AMSER Case of the Month: July 2024

5-year-old male with increased intracranial pressure

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Patient Presentation

HPI: 5-year-old male presented to the ED with increased optic disc swelling found on routine fundoscopic examination by his optometrist. Ophthalmology consult appreciated grade 1-2 papilledema. Patient denied recent head trauma, fever, headache, nausea, vomiting or vision loss.

Medical History: Bannayan-Riley-Ruvalcaba Syndrome (PTEN mutation), developmental and speech delay.

Medications: Flonase, Zyrtec

Physical Exam & Labs: Unremarkable

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

This imaging modality was ordered by the ER physician

Variant 6: **Child. Six months of age or older. Papilledema detected on the ophthalmologic examination or signs of raised intracranial pressure. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head and orbits without and with IV contrast	Usually Appropriate	O
MRI head and orbits without IV contrast	Usually Appropriate	O
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	May Be Appropriate	O
MRV head with IV contrast	May Be Appropriate	O
MRV head without and with IV contrast	May Be Appropriate	O
MRV head without IV contrast	May Be Appropriate	O
CT head without IV contrast	May Be Appropriate	☢☢☢
CTV head with IV contrast	May Be Appropriate	☢☢☢☢
MRA head without and with IV contrast	Usually Not Appropriate	O
MRA head without IV contrast	Usually Not Appropriate	O
MRI orbits without and with IV contrast	Usually Not Appropriate	O
MRI orbits without IV contrast	Usually Not Appropriate	O
CT head and orbits with IV contrast	Usually Not Appropriate	☢☢☢
CT head and orbits without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT head and orbits without IV contrast	Usually Not Appropriate	☢☢☢
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT orbits with IV contrast	Usually Not Appropriate	☢☢☢

Findings (unlabeled)

Axial T2

MRV Time of Flight (TOF)

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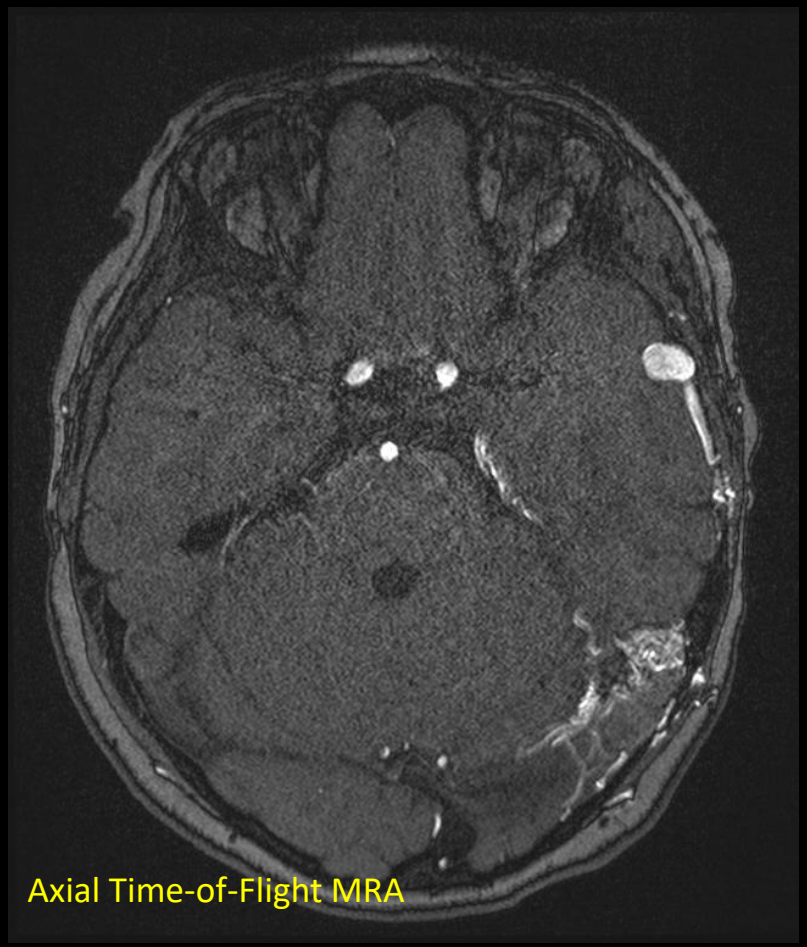
Findings (unlabeled)

