



Ophthalmologic Manifestations of Pheochromocytoma in a 9-year-old Patient

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CASE PRESENTATION

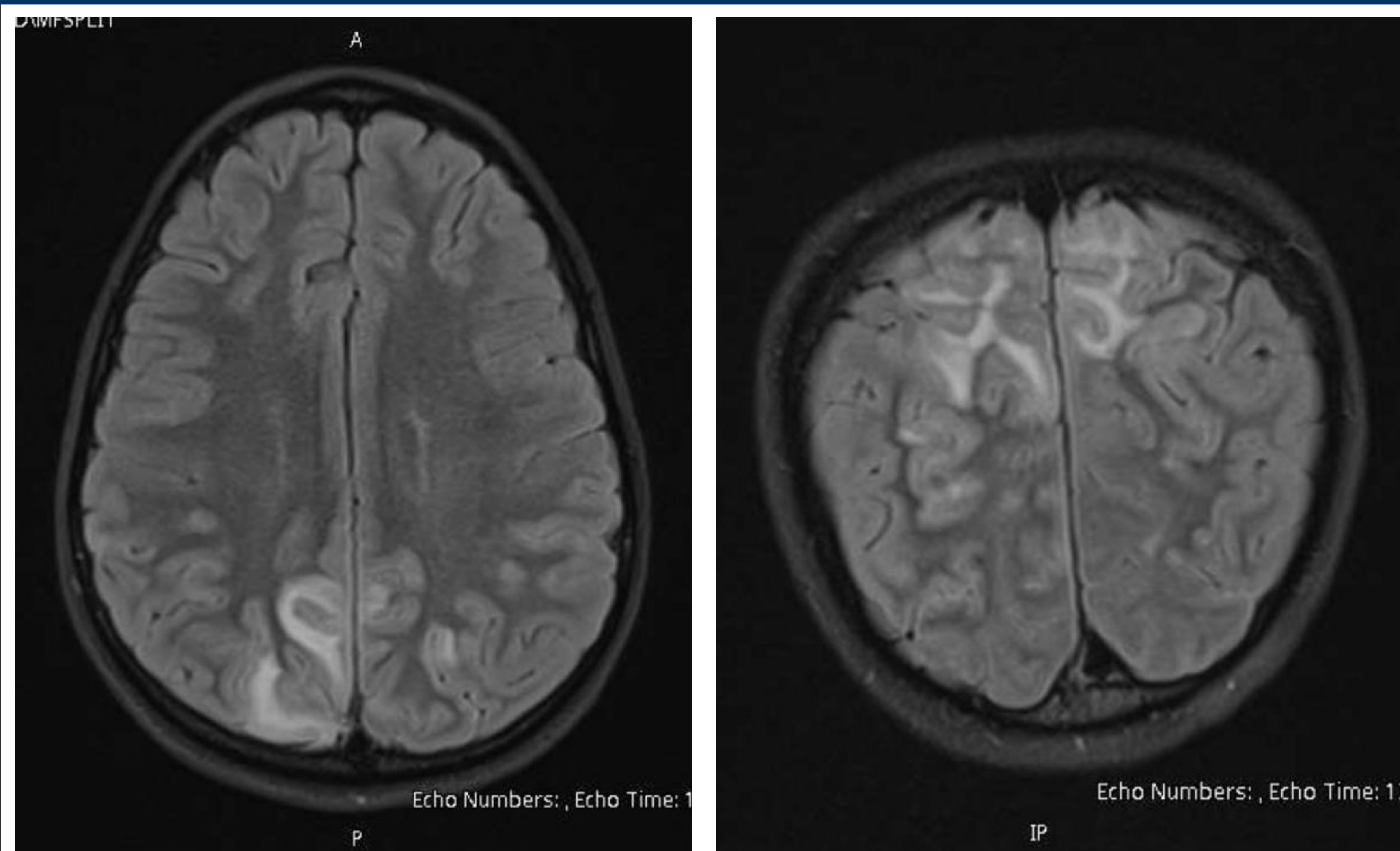
- Inpatient consult for a 9 year-old female admitted for hypertensive emergency and new-onset seizures
- On admission she complains of blurry vision x 3 weeks
- No ocular history, baseline vision unknown
- VA: 20/25 OD, 20/70 OS
- IOP 19/16
- PEERLA, no APD
- EOMS and CVF Full
- On initial consult, her anterior and posterior exam were unremarkable
- Blood pressure was noted to be 190s/130s
- MRI was performed.

