Case 14755

Eurorad

Sickle cell disease cerebral vasculopathy

Published on 10.06.2017

DOI: 10.1594/EURORAD/CASE.14755
ISSN: 1563-4086
Section: Neuroradiology
Area of Interest: Neuroradiology brain
Procedure: Education
Imaging Technique: MR
Imaging Technique: MR-Angiography
Imaging Technique: CT
Special Focus: Pathology Case Type: Clinical Cases
Authors: Jose Alejandro Bacalla MD, Angel Donato MD, Ramon Figueroa MD
Patient: 20 years, male

Clinical History:

20-year-old female patient with history of sickle cell disease and recurrent strokes, previous encephalo-duro-arterio-myo-synangiosis (EDAMS) was performed due to obstruction of both carotid arteries, presenting now with altered mental status.

Imaging Findings:

CT (Figure 1) shows bilateral temporal vertically oriented cranial defects along the coronal sutures from previous superficial temporal artery synangiosis. Diffuse expansion of the diploic spaces in the mandible, skull base and cranial vault are in keeping with diffuse bone marrow haematopoietic recruitment.

MR (Fig. 2) shows chronic encephalomalacia in bilateral superior frontal and left more than right perisylvian distributions in keeping with chronic hypoperfusion infarcts.

MR angiography (Fig. 3) shows high grade carotid occlusive disease with complete right internal carotid artery occlusion and a small calibre left internal carotid artery from cervical segment upwards ending in ophthalmic artery. There is no recognisable carotid siphon or supraclinoid segment on either side. There are prominent external carotid arteries with large calibre middle meningeal arteries and superficial temporal arteries contributing to transdural collaterals.

There are prominent vertically oriented, tortuous, bilateral lenticulostriate and thalamoperforator branches consistent with Moya-Moya Syndrome.

Discussion:

Sickle cell anaemia (SCA) cerebral vasculopathy is the result of endothelial dysfunction believed to result from dynamic barotrauma leading to endothelial damage, in combination with increased expression of adhesion molecules on erythrocytes and endothelial cell surfaces, interactions with leukocytes, increased levels of circulating inflammatory cytokines and enhanced vasa vasorum thrombosis [1].

In young children, vaso-occlusive crises most commonly manifest as dactylitis, a painful swelling of the hands,

fingers, feet, and toes [2]. Other problems in SCA include osteomyelitis, osteonecrosis, splenic infarct, splenic sequestration, acute chest syndrome, stroke, papillary necrosis and renal insufficiency. Up to 25% of SCA patients present with stroke in the first decade of life. 20% of them have white matter infarcts without clinical manifestations ("silent infarcts"). Without adequate blood transfusion treatment programs, these patients reach adulthood with cumulative stroke-related severe cognitive and motor deficits.

The radiological manifestations of SCA cerebral vasculopathy favour carotid occlusion at the top of the carotid siphon and proximal circle of Willis, resulting in cerebral infarcts, usually involving the deep grey matter and watershed areas. Compensatory dilatation of lenticulostriate and thalamoperforating arteries attempt to restore distal blood flow. The tiny perforators appear as a "puff of smoke" ("moyamoya" in japanese) by DSA. Cumulative chronic infarcts result in volume loss and areas of encephalomalacia. These patients express a higher prevalence of fusiform aneurysms, likely high-flow related, even in abnormal locations.

In addition to the thickened diploic space, there is bone marrow low signal from replacement of fatty marrow by red bone marrow. Diffusion-weighted images play a key role in differentiating acute from chronic infarcts.

The only curative treatment is bone marrow transplant, this option being limited due to HLA compatibility issues. Medical treatment includes long-term monthly blood transfusions in order to keep HbS less than 30% and use of hydroxyurea to prevent crises. Other options include encephalodural sinangiosis [3], a procedure in which the superficial temporal artery is attached to the dura in a non-anastomotic manner in order to increase collateral blood flow to hypoperfused areas of the cortex. This is only a palliative measure not preventing disease progression.

