

# Sickle Cell Vasculopathy

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## History

Teenager with sickle cell disease.

## Diagnosis

Pial angiomatosis secondary to sickle cell vasculopathy

## Discussion

Sickle cell disease (SCD) increases the risk for stroke by 200 to 400 times; 5-8% of patients with SCD develop symptomatic cerebrovascular disease. Sickled cells adhere to vessel walls leading to intimal and medial damage and subsequent stenosis and occlusion. Sickle cell vasculopathy (SCV) affects small and large vessels but the distal internal carotid artery and the proximal middle cerebral arteries are the most commonly involved.

Transcranial Doppler is useful as a screening tool to predict SCV. Time averaged mean velocities >170 cm/sec may indicate hemodynamically significant stenosis and require MR angiography for anatomic confirmation. Exchange transfusion may reverse the perfusion disturbance diagnosed by Doppler and MR angiography.

Moya moya is a description of progressive stenosis of the supraclinoid internal carotid artery with enlargement of the perforating arteries and development of collateral vessels at the circle of Willis giving rise to a "puff of smoke" appearance. Moya moya can be seen in SCV, radiation vasculitis, and neurofibromatosis.

## Findings

MR-demonstrates internal carotid artery stenosis with collateral vessels and prominent perforating arteries. Note the pial angiomatosis.

## Reference

Lonergan GJ et al. Sickle Cell Anemia. Radiographics (2001); 21:971-994.

