

# A Case of a Giant Intracranial Arachnoid Cyst Mimicking Cerebrovascular Accident in a Sickle Cell Disease Patient

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## Abstract

Stroke or cerebrovascular accident (CVA) is a common vascular neurological condition. Many cranial lesions produce stroke-like features and may be mistaken for stroke. The aim of case report was created the awareness of the possibility of rare conditions such as this benign giant arachnoid cyst presenting with neurological symptoms mimicking CVA. We present a case of a 53-year-old female sickle cell disease patient who had features highly suggestive of CVA and had been previously managed as such before presenting to us. The patient had cranial magnetic resonance imaging as part of her neurologic evaluation, and this revealed a giant intracranial arachnoid cyst. She subsequently had craniotomy for excision of the mass lesion following which she had an uneventful postoperative recovery and sustained neurological improvement. This case illustrates similarities in the presentation of CVAs and intracranial arachnoid cysts. The value of brain imaging in the evaluation of patients with clinical features of CVA is also shown. This imaging was essential in arriving at an accurate differential diagnoses of CVAs and thus direct appropriate treatment.

**Key words:** Arachnoid cyst, giant, sickle cell disease

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## INTRODUCTION

Arachnoid cysts are benign congenital malformations along the craniospinal axis formed from splitting of the arachnoid layer with cerebrospinal fluid accumulation within the walls.<sup>1,2</sup> They have a predilection for the Sylvian fissure in the temporal fossa but are also found in the posterior fossa and suprasellar region, and on the cerebral convexity.<sup>1,3,4</sup> They are relatively common lesions of the arachnoid, with a reported prevalence as high as 1.1% in the adult population.<sup>5</sup> They are 2–3 times more common in males than females and may be bilateral, multiple, or multiloculated.<sup>1,2</sup> Arachnoid cysts have been documented to be associated with disorders such as tuberous sclerosis, autosomal dominant polycystic kidney disease, neurofibromatosis, Aicardi syndrome, and Proteus syndrome.<sup>6-8</sup> When symptomatic, intracranial arachnoid cysts present with features of space occupying lesions mimicking brain tumors, with the specific clinical features largely depending on the location and size of the cysts.<sup>1,2,9,10</sup>

With increasing availability of diagnostic facilities, in particular, magnetic resonance imaging (MRI), many arachnoid cysts are discovered incidentally.<sup>1,11</sup> We hereby present in this brief report, the clinical and radiological findings, as well as the management of a 53-year-old sickle cell disease (SCD) patient with a giant arachnoid cyst of the left frontoparietal region that was previously misdiagnosed as cerebrovascular accident (CVA).

## CASE REPORT

With the assurance of confidentiality, patient gave consent for her case to be reported.

OA is a 53-year-old female known SCD patient who presented to our neurosurgical outpatient clinic with a 2-year history of right sided weakness and gait impairment. Weakness was of sudden onset and progressively worsened over time. She had persistent dull-aching nonradiating left hemicranial

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headache and problems with word-finding when talking. She had no vomiting, seizures, blurred vision, tremors, auditory deficit, memory impairment, altered sensorium, or neck pain. She had no history of trauma, fever, drenching night sweat, anorexia, or weight loss. Examination findings included motor aphasia, right hemiparesis and hypoesthesia, right supranuclear facial paresis, and right hemiplegic gait. She had been managed as a case of CVA but with no improvement before presenting to us. Brain MRI scan [Figures 1 and 2] showed a massive (10 cm × 12 cm × 15 cm) left frontoparietal nonenhancing hypointense cystic lesion on T1-weighted image with high signal intensity on T2-weighted image, significant midline shift to the right, effacement of the left lateral and third ventricles, and mass effect on the surrounding cerebral tissue. A diagnosis of giant left frontoparietal arachnoid cyst was made. She had craniotomy and excision of the arachnoid cyst. She had an uneventful postoperative recovery with improvement in her speech and in power on the right side. She had sustained neurological gain at the surgical outpatient clinic follow-up.