Sickle cell disease should always be considered as a diagnostic possibility in the setting of an African-American child with cerebral infarction.
Differential Diagnosis List: Sickle cell disease cerebral vasculopathy, Vasculitis, Idiopathic Moya Moya, Thalassaemia

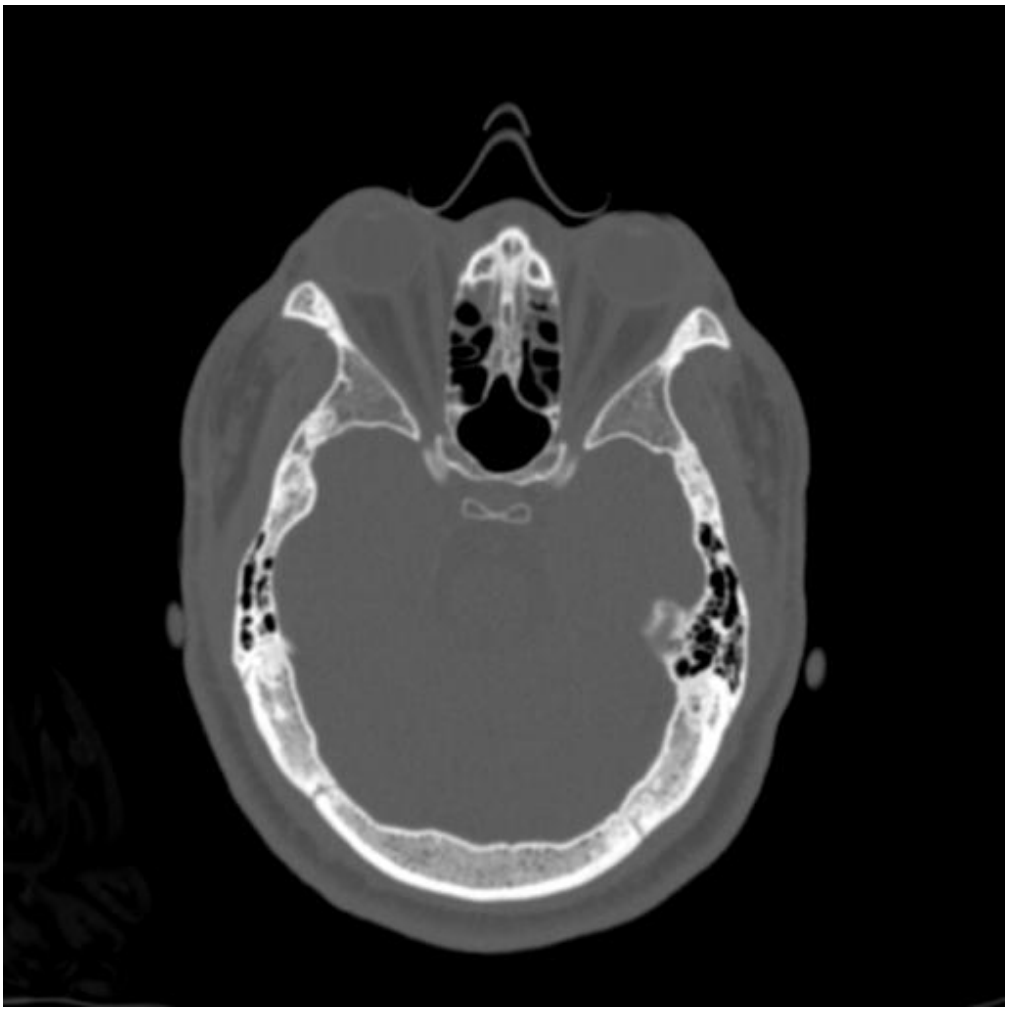
Final Diagnosis: Sickle cell disease cerebral vasculopathy

References:

Claudia Morris (2008) Mechanisms of Vasculopathy in Sickle Cell Disease and Thalassemia. Hematology January 1, 2008 vol. 2008 no. 1 177-185 (PMID: 19074078)
Yaster M, Kost-Byerly S, Maxwell LG (2000) The management of pain in sickle cell disease. Pediatr Clin North Am 47:699 –710 (PMID: 10835998)
Kinugasa (1993) Surgical treatment of moyamoya disease: operative technique for encephalo-duro-arterio-myo-synangiosis, its follow-up, clinical results, and angiograms. Neurosurgery Apr;32(4):527-31 (PMID:8474642)

