A Case of Pediatric Vision Loss

Keegan Nicodemus, MD and Jaron Raper, MD

Emergency Department
University of Alabama at Birmingham

HISTORY OF PRESENT ILLNESS

10-year-old male with no past medical history presents with left eye vision loss over the last 5 days. His mother reports a recent history of Streptococcus pharyngitis one-month prior treated with one week of penicillin. His fever resolved but his sore throat continued for 10 days with malaise and vomiting, after which he was switched to a cephalosporin for 5 days. His symptoms largely improved but he then developed neck pain at the end of his antibiotic course. He underwent outpatient MRI which was reportedly negative, diagnosed with torticollis and placed on methocarbamol. He neck pain fluctuated for one week; however, he then developed blurry vision. He underwent repeat CT/MRV, which were also reportedly negative. Due to concern for diplopia and a possible extraocular palsy, he was transferred to a pediatric tertiary referral hospital for further evaluation. At that time, he was not found to have a palsy and outside images were requested, reviewed and overread as negative as well. After 3 days, his vision continued to worsen and he presented again to the pediatric hospital with frank vision loss on the left. He also endorsed sporadic headache but denied any recurrent fever, development of rash, or recurrence of neck pain.

PHYSICAL EXAM

Vitals: T 99F, HR 96bpm, BP 133/89, RR 24, SpO2 99%

General: Well-appearing, no acute distress, no rash

HEENT: OD 20/40, OS only light perception, pupillary exam seen in Figure 1, bilateral CN VI palsies, oropharynx with exudate/erythema

Neck: FROM, negative Brudzinski

Neuro: A&Ox3, no focal neuro deficits, normal gait, equivocal Babinski

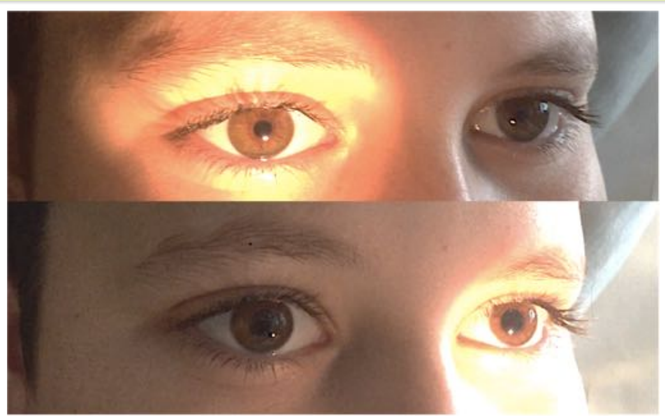

Figure 1. Pupillary exam with left afferent pupillary defect

DIAGNOSTICS

Laboratory: CBC, ESR, CRP, CSF within normal limits

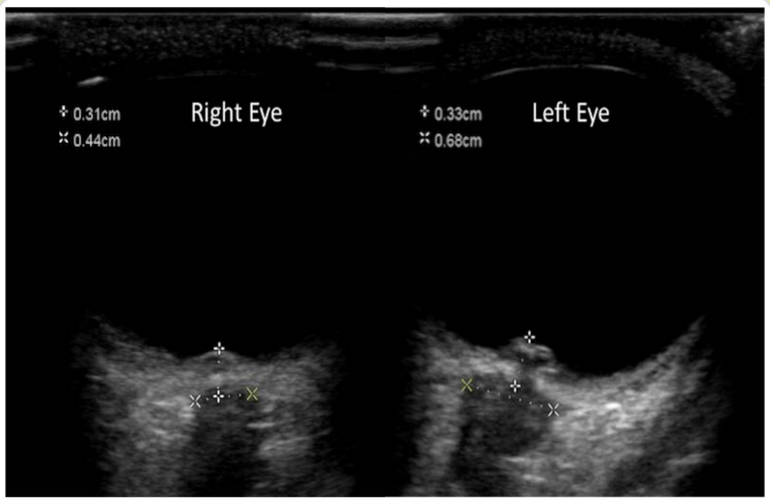

Figure 2. Ocular ultrasound with bilateral papilledema and dilated left optic nerve sheath diameter

Figure 3. MRV with Empty Delta Sign in posterior aspect of Superior Sagittal Sinus

CASE DISCUSSION

Bilateral CN VI palsies and bedside ultrasound (Figure 2) with bilateral papilledema and left optic nerve sheath diameter dilation indicated increased ICP. Review of outside MRV (Figure 3) revealed posterior "empty delta sign" consistent with cerebral venous thrombosis (CVT). Repeat MRV showed 15cm thrombus throughout the Superior Sagittal Sinus, bilateral Transverse Sinuses, Right Sigmoid Sinus and Right Internal Jugular Vein. The patient underwent emergent thrombectomy. No hypercoagulable etiology was found. The patient would go on to have permanent left vision loss and recurrent thrombosis with permanent right vision loss.

Cerebral venous thrombosis is a difficult diagnosis to make. When considering CVT, non-enhanced studies should be scrutinized for the "delta sign" or "dense clot sign" which appears as a triangular enlargement in the anterior or posterior aspects of superior sagittal sinus. Contrasted studies should be scrutinized for the "empty delta sign" which can either show asymmetric filling of the sinus or contrast enhancement outlining the clot.

CONCLUSION

- Bilateral CN VI palsies are a red flag for increased ICP and possible cerebral venous thrombosis
- Ocular ultrasound is an easy, viable alternative to fundoscopy. It can be more sensitive for increased ICP by evaluation of the optic nerve sheath diameter and is standardized in the evaluation of papilledema
  - ➢Diameters < 5mm are considered normal, > 6mm abnormal and any papilledema abnormal
- In cases with subacute neurological deficits, venous-phased imaging should be obtained and scrutinized for "dense clot" and "empty delta" signs as cerebral venous thrombosis can be easily missed

REFERENCES

Farkas, Josh. "Pulmcrit: Algorithm For Diagnosing ICP Elevation With Ocular Sonography". *Emcrit.Org*, 2017

Kuehnen, J., 1998. Cranial nerve syndrome in thrombosis of the transverse/sigmoid sinuses. *Brain*, 121(2), pp.381-388.

Simons, Barbara et al. "The Radiology Assistant : Cerebral Venous Thrombosis". *Radiologyassistant.Nl*, 2021

ACKNOWLEDGMENTS

Thank you to Dr. Jaron Raper for editing, Dr. John Gullett for educational expertise, Dr. Ryan Roddy for assistance in managing the patient, Children's of Alabama Departments of Neurology, Neurosurgery and Critical Care for prompt evaluation and management of the patient. Educational consent obtained for all images shown.