Vision Preservation in COVID-related Cerebral Sinovenous Thrombosis with Optic Nerve Sheath Fenestration

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A 21-year-old female presented to the emergency department (ED) with a one-day history of horizontal diplopia and headache. Three weeks previously, she had been diagnosed with COVID-19 after reporting loss of taste and smell. Past medical history was additionally notable for obesity (BMI 44.1)

On initial ED examination she had a right esotropia. The neurology service pursued neuroimaging, starting with CT head without contrast. This was followed by an MRI brain without and with contrast, performed in conjunction with an MR venogram (MRV) of the head. MRV head revealed cerebral sinovenous thrombosis (CSVT) involving a majority of the superior sagittal sinus, including tributary cortical veins, the right transverse and sigmoid sinuses, and the right jugular bulb. The brain parenchyma was normal on CT head and MRI brain. Hematology was consulted, who started the patient on a heparin drip and admitted her to the neuro intensive care unit. She had an embolic work-up, which was unrevealing. She also had an extensive hypercoagulable work up, which was notable only for an elevated D-dimer (610 ng/mL). Following initiation of anticoagulation, the patient remained stable, was transitioned to Apixaban, and discharged home.

A week later, headache and diplopia worsened. A community ophthalmologist evaluated the patient and found a right cranial nerve VI palsy and bilateral, high-grade optic nerve edema, prompting urgent ED referral. CT head and CT venogram (CTV) were performed, demonstrating extensive CSVT, with a decrease in clot burden since the MRV. Lumbar puncture opening pressure was 40 cm H2O. She was started on Acetazolamide (500mg BID) and instructed to continue oral anticoagulation. She was then referred to our neuro-ophthalmology clinic.

On examination vision was 20/20 OU. Pupils were equally reactive without an afferent pupillary defect (APD). She had a 12° right esotropia and -1 limitation to abduction of the right
eye, consistent with a right sixth nerve palsy. She also had Frisen grade III papilledema (Figure 1). Humphrey Visual Field (HVF) revealed paracentral areas of depression in the right eye and an enlarged blind spot in the left. Retinal nerve fiber layer (RNFL) global thickness was 259µm OD and 220µm OS. Treatment options were discussed, including continued medical therapy versus surgical interventions. Given resolved headache, she elected to continue her medications at the current doses.

The patient was re-examined within one week. Vision decreased to 20/30 OD, there was a right APD, and HVF revealed an enlarged blind spot with superotemporal areas of depression OD. OS remained stable. RNFL thickness increased to 274µm OD and 221µm OS. A plan was made for optic nerve sheath fenestration (ONSF) of the right eye the following day.

The patient underwent an uncomplicated ONSF. At three weeks post-operatively, vision improved to 20/25 OD. She had a residual, mild right APD. HVF showed only a stable enlarged blind spot. RNFL OD improved to 151µm. OS remained stable (Figure 2). At 2.5 months post-operatively, headaches resolved, vision improved (20/20 OU), visual fields were full, diplopia had largely resolved, with a 12° esotropia in right gaze only, and RNFL global thickness improved OU, without ganglion cell layer loss on OCT. MRV showed resolution of the right sided clot with only residual clot in the superior sagittal sinus. She was continued on Acetazolamide and Apixaban.

Cerebral sinovenous thrombosis (CSVT) is a relatively uncommon thromboembolic event within the general population (1). While there has not been an increase in the reported number of CSVT cases during the 2020 COVID pandemic, there is emerging evidence of COVID-19 associated coagulopathy (1-4), with (CSVT) as a complication with neuro-ophthalmologic implications (1,5,6). Although the pathophysiology remains unknown, there is a
growing consensus that viral infections may lead to profound dysregulation between inherent procoagulant and anticoagulant mechanisms, leading to endothelial dysfunction, inhibition of fibrinolysis, increased blood viscosity, and septic-associated coagulopathy (7, 8). Patients with COVID-19 have been found to have anticoagulant dysregulation resulting in elevated D-dimer levels and fibrinogen (9). These patients are therefore at a higher risk for thromboembolic events such as CSVT and other sequelae. Papilledema is often a consequence of venous hypertension from CSVT. Early detection and treatment are critical in preventing vision loss.

The mainstay of treatment for CSVT is anticoagulation to prevent further clot formation and promote clot dissolution (10). Management of CSVT sequelae, such as papilledema, is an important consideration. Mild papilledema can often be managed with anticoagulation and oral carbonic anhydrase inhibitors (acetazolamide). Severe cases, at high risk of vision loss, may require serial lumbar punctures, however, are also treated with ONSF and other cerebrospinal fluid (CSF) diversion procedures (e.g. ventriculoperitoneal, lumboperitoneal shunt) (11). Several endovascular interventions have been reported for the treatment of CSVT: thrombolysis, thrombectomy, stenting, or a combination of these. The efficacy of these treatments has not been established, and most endovascular treatments have been deemed as salvage therapies for refractory CSVT (12).

We present a female with CSVT, elevated ICP, high-grade papilledema, sixth nerve palsy, and reduced vision, following a symptomatic COVID infection. This is a timely and interesting case due to increasing recognition of thrombotic events associated with COVID infections. Several studies have reported significantly increased rates of venous and arterial thromboses in patients with COVID-19, but most of these events have been in hospital settings,
particularly the ICU (13-16). CSVT has only rarely been reported in the context of COVID-19, especially in the outpatient setting.

Unique to this case is our patient having only mild COVID symptoms, yet remaining at risk for hypercoagulable events, given her elevated D-dimer—which has been shown to not only predict likelihood of venous thromboembolism, but also stratify risk (15). While evidence for direct oral anticoagulant (DOAC) use in CSVT is limited, there is a growing tendency to consider DOACs for the treatment of hypercoagulable states, including CSVT. Apixaban was the preferred (DOAC) in this case (17,18). Her high-grade papilledema, despite imaging signs of decreased clot burden, placed her at risk for permanent vision loss. Therefore, it is important to consider this vision threatening condition in COVID-positive patients who also present with visual or neurologic complaints, even in ambulatory settings. This case also highlights the importance of monitoring patients with known idiopathic intracranial hypertension (IIH) or IIH risk factors closely, as acquiring COVID could exacerbate symptoms resulting from CSVT.

ONSF was pursued due to worsening vision, high-grade papilledema, and field defects. ONSF is particularly useful for vision preservation in patients with intractable papilledema in the setting of CSVT to prevent optic nerve damage in the acute setting, while allowing time for clot dissolution. In this way, patients are protected from vision loss while also avoiding more invasive procedures with permanent implanted devices.


12. Effect of Endovascular Treatment With Medical Management vs Standard Care on Severe Cerebral Venous Thrombosis: The TO-ACT Randomized Clinical Trial. JAMA Neurol. 2020 Aug 1;77(8):966-973.


Figure 1. High-grade, bilateral papilledema at the time of presentation to the neuro-ophthalmology clinic.

Figure 2. Reduced papilledema in the right eye three weeks following ONSF (image on left).
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