It’s more than meets the eye: A unique case of neuromyelitis optica presenting with cerebral venous sinus thrombosis.

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INTRODUCTION

Neuromyelitis Optica spectrum disorder (NMOSD) represents a severe CNS inflammatory- demyelinating disorder. Pathogenetic mechanisms such as coagulation cascade dysregulation and systemic autoimmune activation have been linked with an increased prevalence of vascular thrombosis in these patients.1

We discuss a catastrophic initial presentation of NMOSD complicated by cerebral venous sinus thrombosis (CVST).

CASE PRESENTATION

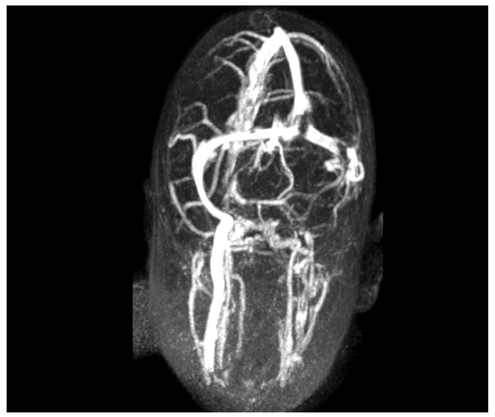
A 43-year-old female with a past medical history of chronic pain and progressive functional decline presented with altered mental status, vision loss, and vomiting.

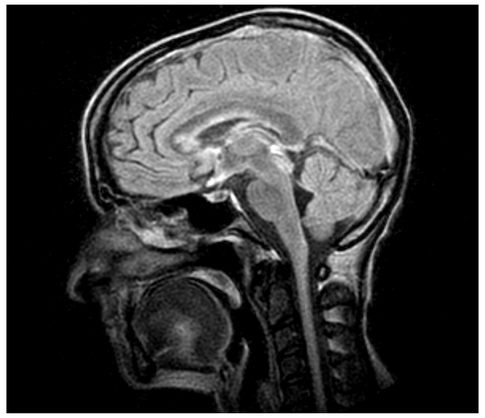
- On exam, she was afebrile, hypotensive, minimally responsive, and bilaterally paraplegic.
- An Initial head CT and CTA suggested left transverse and sigmoid sinus thrombi extending into left internal jugular vein.
- **Clinical course:**
  - The patient was admitted to the neuro-ICU and therapeutic anticoagulation with unfractionated heparin and fluid resuscitation was initiated.
  - An MRV of the brain showed CVST with foci of enhancements in brain stem, corpus callosum, and periventricular region;. MRI of the spine showed diffuse enhancement throughout the cord indicating extensive myelitis.
  - Seropositivity of NMO/Aquaporin 4 IgG (AQP4-IgG) confirmed the diagnosis of NMOSD, she was started on 1 gram of IV solumedrol per day.

CASE PRESENTATION (cont.)

- Workup for hypercoagulable causes and associated autoimmunity included a negative antinuclear antibody, myelin oligodendrocyte glycoprotein antibody, rheumatoid factor, C4, C3, IGA, Anti SS-A & B, beta-2-microglobulin and anticardiolipin antibody.
- The hospital course was complicated by dysautonomia including hypotension and hypothermia with requirement if pressors. Workup for sepsis was negative and the cause was considered secondary to a hypothalamic demyelinating lesion or due to NMOSD.
- She had plasma exchange therapy with subsequent stabilization of temperature and blood pressure. Her encephalopathy improved; she regained some motor function in her toes and a slight improvement in visual acuity. She was discharged with a plan for an outpatient eculizumab infusion.

IMAGING





- Figure 1 : MRV, Left posterior view shows filling defects in region of torcula and patchy filling defects in left distal transverse sinus
- Figure 2: MRI: enhancements seen in the periventricular and subcortical white matter regions with corpus callosum involvement and areas of punctate enhancement

DISCUSSION

- Thrombo-inflammatory pathophysiology has been described in association with CNS disorders including multiple sclerosis and NMOSD.2
- Autoimmunity and conditions like antiphospholipid syndrome in association with NMOSD can be a contributing factor for the development of vascular thrombosis.3
- In our case, a presentation with CVST and bilateral paraplegia prompted a workup with the discovery of longitudinally extensive transverse myelitis and AQP4-IgG seropositive NMOSD.

CONCLUSION

The association between demyelinating CNS disorders and vascular thrombosis has been described.

This case highlights the importance of considering this association and look for underlying demyelinating pathologies in patients presenting with CVST

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