Axial Time-of-Flight MRA



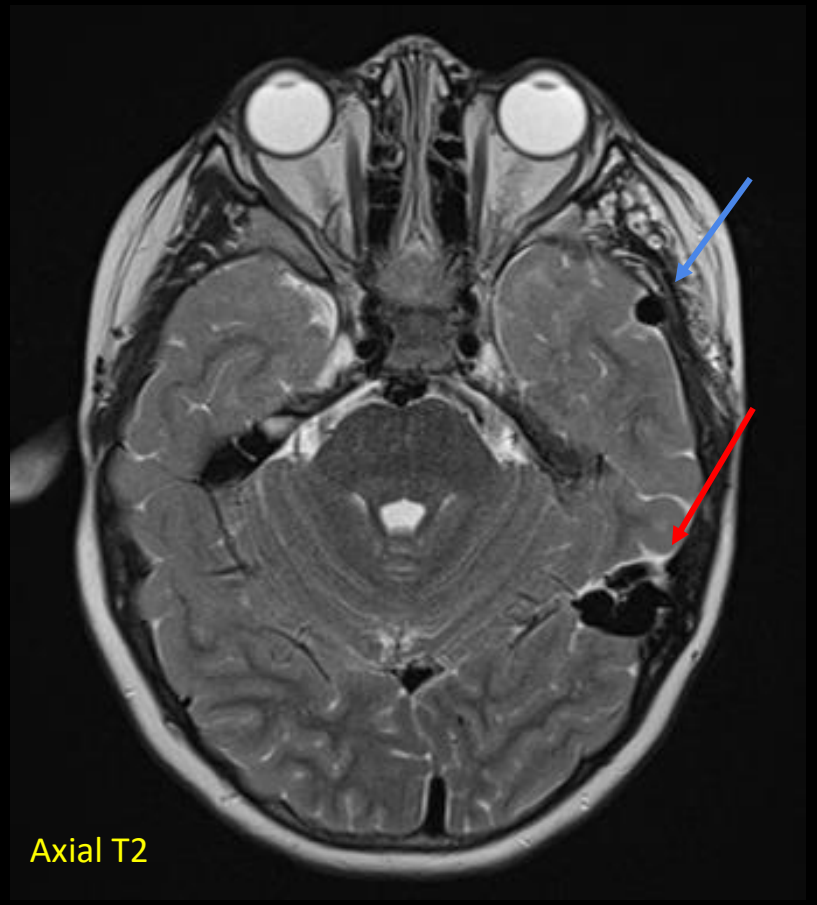
Axial Time-of-Flight MRA

Findings (unlabeled)

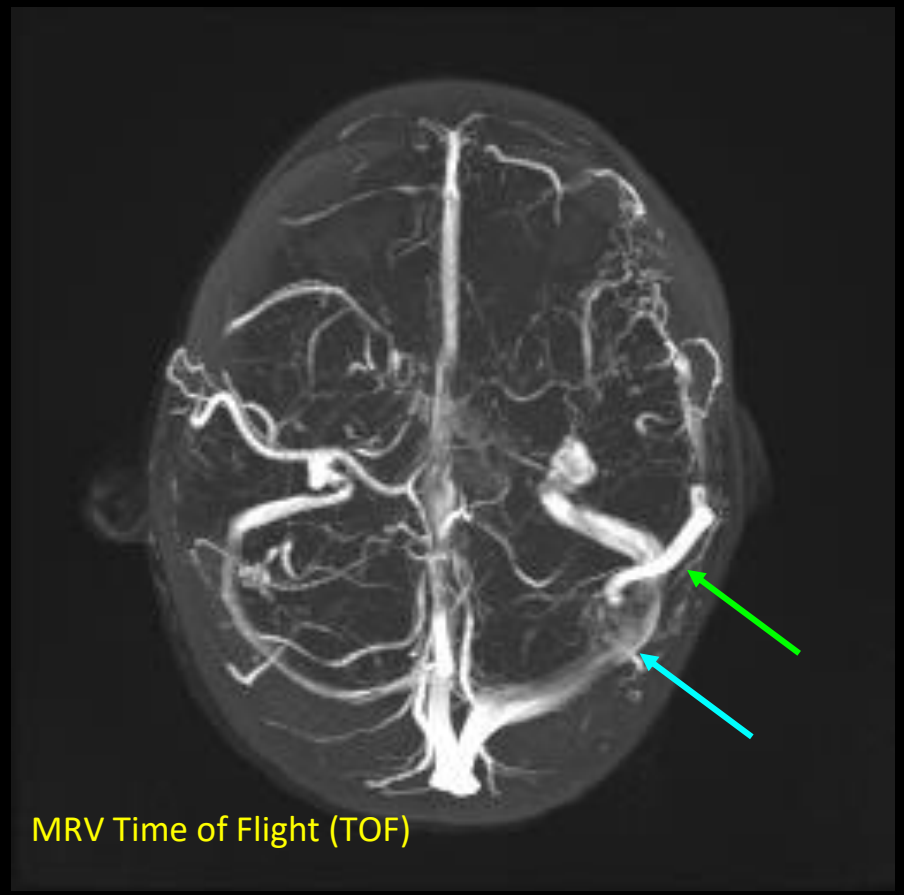
Digital Subtraction Angiography (DSA)