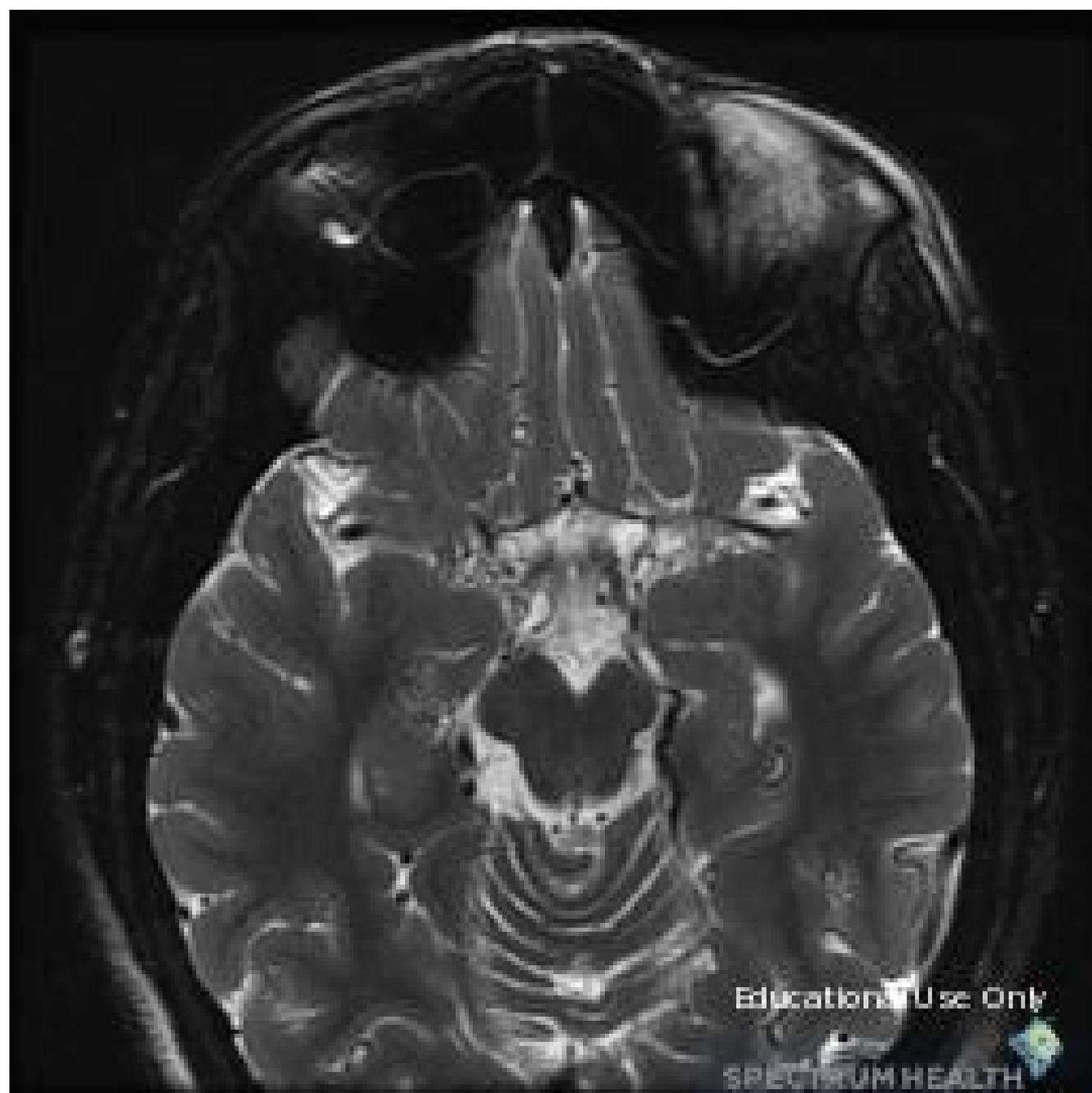
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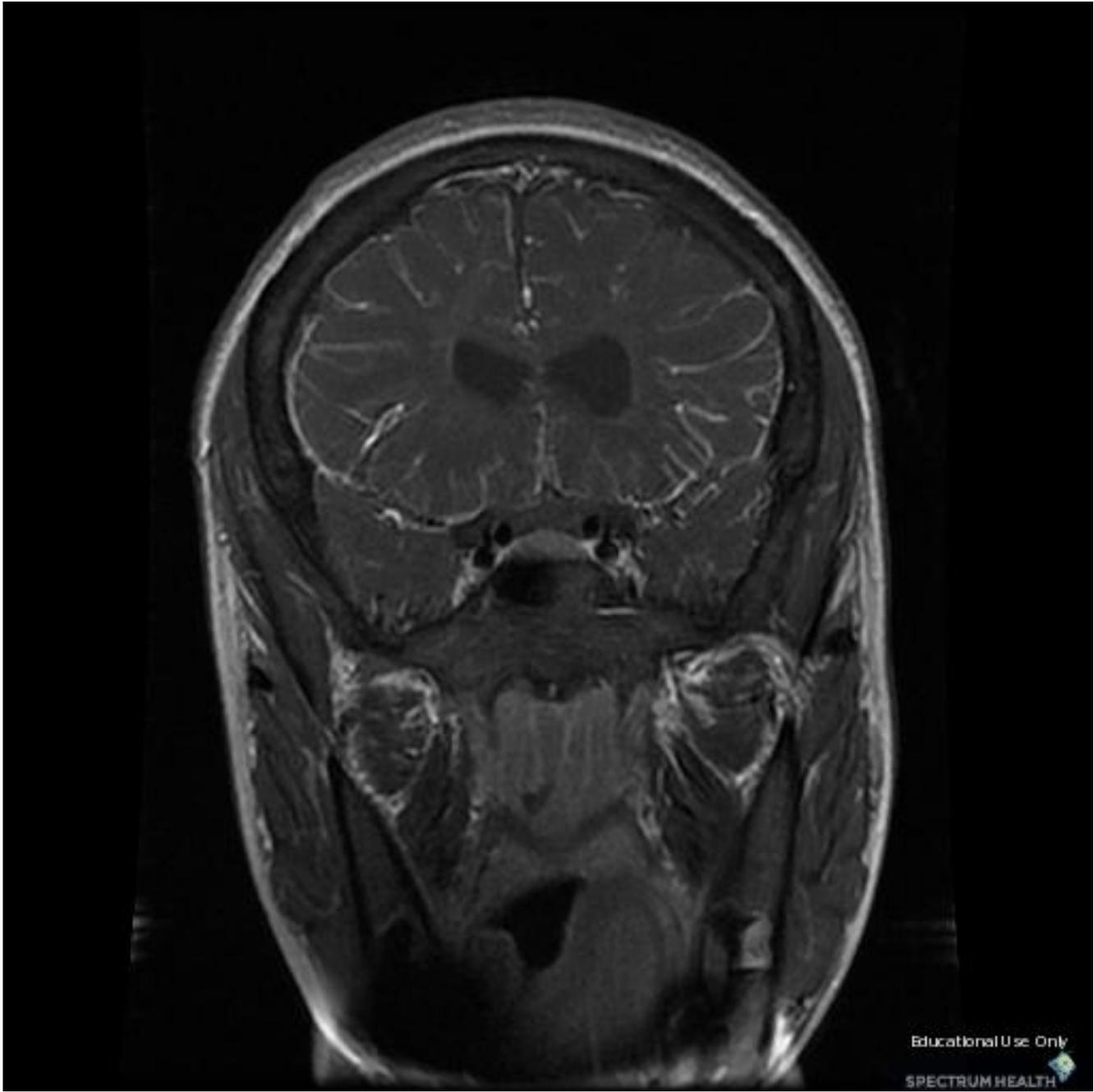




