

Clinical and Neuroimaging findings of an Unusual Pediatric

Presentation of Sturge Weber Syndrome Type III

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BACKGROUND

- Sturge Weber Syndrome (SWS) is a neurocutaneous disorder, of which there are three types:

Type I	Type II	Type III
<ul style="list-style-type: none"> Facial capillary malformation Brain involvement Glaucoma 	<ul style="list-style-type: none"> Facial capillary malformation No brain involvement 	<ul style="list-style-type: none"> Leptomeningeal angiomas No skin or ocular involvement

CASE PRESENTATION

- 13 yo male with no PMH presented with confusion after mild trauma without loss of consciousness.
- He re-presented the following day with right face & extremity weakness, when a stroke code was activated.
- He had persistent right sided hemiparesis and was found to be in status epilepticus.
- There was no evidence of dermatologic or ophthalmologic stigmata associated with SWS.
- Head CT scan concerning for arteriovenous fistula vs. cerebral venous thrombosis.
- MRI showed leptomeningeal enhancement of left hemisphere, consistent with SWS Type III.

CONCLUSIONS

- Sturge Weber Syndrome Type III should be considered in a patient presenting with refractory epilepsy without neurocutaneous stigmata.
- CT and/or MRI findings suggestive of leptomeningeal angiomas should prompt consideration of SWS.

NEUROIMAGING

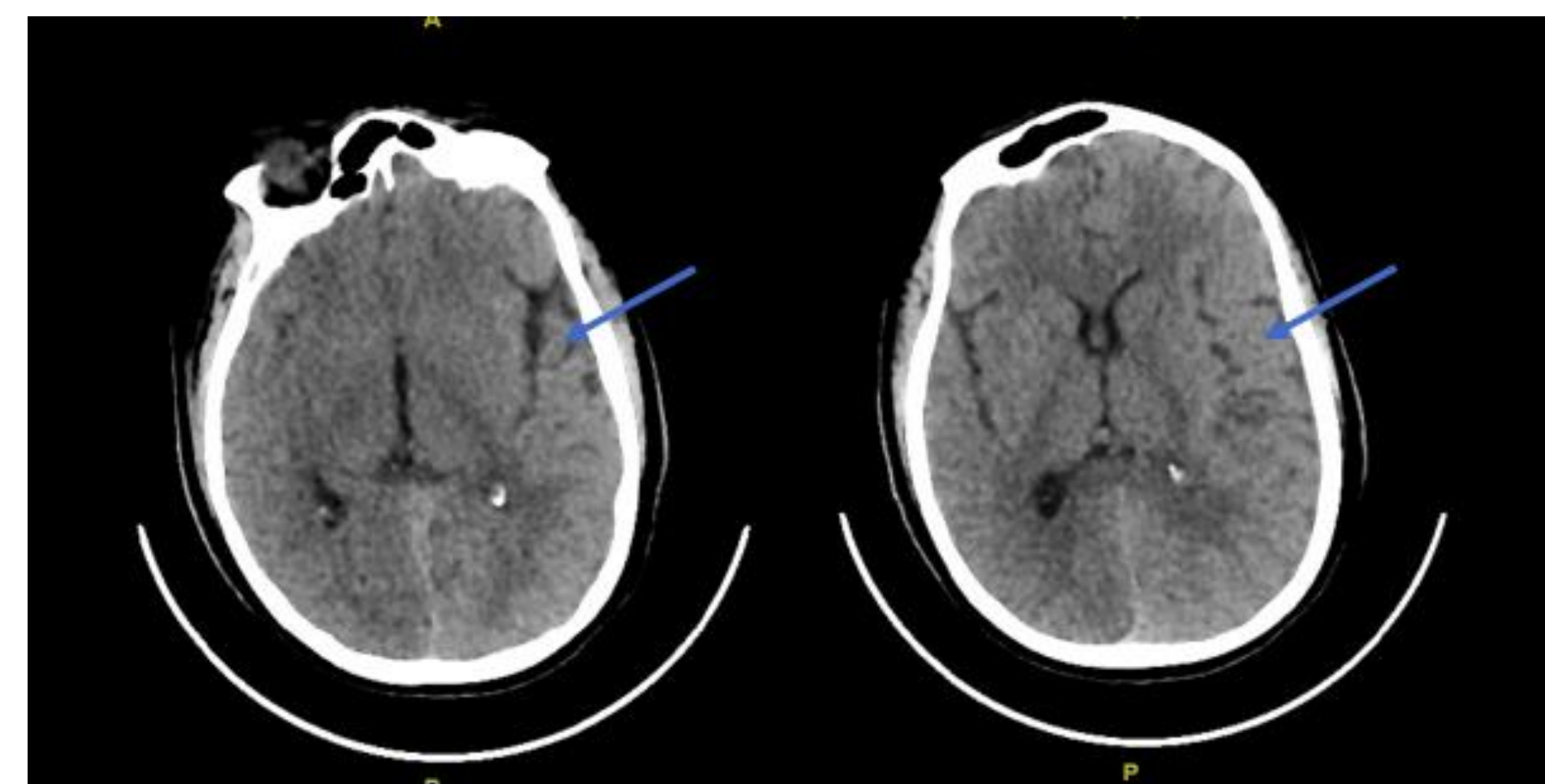


Figure 1. CT head. Left cerebral edema and cortical thickening.



