Reversible cerebral vasoconstriction syndrome as an initial presentation of SLE: a case report.

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Introduction:

Headache is a frequent symptom in patients with SLE, with prevalence similar to that of the general population; nonetheless, individuals with SLE are at a higher risk of experiencing headaches related to vascular issues such as cerebral venous sinus thrombosis, stroke, reversible cerebral vasoconstriction syndrome, posterior reversible encephalopathy syndrome, and vasculitis. Other potential causes of headaches in SLE patients include aseptic meningitis, neuroinfections, intracranial neoplasms, and intracranial hypertension or hypotension. This brings a challenge to physicians, as early diagnosis and treatment in such cases can alter the outcome of the disease (1).

Abstract:

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune multi-system disease. Despite the usual common insidious presentations of SLE, abrupt and life-threatening presentations still occur. Even with diagnostic challenges, early recognition and management can alter the disease course. Headache, a common symptom in the general population, can be a leading clue for the diagnosis, like in our patient. A 28-year-old African lady who was admitted for ovarian torsion and suddenly developed a severe headache and bilateral vision loss. An urgent MRI/MRA revealed subarachnoid haemorrhage, complicating reversible cerebral vasoconstriction syndrome. She was placed for angiography the next day, which showed luminal irregularities at distal intracranial arteries, considering vasculitis.

Case History and Examination:

A 28-year-old African lady was admitted under the supervision of the gynecology team for a left ovarian hemorrhagic cyst. On day 2 post surgery, while being recovered, she developed a sudden-onset thunderclap headache with bilateral vision loss. Examinations wise were positive for retinal microhemorrhages and hyperreflexia all over with pathological reflexes and mute planters.

Methods:
An urgent MRI was ordered, which revealed reversible cerebral vasoconstriction syndrome (RCVS) complicated by acute subarachnoid haemorrhage (figure A, B, C); hence, the patient was started on nimodipine. The case was referred to the neurosurgery team, who decided to proceed with Digital Subtraction Angiography that showed luminal irregularities at distal intracranial arteries considering vasculitis (figure D), reviewing the patient medical records showed that she presented recently to the emergency service with neck pain, severe anemia (6.8mg/dl , normal 12-15 mg/dl) , polyarthritis and arthralgia. She was treated symptomatically, transfused, and basic immunological works up were sent, with arranged follow up with a rheumatologist. These labs came positive for anti-nuclear antibodies with high titer (>=1:1280), anti-double strand DNA antibodies and low complements [C3 0.7 (normal: 0.9-1.8), C4 0.04 (normal 0.1-0.4)].

Results:
In light of the previously stated, the patient was started on nimodipine, pulse steroids 1 gram for 3 days followed by 40 mg/day, hydroxychloroquine and later mycophenolate. The patient's headache resolved, she regained her vision, yet she developed an episode of seizure, which was controlled by levetiracetam.
She was discharged on immunomodulators with arranged follow-ups.

Discussion:

The nervous system is one of the major organs affected by SLE, termed neuropsychiatric SLE (NPSLE), Despite its high prevalence, which ranges from 12 to 95% -10 , it has been a major challenge for clinicians to early diagnose and manage due to the highly variable presentations. Some of the presentations can be as simple as tension headaches, while other rare ones could be as deadly as intracranial hemorrhage. The variable presentations are probably attributed to a variety of pathological mechanisms (including thrombosis, autoantibodies, cytokines, and cell-mediated inflammation) 2. .SLE effect on the cerebral vasculature is an area of investigation, with many studies suggesting BBB dysfunction; such findings are suspected to play a role in anxiety and depression. 3-4. Small-vessel vasculitis is a rare occurrence in SLE, affecting approximately 1% of SLE patients. Its clinical presentation is similar to NPSLE but is rarely confirmed through brain biopsy, which is considered the gold standard test that carries a high surgical risk and low sensitivity. Cerebral angiography is another available option for diagnosis, revealing segmental narrowing and dilation in various vascular regions. However, this technique has limitations, as it can only identify large vessel involvement and may overlook small vessel disease.5 , A major deferential for NPSLE presenting with headache is reversible cerebral vasoconstriction syndrome (RCVS), a condition characterized by acute onset of thunderclap headache with or without neurologic symptoms, with radiological findings of “beading” of the affected blood vessels with a particular tendency to affect the posterior circulation. The