## DISCUSSION

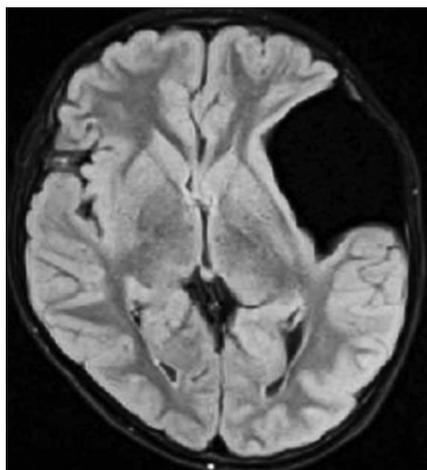
SCD is common in malaria endemic areas and occurs when an individual is homozygous for the sickle hemoglobin (HbS) mutation or is a compound heterozygote for HbS and  $\beta$ -thalassemia, hemoglobin C, or some less common  $\beta$ -globin mutations.<sup>12</sup> The disease is characterized by hemolytic anemia and three types of crises: Painful vaso-occlusive, sequestration, and aplastic. Complications include splenic infarction, autosplenectomy, bone infarcts, aseptic necrosis of the femoral head, leg ulcers, priapism, pulmonary hypertension, and renal failure.<sup>13</sup> Neurological complications such as hemorrhagic and ischemic CVAs (strokes) are also seen in SCD patients.<sup>12,13</sup> Since vascular complications are common in these patients,<sup>13</sup> it is usual to think of such when they present with suggestive clinical features.

Ay *et al.* investigated 782 patients with stroke-like deficits and found 27 of them to have normal diffusion-weighted MRI without imaging features of stroke. Of these 27, one was found to have a left temporal arachnoid cyst. He was a 58-year-old man who had presented with motor aphasia.<sup>14</sup> This was particularly similar to our patient who was in the same decade of life and also had aphasia as one of her symptoms. Others have reported patients who have intracranial arachnoid cysts presenting with headache,<sup>15</sup> also, a major symptom our patient had.

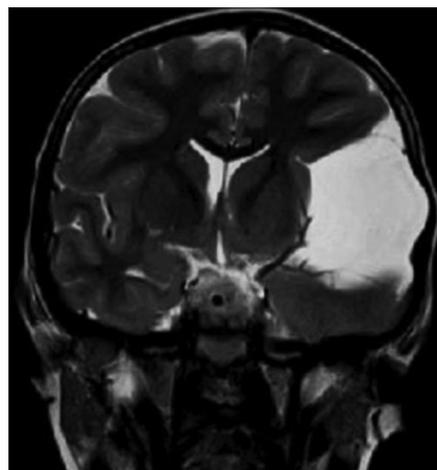
Our patient's cyst was found in the Sylvian fissure, the most common intracranial location of arachnoid cysts reported in the literature.<sup>1,3,4</sup> Cranial imaging findings as seen in other case reports have also shown this predilection of intracranial arachnoid cysts for the middle fossa<sup>16</sup> as well as the mass effect they can cause.<sup>16-18</sup> Arachnoid cysts presenting such as stroke have not been previously reported in SCD patients.

Options of treatment of symptomatic arachnoid cysts, whether associated with SCD or not, include resection of the cyst walls, endoscopic or microsurgical fenestration of the cyst to the subarachnoid cisterns, and shunting techniques such as ventriculocystostomy, cystocisternostomy, and cystoperitoneal and cystosubdural shunts which have all been tried with varying success rates.<sup>1,2,19,20</sup>

This case further emphasizes the importance of having imaging studies of the brain done when a diagnosis of stroke is entertained, even when the presence of factors such as SCD strongly support the diagnosis. Misdiagnosis of stroke has significant implication for definitive therapy. The patients with clinical diagnosis of stroke should, therefore, have at least brain computed tomography scan evaluation to ensure that patients who can be helped surgically are identified early and appropriately treated.<sup>21</sup> The prompt and correct definitive diagnosis with timely adequate management will ensure a better outcome.



**Figure 1:** An axial T1-weighted image in which the cyst is seen as a hypointense lesion occupying the left Sylvian fissure with mass effect on the ipsilateral temporal and frontal lobes, insula, basal ganglia, internal capsule and thalamus.



**Figure 2:** A coronal T2-weighted image in which the cyst is now seen to be hyperintense with mass effects as described for Figure 1 and effacement of the frontal horn of the ipsilateral lateral ventricle. Both figures also show a midline shift to the right.