Figure 2. MRI Brain. Axial view (A, B), QSM; SWI (C). Showing prominent leptomeningeal veins seen in the left cerebral hemisphere, including prominent deep medullary and thalamostriate veins.

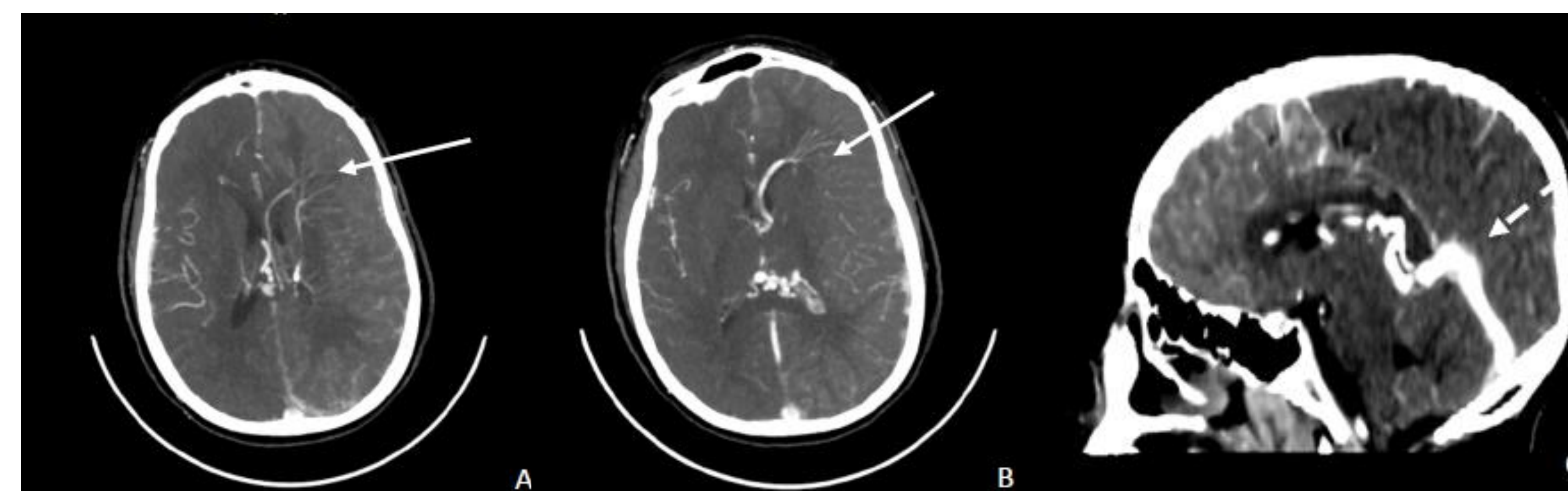


Figure 3. CT angiogram. Axial view (A,B) coronal (C). Left cerebral leptomeningeal angiomas and 2 enlarged veins within the left periventricular left frontal lobe and in the corona radiata (arrows) which drain into an enlarged Vein of Galen (dashed arrow).