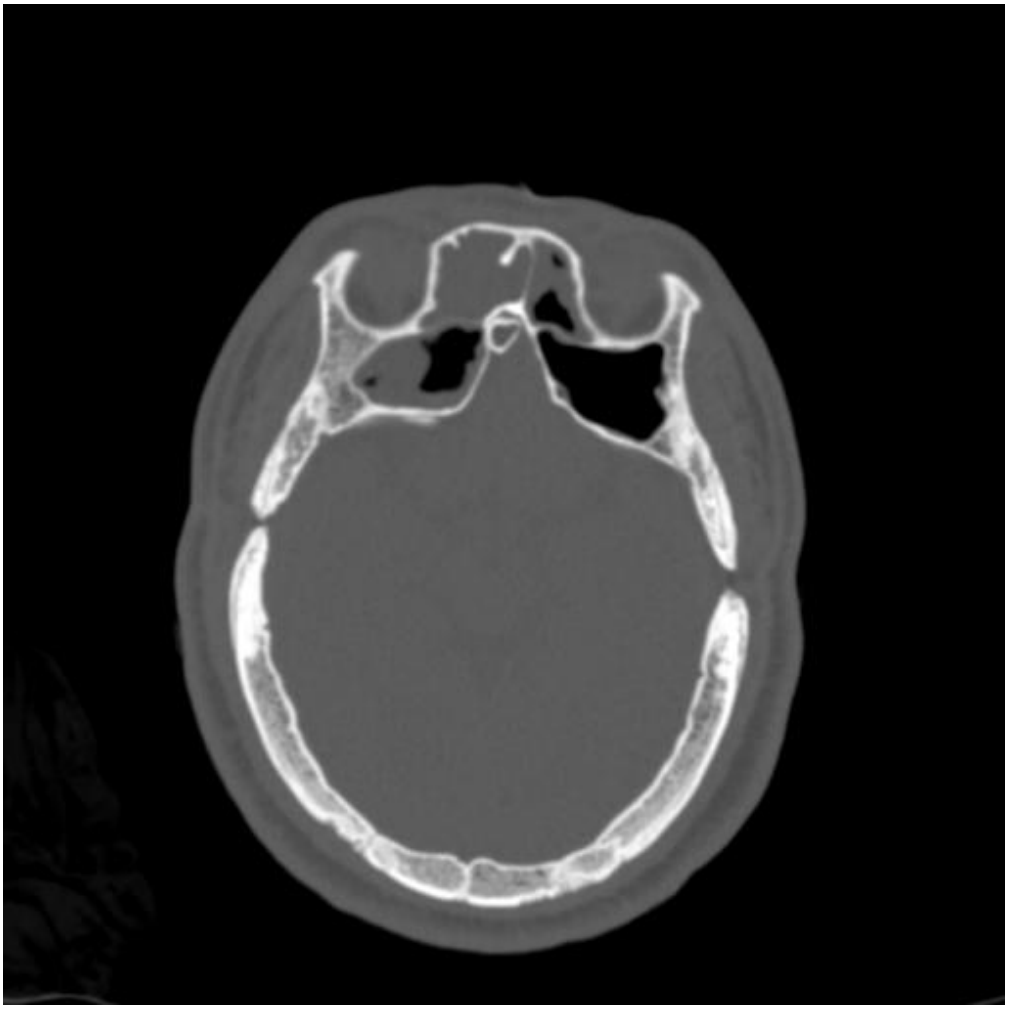
Figure 1

a



Description: There is global thickening of the calvarium, resulting from haematopoietic diploic space recruitment. There are also cortical defects at the coronal sutures, from prior EDAMS. **Origin:** Augusta University