DSA post sclerotherapy

Findings (labeled)



Axial T2



MRV Time of Flight (TOF)

Axial T2 weighted imaging demonstrates T2-dark flow voids from prominent vessels in the region of the left transverse sinus, as well as a prominent cortical vessel.

MRV TOF imaging demonstrates a prominent abnormal vessel in the region of the left temporal convexity, with an apparent communication with the left transverse sinus near the transverse/sigmoid junction.

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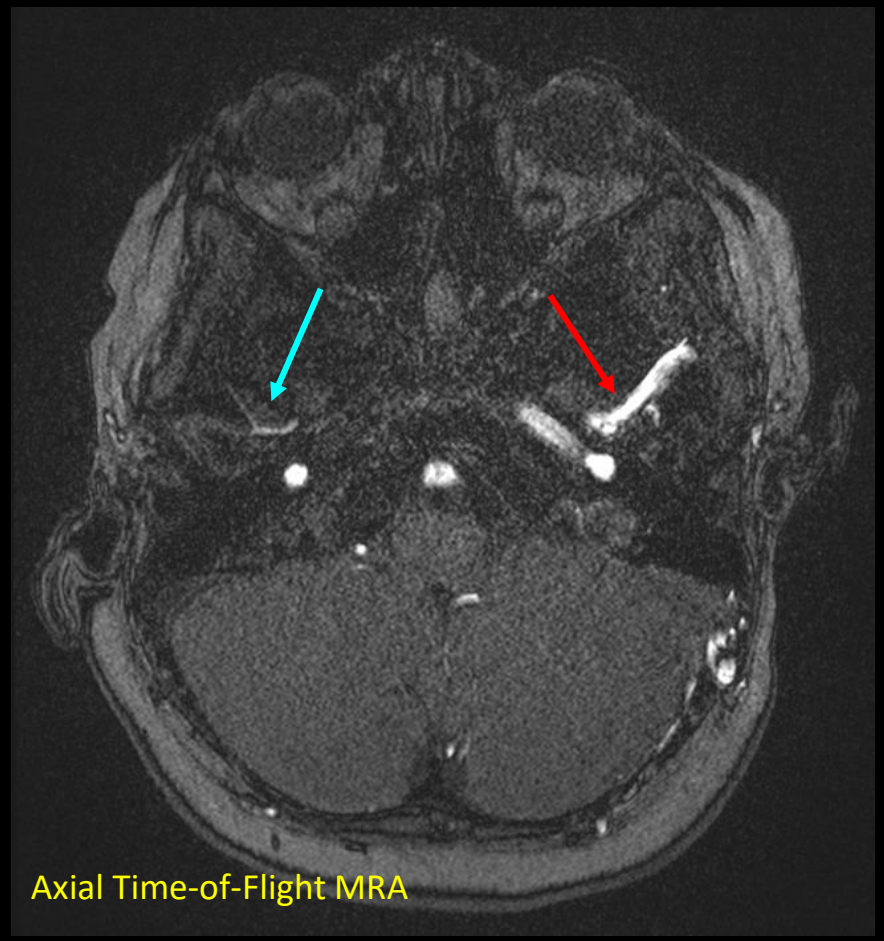
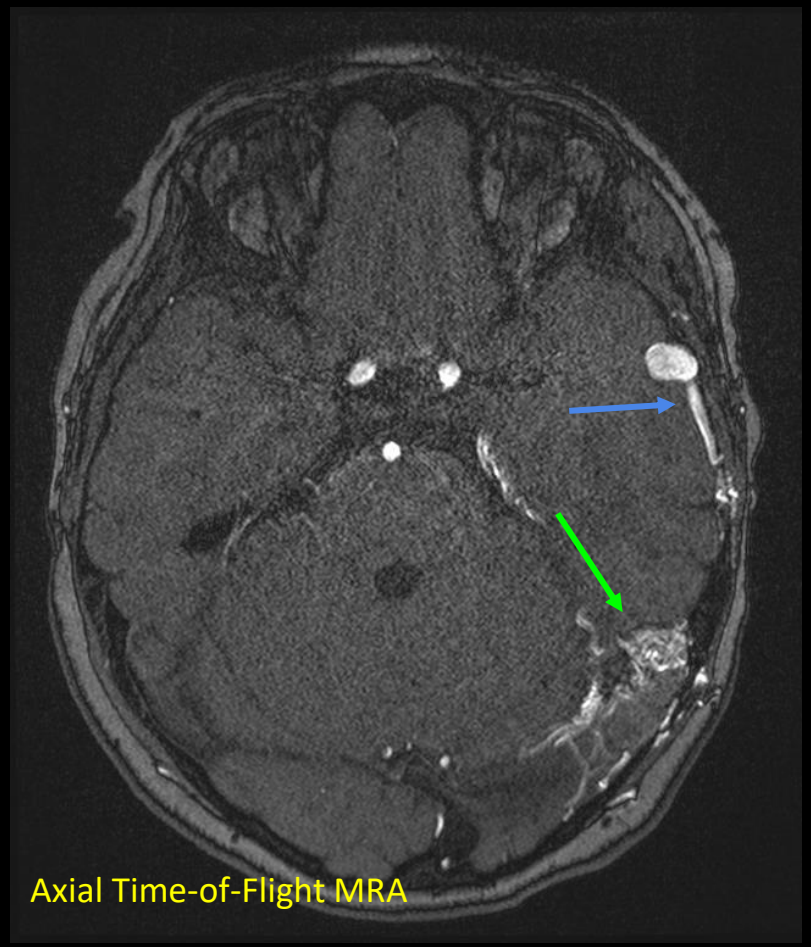
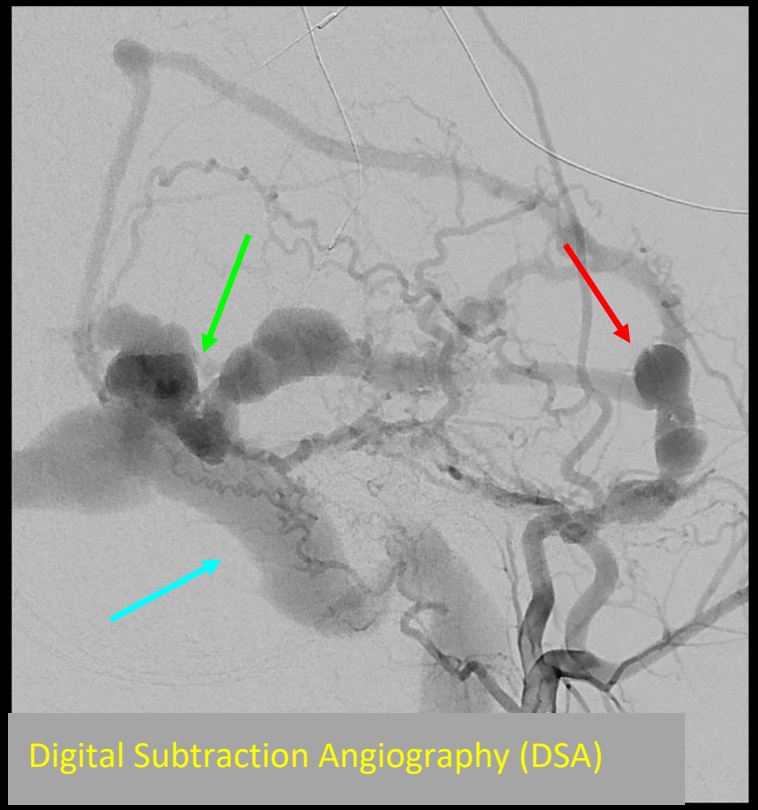
Findings (labeled)