reversibility of the angiographic findings is a key component to the diagnosis. Distinguishing the exact cause of headache in such cases can be difficult as they can exhibit similar radiological findings; however, they have different modalities of treatment and carry a distinctive course of progression 6/7. Although SLE is frequently accused as being the cause for angiographically apparent cerebral vasculitis, only very few cases of SLE cerebral vasculitis have been reported. Despite the above, physicians remain hesitant to postpone immunosuppressive treatment. Our patient had a very recent presentation resembling active SLE, with radiological evidence of RCVS; hence, she was initially given nimodipine followed by pulse steroids, which resulted in clinical resolution of her symptoms. However, a previously observed trend for poor outcomes in such patients receiving steroids was reported in a retrospective analysis of similar cases, emphasizing the need for urgent and correct diagnosis 8.

Key clinical message:

Reversible cerebral vasoconstriction syndrome is rare and serious condition, which requires early recognition and direct taking action to prevent severe damage to the brain. Early using calcium channel blockers can result in better outcomes and avoid the potentially harmful effects of unnecessary steroid administration.

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The case approved by Hamad Medical Corporation Medical Research center and the patient signed written informed consent for the publication of any potentially identifiable images or data included in this article.

Authorship contributions
The authors contributed equally to the manuscript.

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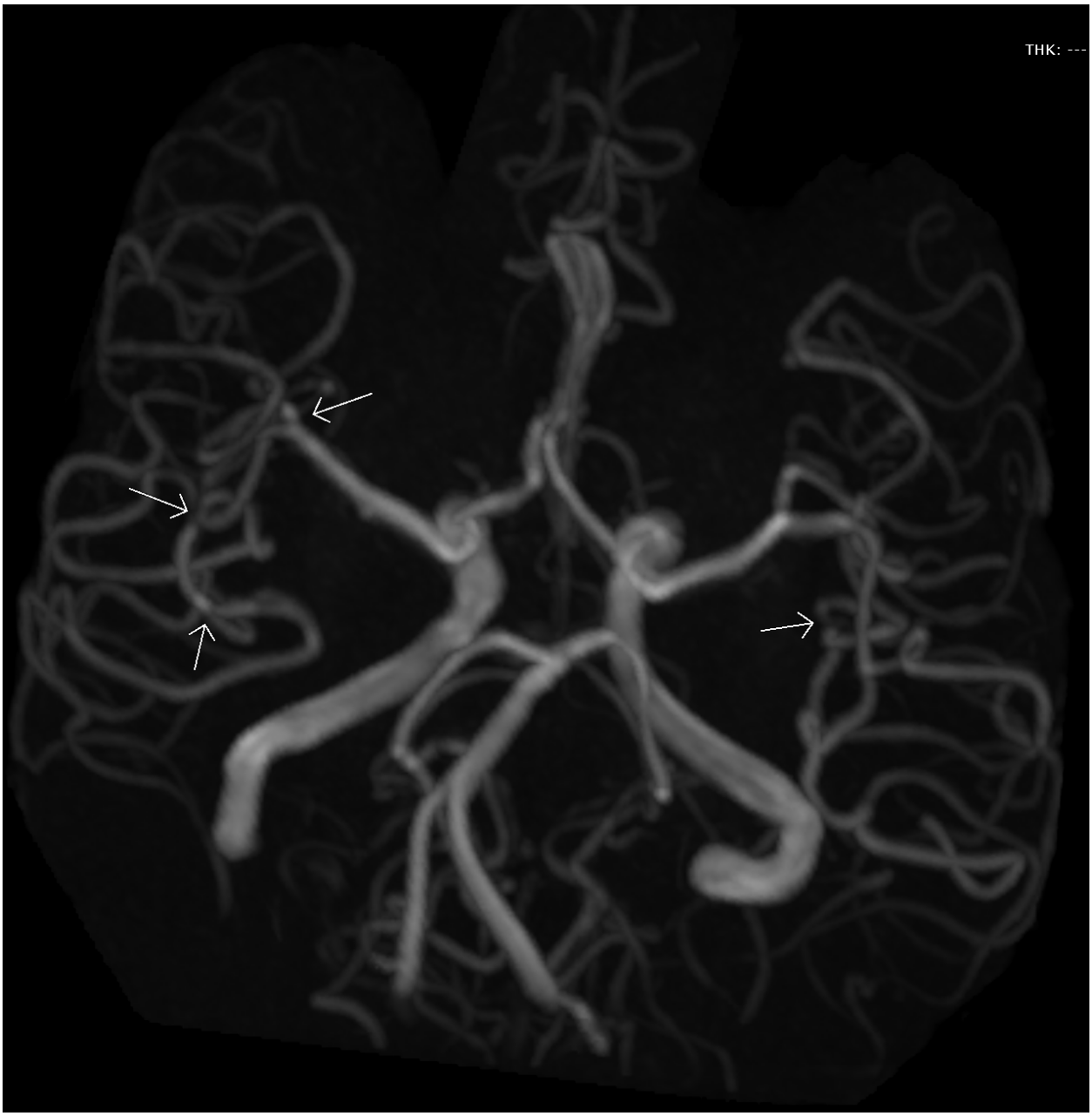