b



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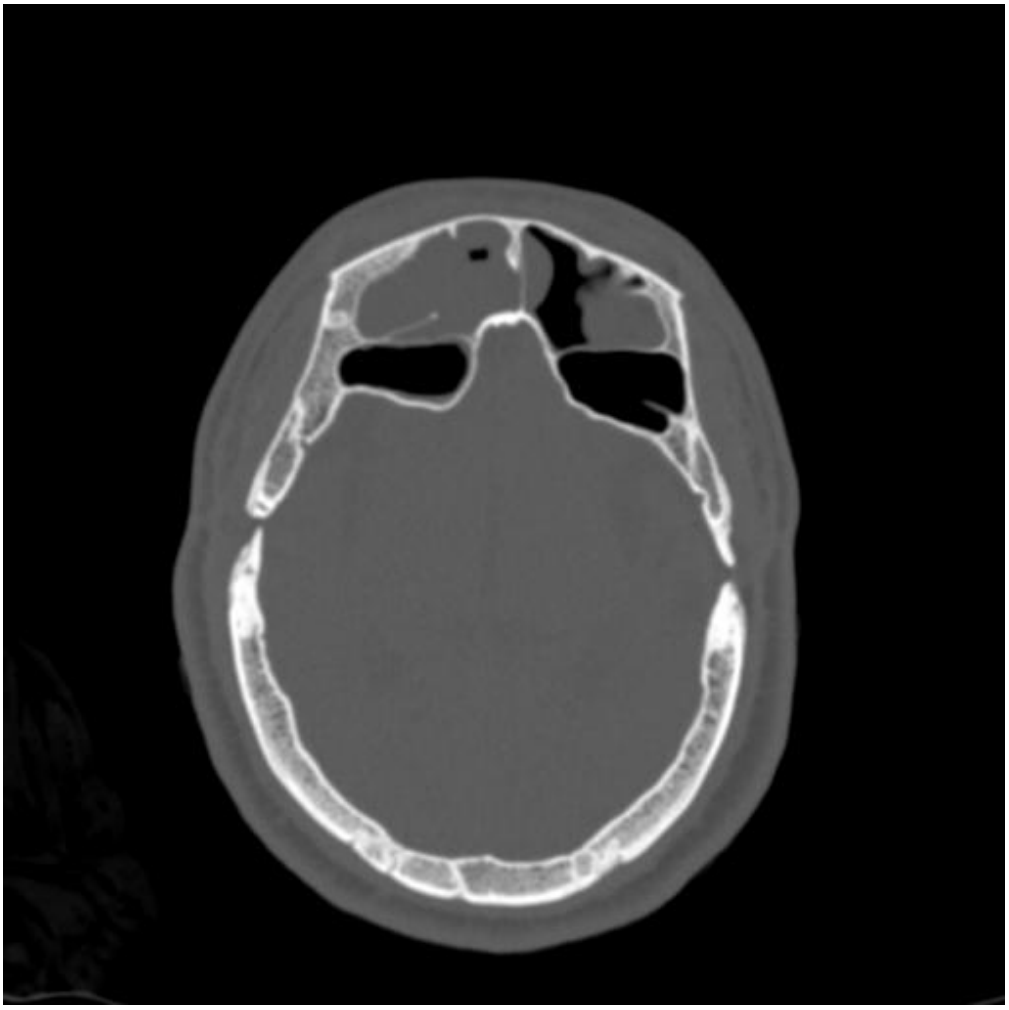

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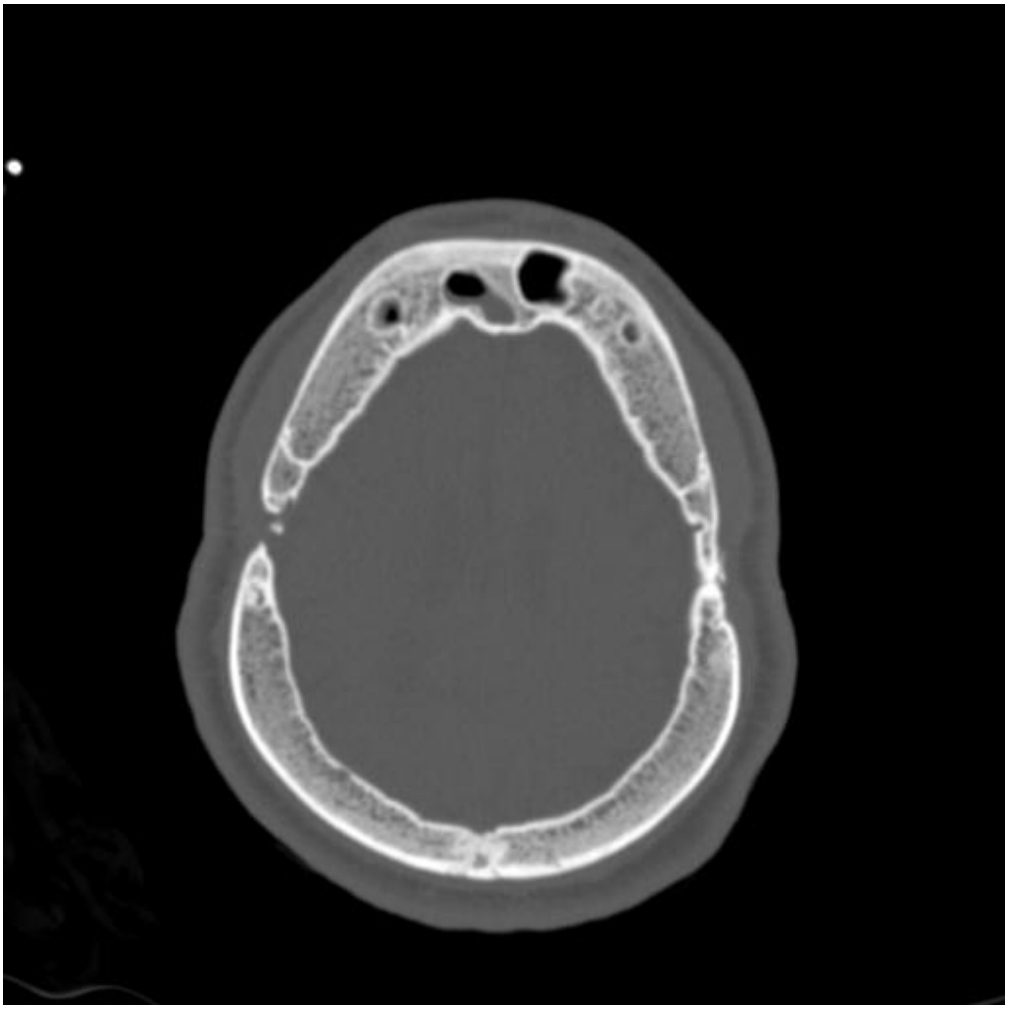