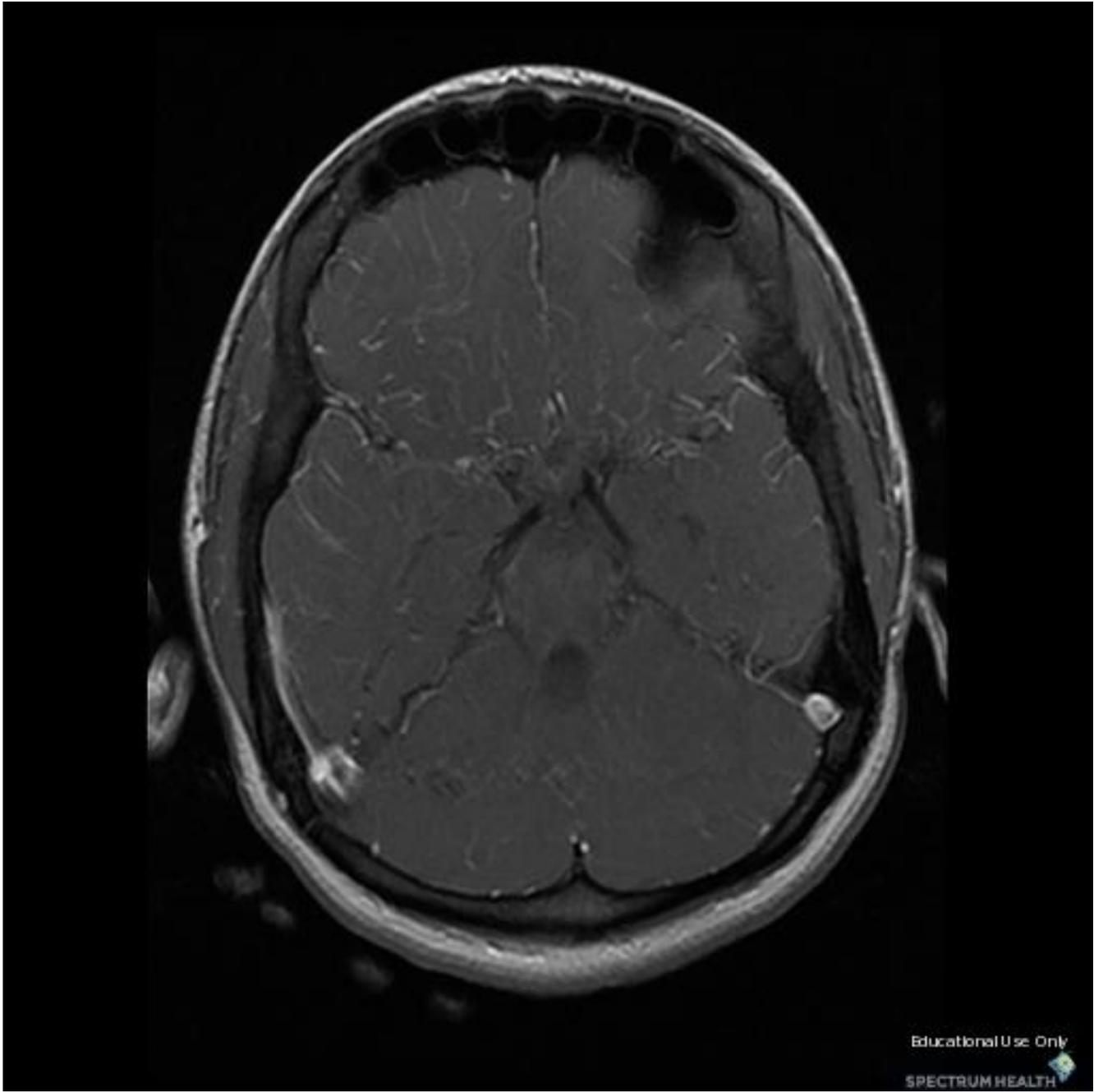
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