Figure A :
MRA Showing multifocal stenosis in the intracranial arteries as pointed by arrows

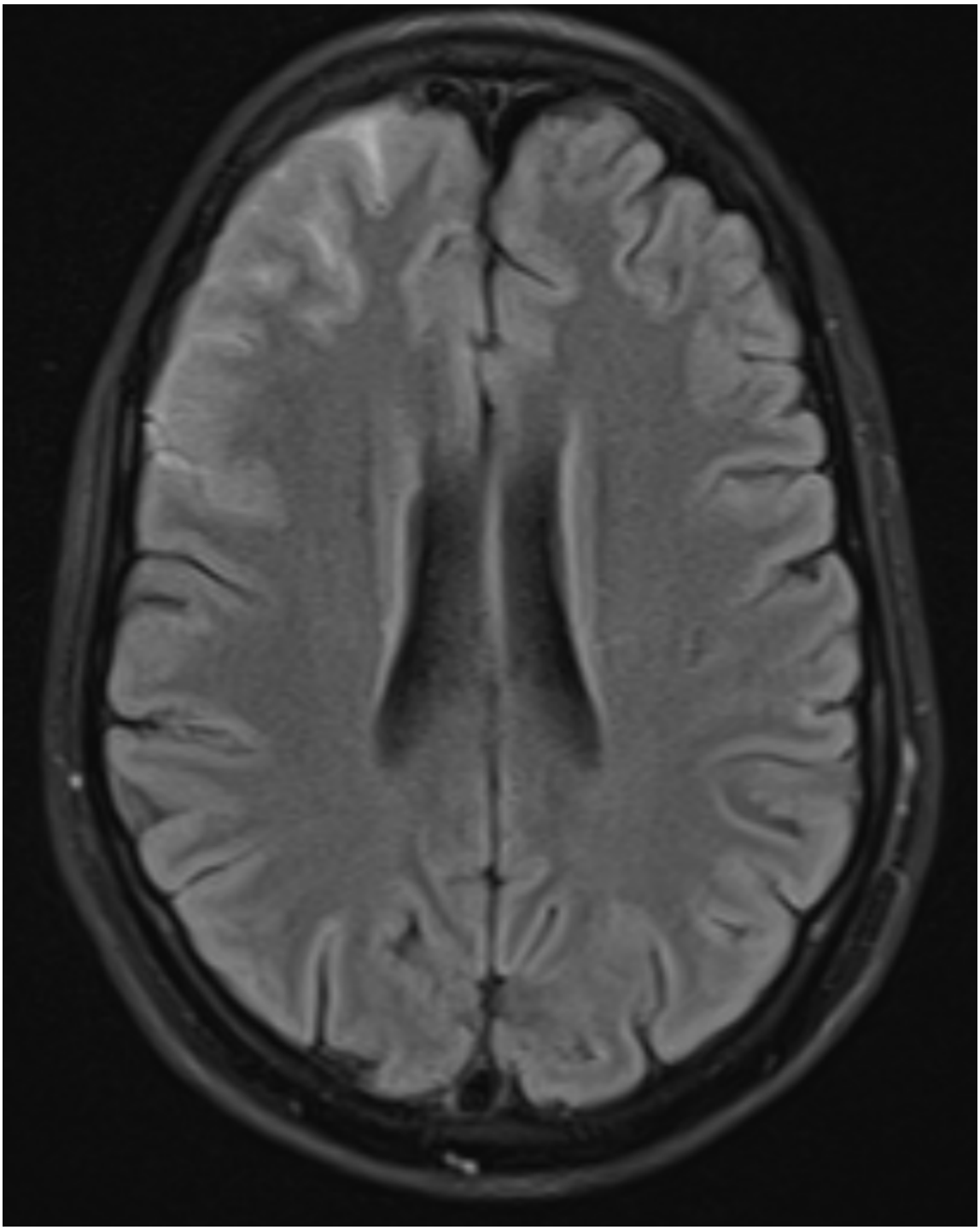

Figure B:
FLAIR sequence showing Right frontal convexity acute sub arachnoid hemorrhage