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Figure 2

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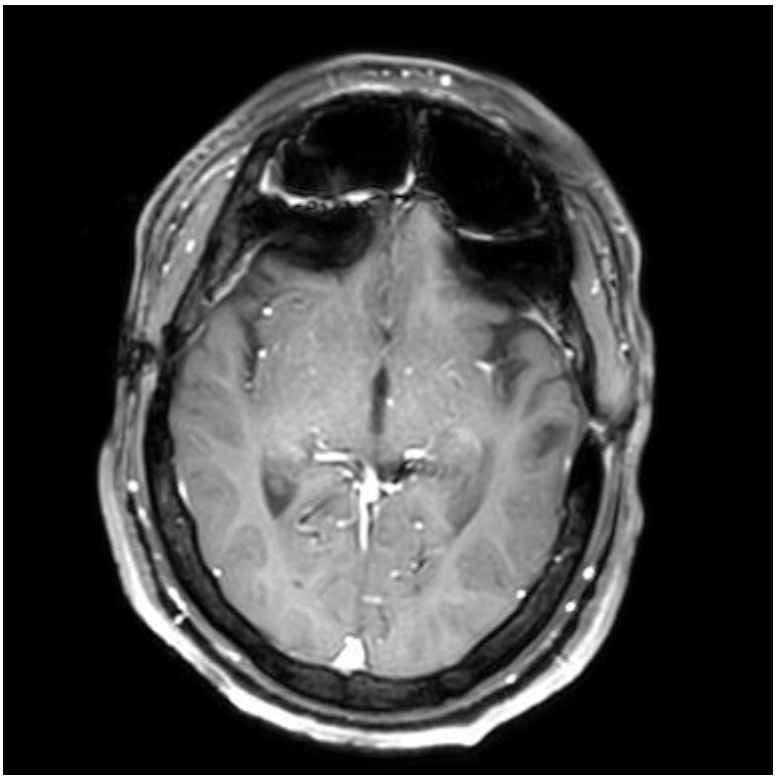
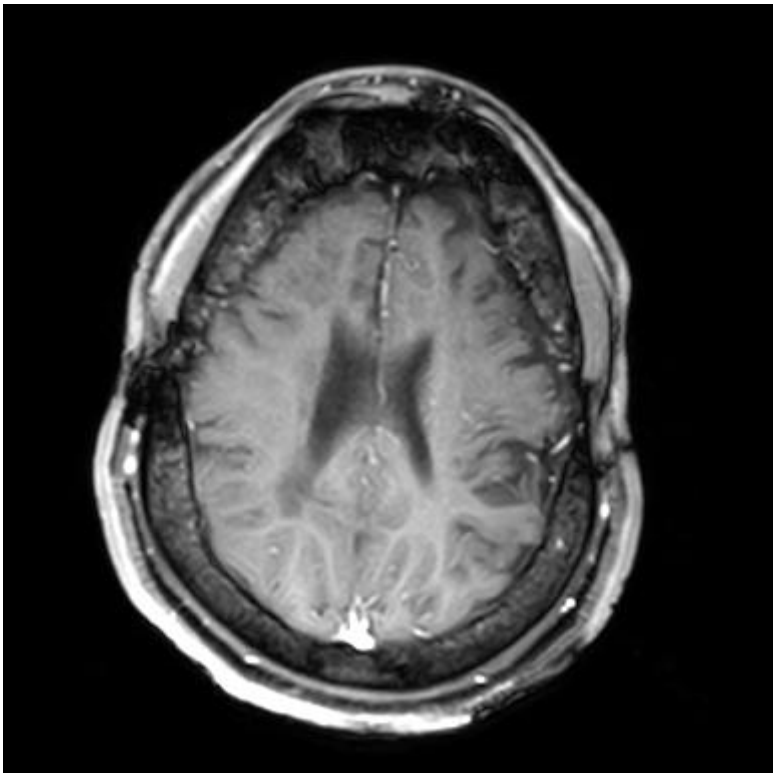