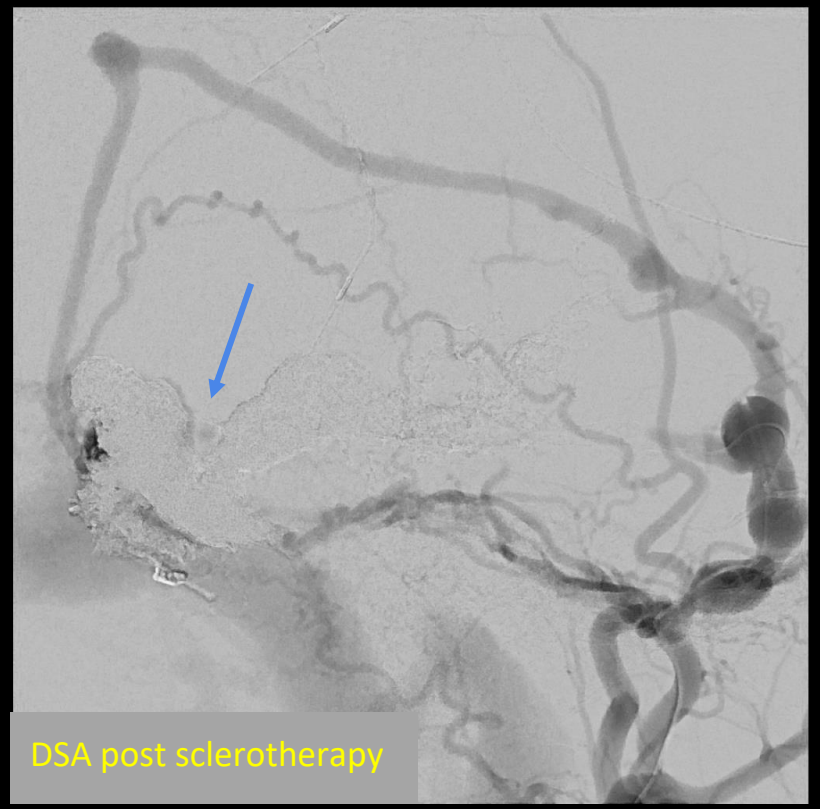


Axial Time-Of-Flight (TOF) imaging demonstrates marked asymmetric enlargement of the left middle meningeal artery when compared to the right, which can be seen feeding the highly vascular lesion along the left transverse sinus.

Findings (labeled)



Digital Subtraction Angiography (DSA)



DSA post sclerotherapy

Digital subtraction angiography (DSA) demonstrates the enlarged, tortuous middle meningeal artery arising from the external carotid artery feeding into the vascular lesion, which can be see draining into the transverse/sigmoid sinus and through the internal jugular vein.

Post sclerotherapy DSA demonstrates absence of flow within the lesion.

Final Dx:

Dural Arteriovenous Fistula (DAVF)
of the Left Transverse-Sigmoid Sinus

Case Discussion

Epidemiology: Most commonly diagnosed between ages 40-60 and accounts for approximately 10-15% of intracranial vascular malformations.

Etiology: The causes of DAVFs largely remain misunderstood but most cases are thought be acquired (secondary to head trauma, infection, dural sinus thrombosis, etc). However, in the pediatric population, most DAVFs are congenital and associated with structural venous abnormalities. Additionally, multiple cases of PTEN hamartoma tumor syndromes, such as BRRS and Cowden Syndrome, have been described in association with DAVFs.

Case Discussion

Clinical Features: DAVFs are asymptomatic for most patients and are often discovered incidentally in the diagnostic workup of other neurological conditions. However, symptomatic patients may experience:

- pulsatile tinnitus
- headache
- seizures
- nausea, vomiting
- papilledema
- speech, language, and coordination deficits
- vision loss
- apathy, parkinsonism, dementia

Case Discussion

Imaging Findings: CT and MR aid in the initial diagnosis of DAVFs. However, digital subtraction angiography (DSA) is considered gold standard for accurate characterization and classification.

- Noncontrast CT reveals intracranial hypertension and edema caused by venous congestion while MR evaluates the anatomy of involved vessels, cortical vein outpouching, and signs of venous hypertension in high-grade lesions.
- With DSA, the presence of arteriovenous shunting most typically with multiple feeders and no intervening capillary structures is characteristic.
- The most common location for DAVFs is the transverse-sigmoid junction with a slight left-sided predominance, and generally are more likely to be supratentorial than infratentorial.

Case Discussion

Management: High-risk, symptomatic lesions with cortical venous drainage can be treated with open neurosurgery, stereotactic radiosurgery, and/or neuroendovascular interventions. Benign lesions are managed conservatively with serial monitoring. The Borden and Cognard classifications are most commonly used in stratifying the severity of the neurological course.

Table 1 Summary of the Cognard and Borden Classification

Cognard	Borden	condition	hemorrhage risk
I , II a	I	DVS/MV outflow only	none
II b, II a+b	II	DVS/MV outflow and CVR	low
III, IV	III	CVR only (isolated sinus)	high

DMS：dural venous sinus, MV：meningeal vein, CVR：cortical venous reflux

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