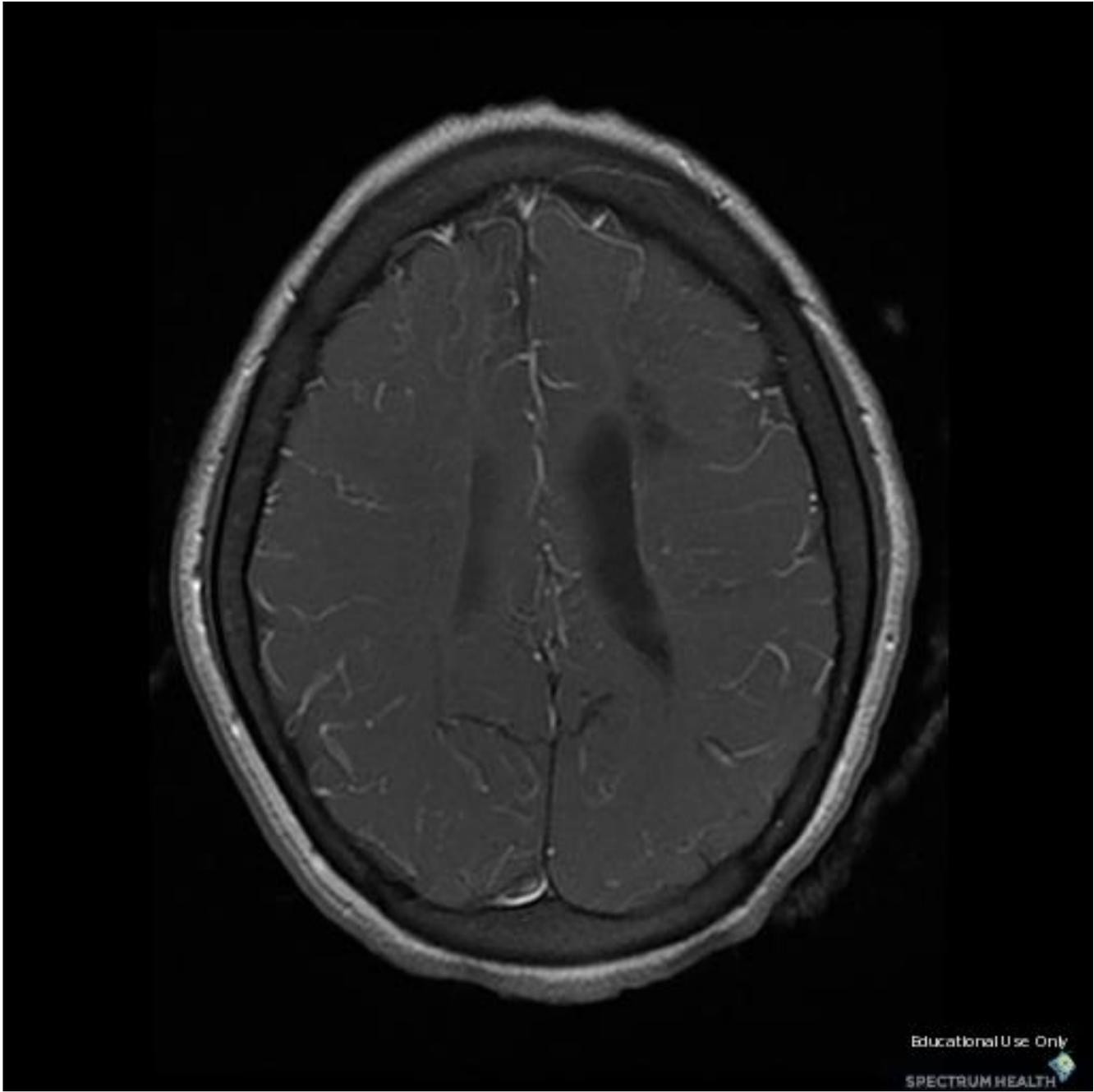


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