Description: There are vertically oriented bilateral temporal cranial defects along the coronal sutures from superficial temporal artery synangiosis. **Origin:** Augusta University

b



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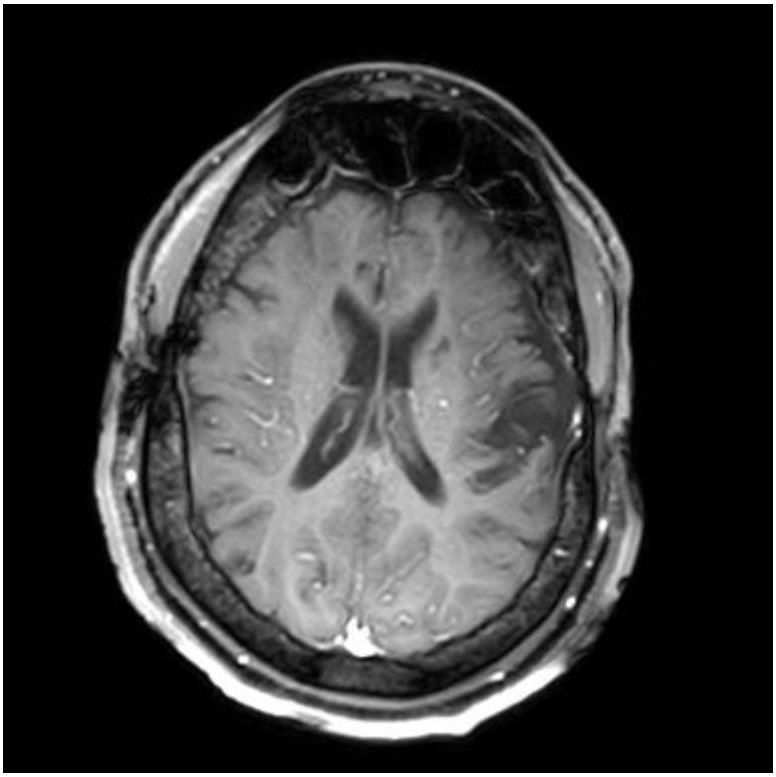

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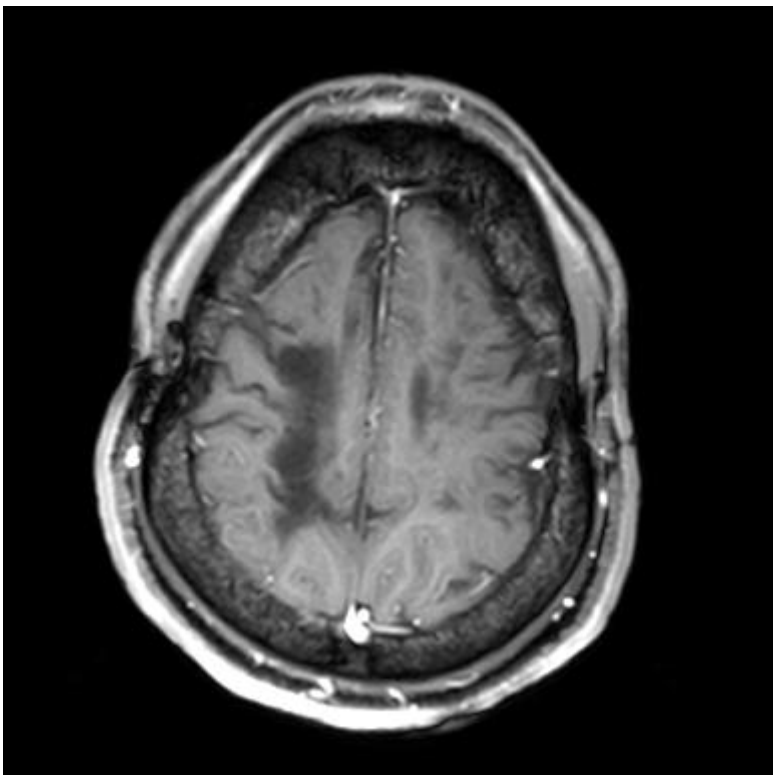