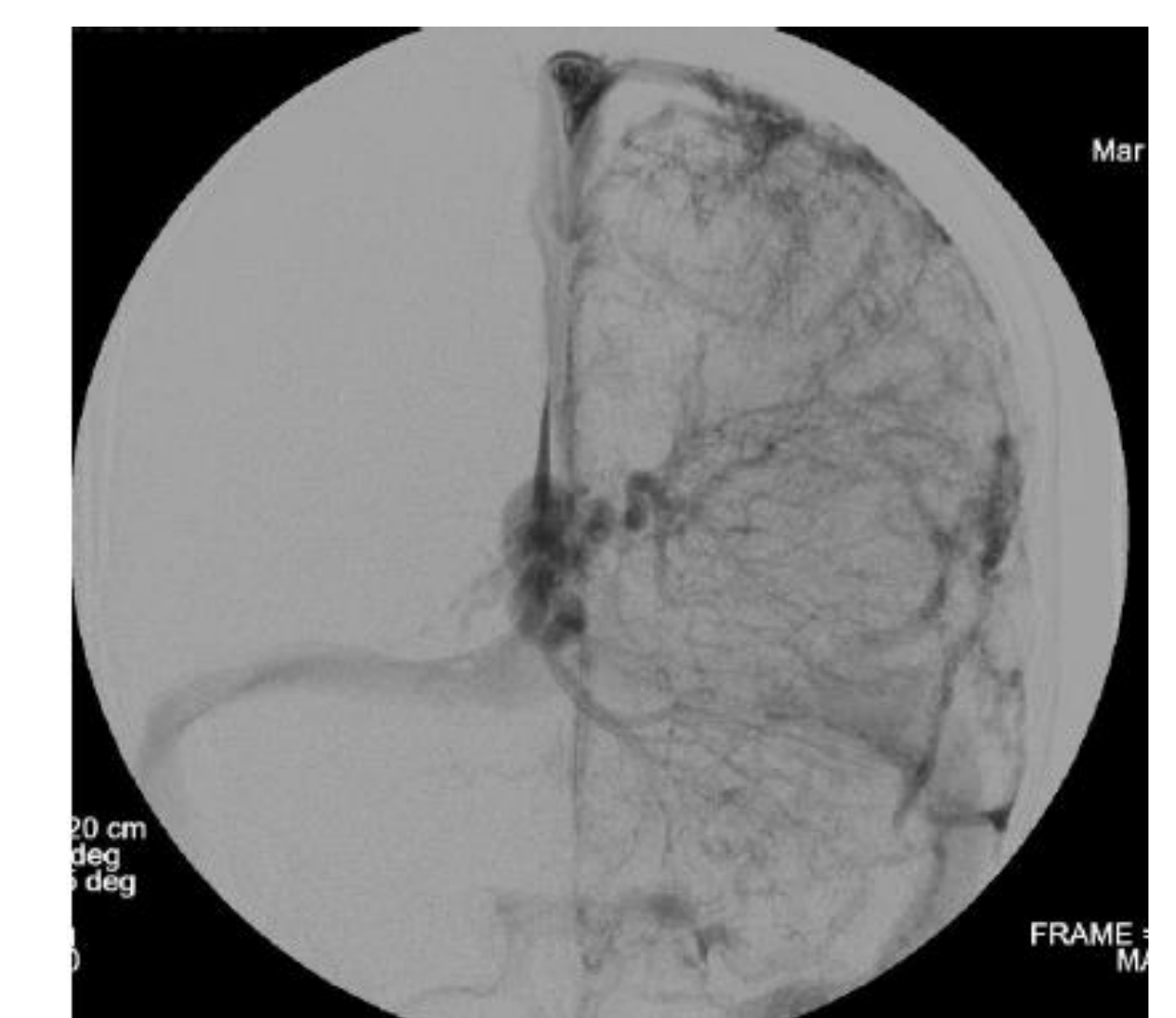


Figure 4. Digital Angiography. During left internal carotid angiogram, there is diffuse leptomeningeal angiomas with early venous filling with enlargement of the left internal cerebral vein, Vein of Galen.

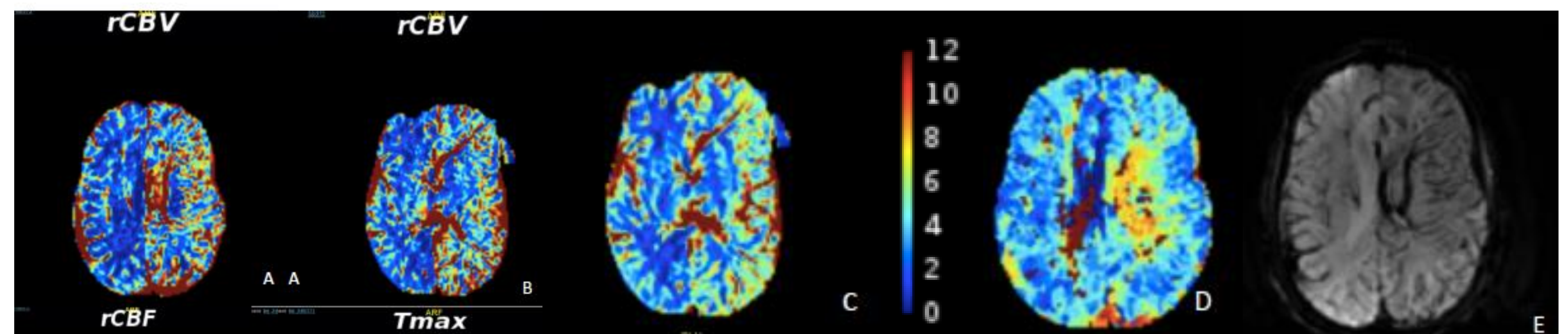


Figure 5. MRI Brain perfusion. Increased cerebral blood flow and blood volume throughout the cortical surface in the left hemisphere compared to the right (A, B, C). There is elevated Tmax in the left cerebral hemisphere likely due to venous congestion (D). RAPID AIF/VOF (E), prominent deep medullary and thalamostriate veins.