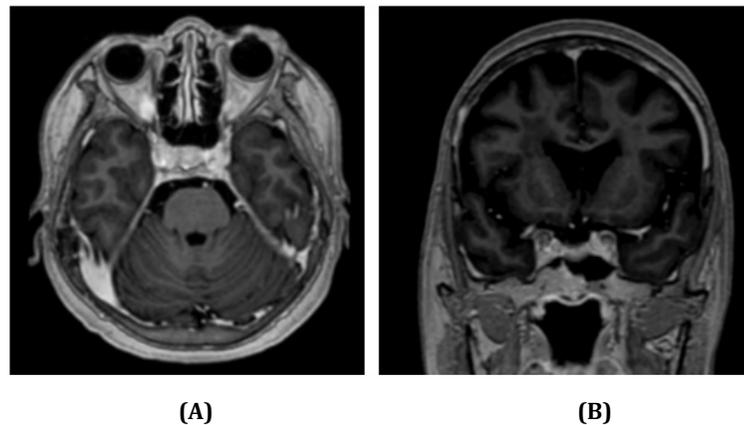


Figure 2: Post-treatment (26/4/16) T1-post gadolinium magnetic resonance imaging in axial (A) and coronal (B) planes; the left cavernous sinus wall thickening has been reduced in size.

Discussion

NMOSD is a rare immune-mediated disease which has evolved in recent years in terms of diagnosis, clinical description and disease associations. Several other conditions have been associated with NMOSD. There has been no association with cavernous sinus syndrome or thrombosis reported to date. Our patient presented with visual symptoms and she was diagnosed and treated for optic neuritis as that would be the commonest diagnosis for her presentation. She was given intravenous steroids. She did not present with typical features such as impaired extraocular muscle movement, chemosis or congestion of the eyes, Horner's syndrome or trigeminal nerve involvement for venous thrombosis in the cavernous region. In view of the patient receiving immunosuppressants in the past, she was investigated for chronic infective central nervous lesions such as tuberculosis but the investigations did not reveal any positive results.

Neuroimaging done after a course of methylprednisolone showed no evidence of cavernous sinus thrombosis. The authors feel that the pathophysiology behind cavernous sinus thrombosis in this patient could be due to an inflammatory process associated with her background autoimmune condition. A histopathological diagnosis would have been a valuable assessment because such a presentation has never been reported and studied. Our patient did not consent to a biopsy for confirmation of the diagnosis.

We feel that rare associated conditions will need to be considered in patients with demyelinating disorders such as NMOSD so that all visual symptoms are not diagnosed as optic neuritis. That the cavernous sinus lesion improved after steroids is an indication that her cavernous sinus abnormalities could be immune mediated. In their study of 151 cases of cavernous sinus syndrome, Keane, et al. [5] found self-limited inflammation is the third most common cause after tumour and trauma. The authors feel more case studies and research are required to confirm this association.

Conclusion

Cavernous Sinus thrombosis is rare in Neuromyelitis Optica Syndrome Disorder and can be an asymptomatic manifestation

Conflict of interest: None

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