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Figure 3

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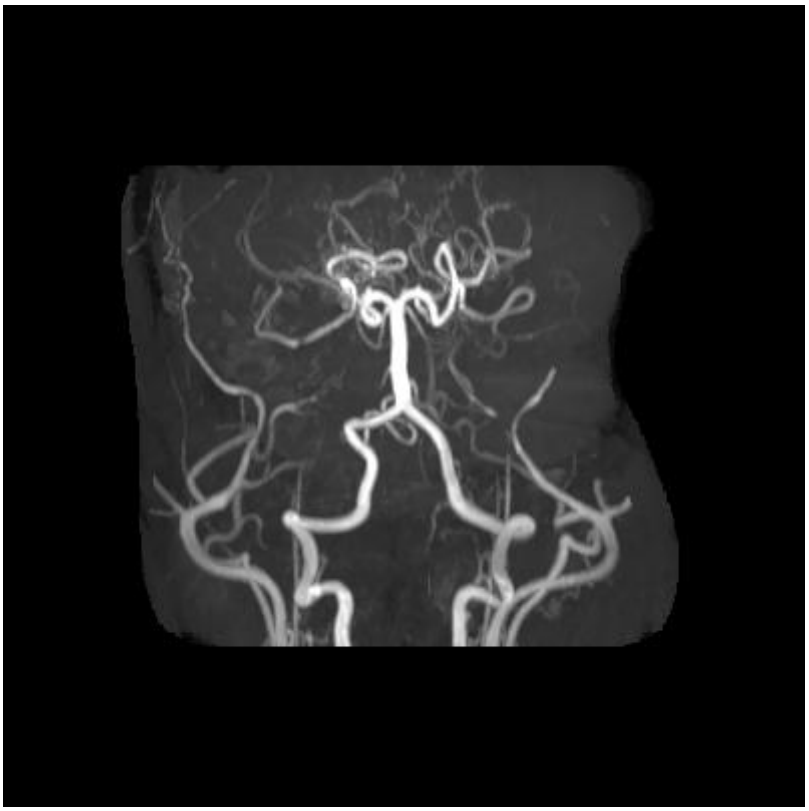
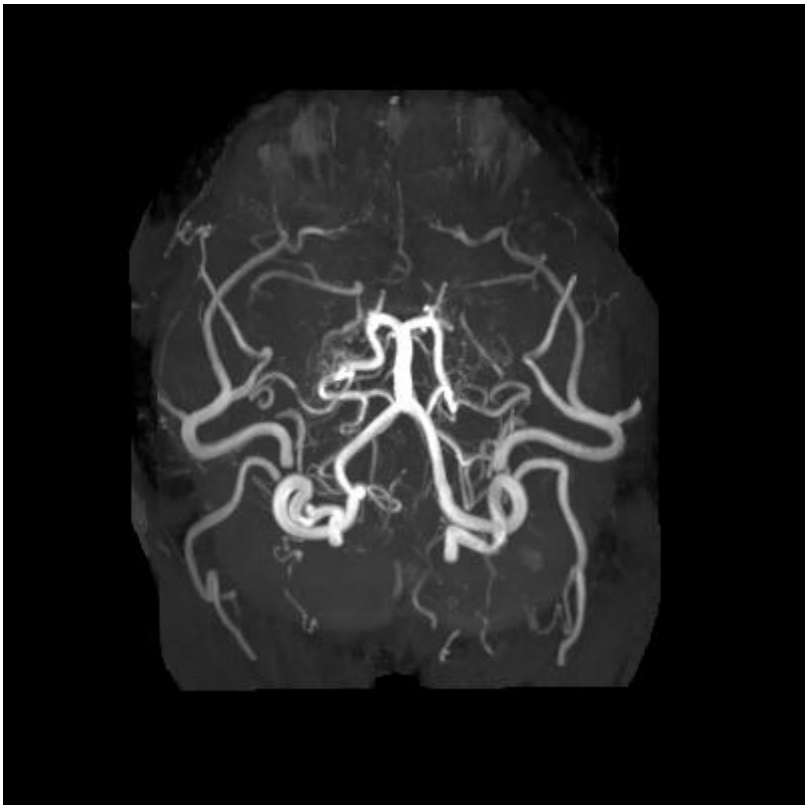