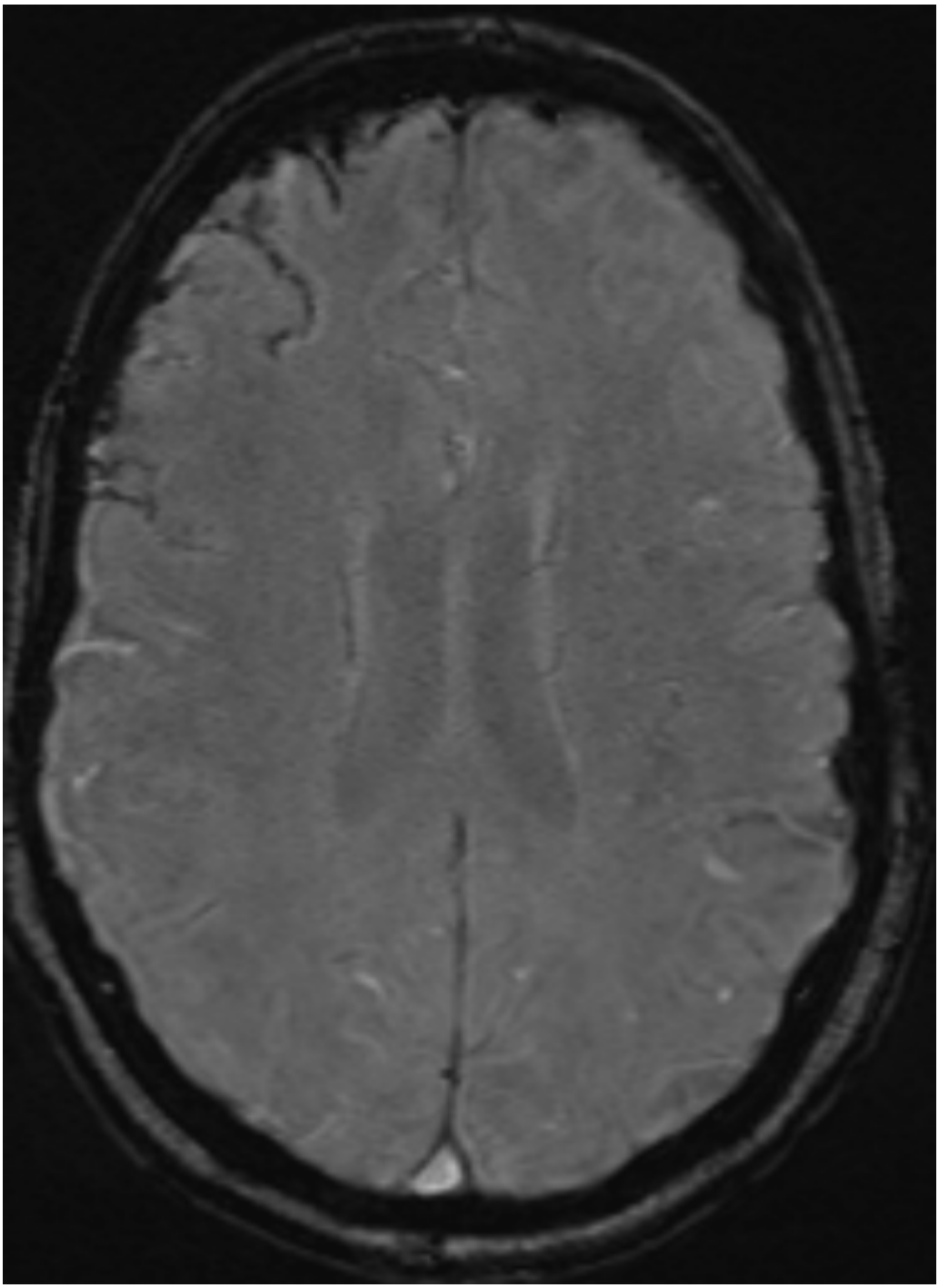

Figure C
SWI sequence showing Right frontal convexity acute sub arachnoid hemorrhage