- One week later, vision has decreased
- VA 20/20 OD, 20/400 OS
- Posterior exam at that time notable for multiple peripapillary cotton wool spots and macular edema in the left eye
- No optic nerve edema OU

DIAGNOSIS

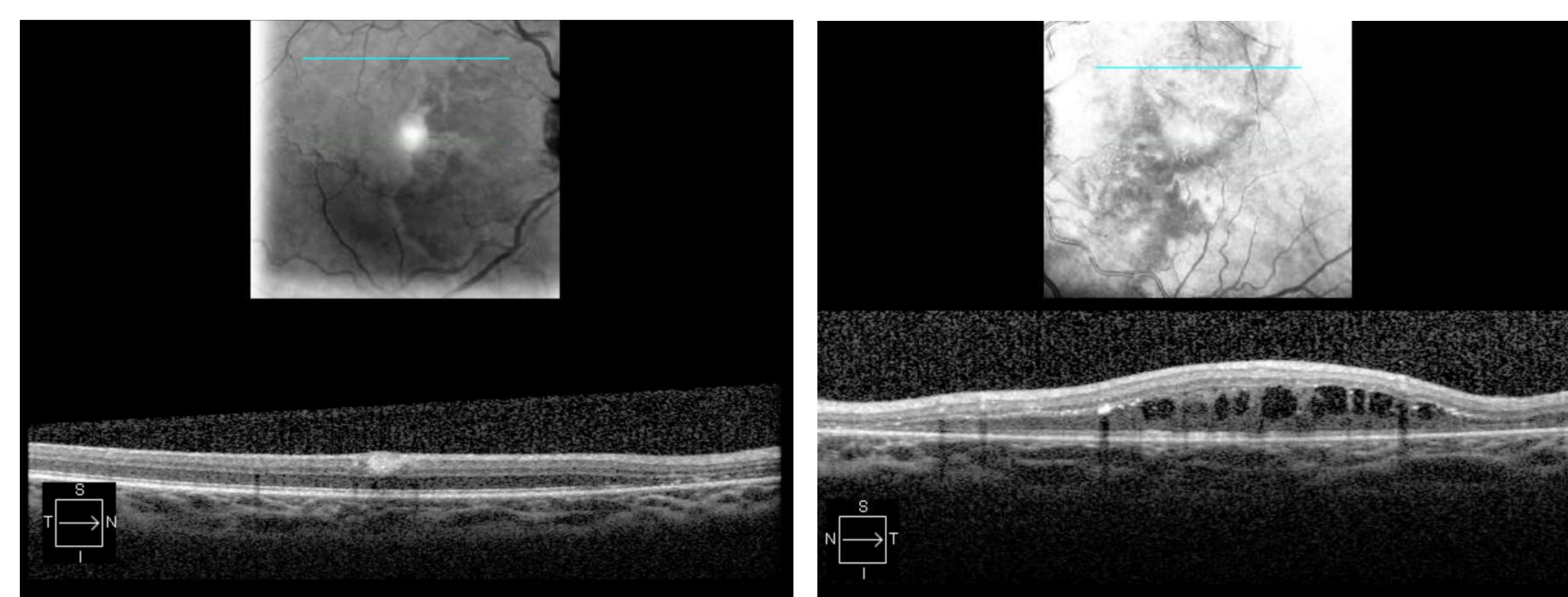
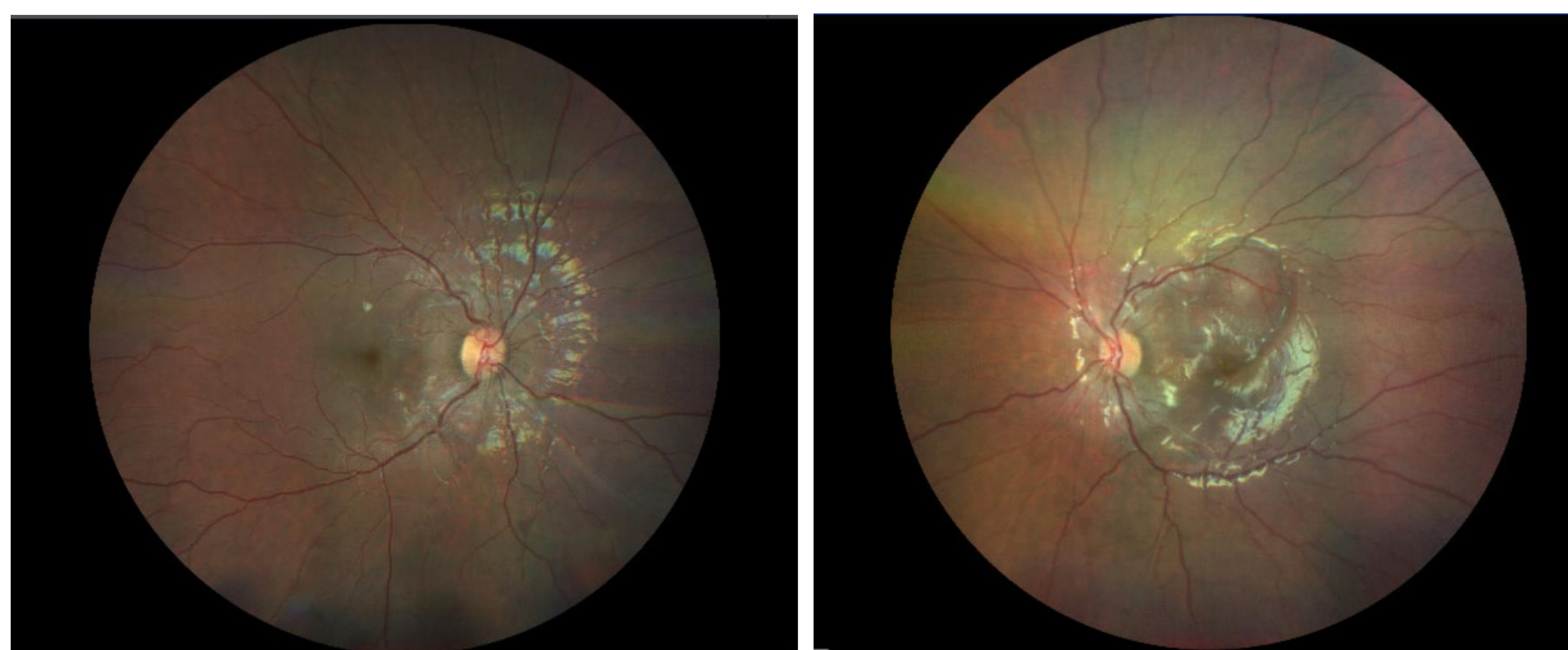
- Based on radiographic findings, she was diagnosed with posterior reversible encephalopathy syndrome (PRES)
- PRES is a neurotoxic process secondary to cerebral edema—typically in the parieto-occipital lobes in watershed areas, but not necessarily always posterior
- Predisposing factors: multiple, esp. systemic inflammatory responses; severe hypertension
- Pathogenesis: heterogenous, although end result is a breakdown of the blood-brain barrier leading to vasogenic edema