Description: Sickle cell vasculopathy with occlusion of internal carotid arteries and moyamoya pattern supported by posterior circulation from vertebrobasilar system. Notice patent bilateral superficial temporal artery synangiosis. **Origin:** Augusta University

b



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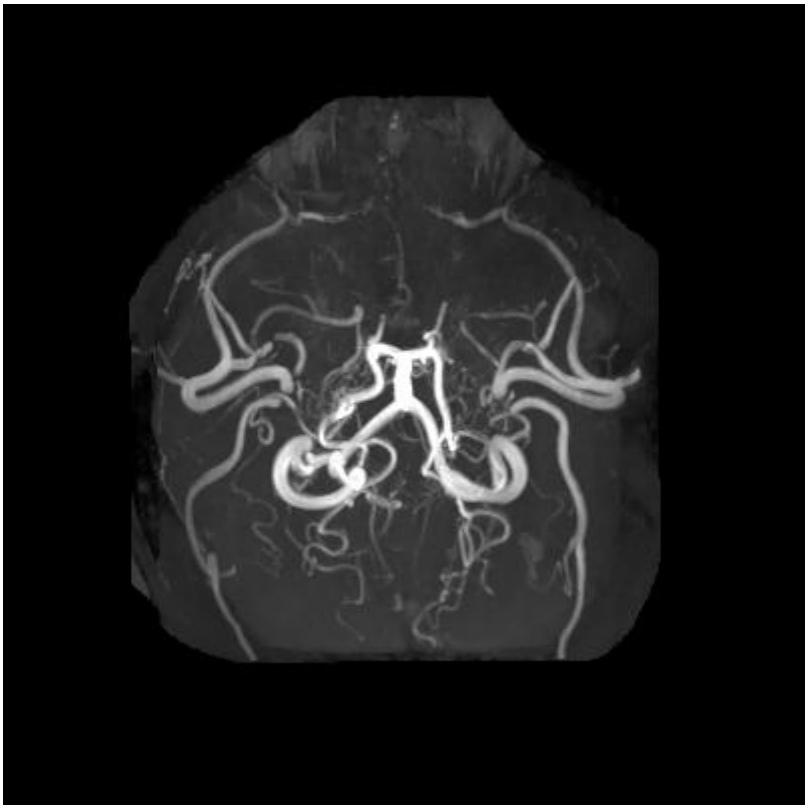

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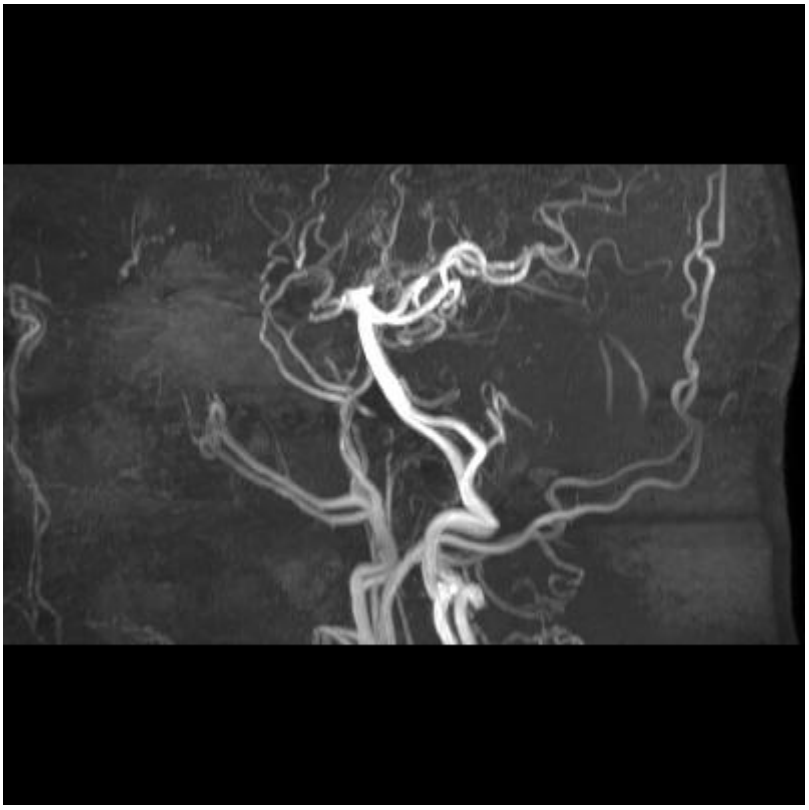

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