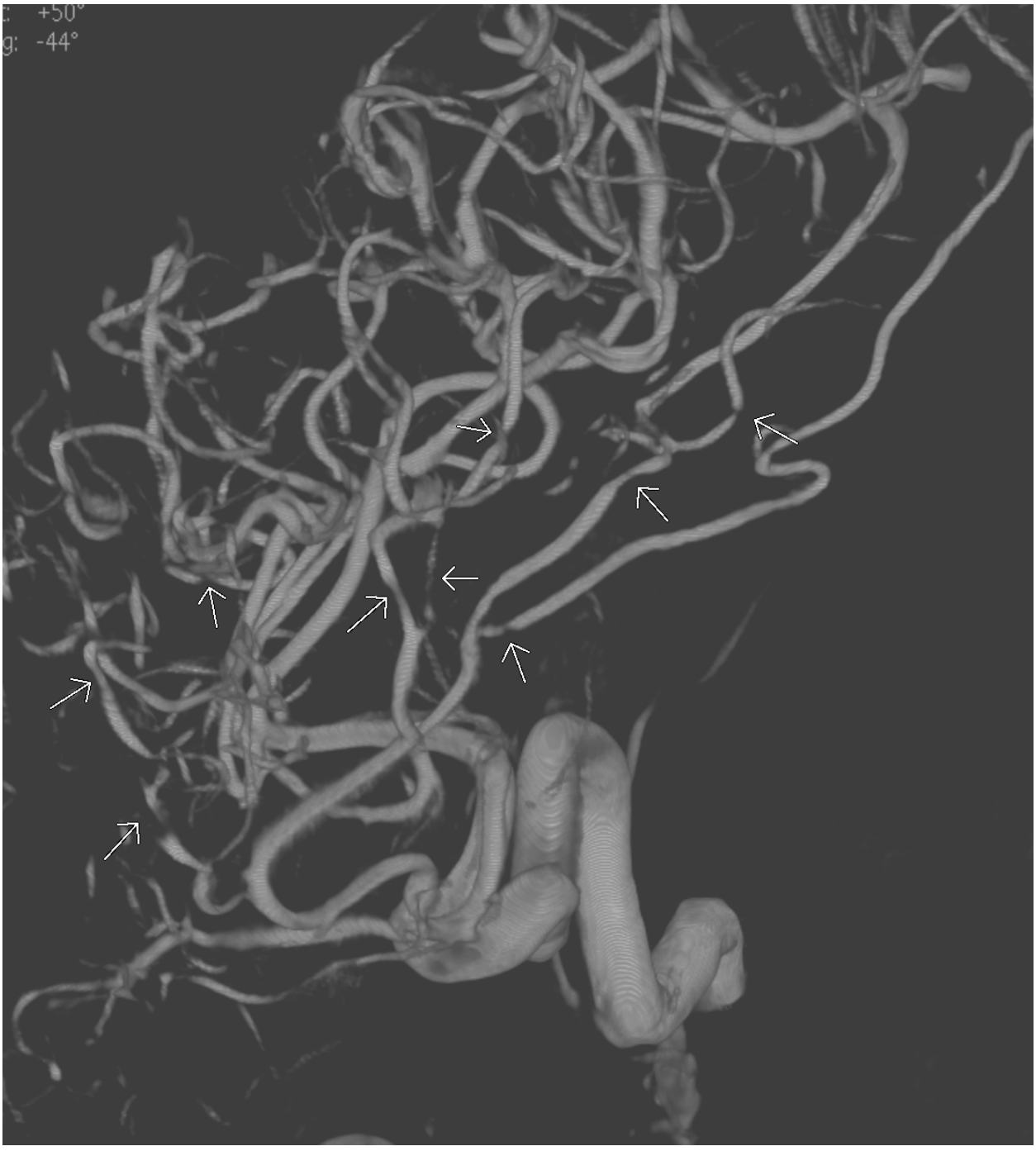

Figure D:
Digital subtraction Angiography showing multifocal stenosis in the intracranial arteries as pointed by arrows