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## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

1. Wetjen NM, Walker ML. Arachnoid cysts. In: Winn HR, editor. Youmans Neurological Surgery. 6<sup>th</sup> ed., Vol. 2. Philadelphia: Elsevier Saunders; 2011. p. 1911-7.
2. Balasubramaniam C, Santosh V. Intracranial arachnoid cysts. In: Tandon PN, Ramamurthi R, editors. Textbook of Neurosurgery. 3<sup>rd</sup> ed., Vol. 1. New Delhi: Jaypee Brothers Medical Publishers (P) Ltd.; 2012. p. 208-23.
3. Rengachary SS, Watanabe I. Ultrastructure and pathogenesis of intracranial arachnoid cysts. *J Neuropathol Exp Neurol* 1981;40:61-83.
4. Pradilla G, Jallo G. Arachnoid cysts: Case series and review of the literature. *Neurosurg Focus* 2007;22:E7.
5. Vernooij MW, Ikram MA, Tanghe HL, Vincent AJ, Hofman A, Krestin GP, *et al.* Incidental findings on brain MRI in the general population. *N Engl J Med* 2007;357:1821-8.
6. Alehan FK, Gürakan B, Agildere M. Familial arachnoid cysts in association with autosomal dominant polycystic kidney disease. *Pediatrics* 2002;110 (1 Pt 1):e13.
7. Hunter AG. Arachnoid cysts. In: Stevenson RE, Hall JG, editors. Human Malformations and Related Anomalies. 2<sup>nd</sup> ed. New York: Oxford University Press; 2006. p. 689-97.
8. Jimenez DF, Savage JG, Samuelson M. Developmental anomalies: Arachnoid cysts, dermoids and epidermoids. In: Ellenbogen RG, Abdulrauf SI, Sekhar LN, editors. Principles of Neurological Surgery. 3<sup>rd</sup> ed. Philadelphia: Elsevier Saunders; 2012.
9. Harsh GR 4<sup>th</sup>, Edwards MS, Wilson CB. Intracranial arachnoid cysts in children. *J Neurosurg* 1986;64:835-42.
10. Parsch CS, Krauss J, Hofmann E, Meixensberger J, Roosen K. Arachnoid cysts associated with subdural hematomas and hygromas: Analysis of 16 cases, long-term follow-up, and review of the literature. *Neurosurgery* 1997;40:483-90.
11. Katzman GL, Dagher AP, Patronas NJ. Incidental findings on brain magnetic resonance imaging from 1000 asymptomatic volunteers. *JAMA* 1999;282:36-9.
12. Hebbel RP. Pathobiology of sickle cell disease. In: Hoffman R, Benz EJ, Silberstein LE, Heslop H, Weitz J, Anastasi J, editors. Hematology: Basic Principles and Practice. 6<sup>th</sup> ed. Philadelphia: Elsevier Saunders; 2013. p. 536-47.
13. Sauntharajah Y, Vichinsky EP. Sickle cell disease: Clinical features and management. In: Hoffman R, Benz EJ, Silberstein LE, Heslop H, Weitz J, Anastasi J, editors. Hematology: Basic Principles and Practice. 6<sup>th</sup> ed. Philadelphia: Elsevier Saunders; 2013. p. 548-72.
14. Ay H, Buonanno FS, Rordorf G, Schaefer PW, Schwamm LH, Wu O, *et al.* Normal diffusion-weighted MRI during stroke-like deficits. *Neurology* 1999;52:1784-92.
15. Kandogan T, Olgun L, Gültekin G, Aydar L, Sezgin O. A suprasellar arachnoid cyst destructing sphenoid sinus: An unusual cause of headache in an elderly female. *Swiss Med Wkly* 2004;134:28-9.
16. Mondal T, Kumar KJ, Anand R, Dubey NK, Sharma D. Large arachnoid cyst. *Indian Pediatr* 1995;32:927-9.
17. Laribi S, Chakroun O, Segal N, Hamdi S, Plaisance P. Intracranial symptomatic giant arachnoid cyst. *Signs of Life* 2012;7:43-5.
18. Gabelic T, Klepac N, Mubrin Z, Ozretic D, Habek M. Giant arachnoid cyst. *Acta Neurol Belg* 2009;109:244.
19. Boutarbouch M, El Ouahabi A, Rifi L, Arkha Y, Derraz S, El Khamlichi A. Management of intracranial arachnoid cysts: Institutional experience with initial 32 cases and review of the literature. *Clin Neurol Neurosurg* 2008;110:1-7.
20. Ciricillo SF, Cogen PH, Harsh GR, Edwards MS. Intracranial arachnoid cysts in children. A comparison of the effects of fenestration and shunting. *J Neurosurg* 1991;74:230-5.
21. Ogun SA, Oluwole O, Ogunseyinde AO, Fatade B, Odusote KA. Misdiagnosis of stroke – A computerised tomography scan study. *West Afr J Med* 2000;19:19-22.