NEUROIMAGING



MRI was remarkable for edema involving the bilateral parieto-occipital regions with enhancement of the sulci in the affected regions, as seen on axial (left) and coronal (right) T2 FLAIR images.

FUNDUS FINDINGS



OCT of the macula showing a cotton wool spot in the nerve fiber layer (OD) and intraretinal fluid in the superior macula (OS)

Dilated fundus exam revealed Grade III-IV Hypertensive Retinopathy

MANAGEMENT

- Management is centered on treating the underlying pathology

CASE FOLLOW UP

- Our patient was found to have a norepinephrine-secreting pheochromocytoma
- Pheochromocytoma has an estimated incidence of 5 per 1 million people
- Genetic testing was sent on our patient as pheochromocytoma can be associated with other syndromes such as MEN2
- While awaiting surgery, BP was controlled with multiple anti-hypertensive agents
- At 1 month follow up, VA was stable from our initial exam
- Cotton wool spots were noted in both eyes OS>OD
- She underwent surgical resection ~1 month after diagnosis
- She was unfortunately lost to follow up at Upstate CVC

SELECTED REFERENCES

❖ Sudulagunta SR, Sodalagunta MB, Kumbhat M, Settikere Nataraju A. Posterior reversible encephalopathy syndrome(PRES). *Oxf Med Case Reports*. 2017 Apr 3;2017(4):omx011. doi: 10.1093/omcr/omx011. PMID: 28473920; PMCID: PMC5410886

❖ Tsukikawa, M., & Stacey, A. W. (2020). A Review of Hypertensive Retinopathy and Chorioretinopathy. *Clinical optometry*, 12, 67–73. <https://doi.org/10.2147/OPTO.S183492>

❖ <https://www.cancer.net/cancer-types/pheochromocytoma-and-paraganglioma/statistics>.



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