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SHORT REPORT



## Diffuse cerebral angiomatosis associated to basilar apex aneurysm

Giuseppe Emmanuele Umana<sup>a</sup> , Angelo Spitaleri<sup>a</sup>, Marco Fricia<sup>a</sup>, Ignazio Indorato<sup>b</sup>, Giovanni Federico Nicoletti<sup>c</sup> , Salvatore Cicero<sup>a</sup> and Gianluca Scalia<sup>c</sup> 

<sup>a</sup>Department of Neurosurgery, Trauma Center, Gamma Knife Center, Cannizzaro Hospital, Catania, Italy; <sup>b</sup>Department of Radiology, S. Elia Hospital, Caltanissetta, Italy; <sup>c</sup>Department of Neurosurgery, Highly Specialized Hospital and of National Importance "Garibaldi", Catania, Italy

### ABSTRACT

**Background:** Diffuse cerebral angiomatosis (DCA) is a diffuse infiltration of normal brain by complex vascular structures. It differs from arteriovenous malformations (AVMs) that are composed of a nidus of vessels through which arteriovenous shunting occurs without interposed functional brain parenchyma. A rare subgroup of AVMs is diffuse with no recognizable nidus with functional neuronal tissue interspersed within the malformed vessels. We present a rare association of DCA and cerebral arterial aneurysm, which dramatically influenced the patient's prognosis.

**Case description:** A 43-year-old male patient with right hemispheric diffuse cerebral angiomatosis presented with a ruptured basilar tip aneurysm that was successfully embolized. Unfortunately, the patient developed a locked-in syndrome.

**Conclusion:** The present report shows a possible association between diffuse cerebral angiomatosis and cerebral aneurysms, but this association appears to be less strong than it is with other AVMs.

### ARTICLE HISTORY

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### KEYWORDS

Diffuse cerebral angiomatosis; proliferative cerebral angiopathy; hemispheric vascular malformation; aneurysm; subarachnoid hemorrhage; angiography

### Introduction

Diffuse cerebral angiomatosis (DCA) is characterized by a diffuse infiltration of normal brain parenchyma by complex vascular structures. It differs from AVMs that are composed of a nidus through which arteriovenous shunting occurs without interposed functional brain parenchyma. A rare subgroup of AVMs is diffuse with no recognizable nidus and functional neuronal tissue interspersed between malformed vessels.<sup>1-3</sup> Multiple AVMs usually present in patients affected by Osler-Weber-Rendu, Divry-Van Bogaert or Sneddon's syndromes<sup>4,5</sup> and DCA is distinct from these. Few case reports have been documented to date.<sup>3,6</sup> In the literature DCA is identified in 3.4% of patients affected by AVMs, with an age ranging from 10 to 65-year-old, frequently in young females (67%).<sup>7</sup> Clinically, patients affected by DCA usually present epilepsy, headache and progressive rather than an acute neurological deficit, rarely with hemorrhage, unlike classical brain AVMs.<sup>8</sup> There is only one report of fatal hemorrhage in an elderly female.<sup>9</sup> DCA is more common in adolescent and middle-aged females; presentations include seizures, headaches, and stroke-like symptoms due to incompetent angiogenesis.<sup>7,10,11</sup> These vascular anomalies are usually larger than 6 cm and dural supply is a common finding.<sup>7</sup> There are of possible progression.<sup>10</sup>

DCA presents a unique therapeutic challenge because the nidus is interspersed with normal brain. Functional studies including perfusion-weighted imaging (PWI) or single-photon emission computed tomography (SPECT) have shown impaired perfusion within the vascular malformation as well as the lesional areas<sup>7,12,13</sup> likewise to the 'ischemia' in moyamoya disease. Revascularization surgery has been proposed to increase cortical blood supply<sup>9,14,15</sup> with encouraging results. Gamma knife

radiosurgery is reported to partially relieved headaches, but the other neurological deficits did not improve<sup>[8]</sup>. Thus, there is still no clear indication for the type of treatment for DCA.

### Case presentation

A 43-year-old male patient with a history of epilepsy and stroke with left hemiplegia. The diagnosis of right hemispheric DCA was made at the age of 36. The patient was referred to the emergency department after an episode of sudden headache and loss of consciousness, Hunt-Hess grade 4. The patient was sedated, intubated and performed a brain angio-CT scan with 3D-reconstruction revealed the known hemispheric DCA with extracranial vascular abnormalities (Figure 1) that were unchanged from previous studies and massive subarachnoid hemorrhage (SAH) Fisher grade 3, with a basilar tip aneurysm (Figure 2). Angiography confirmed the vascular malformation as unchanged (Figure 3) and defined the new irregular, wide necked 8 mm basilar aneurysm (Figure 4). The aneurysm was embolized with coils and a Scepter XC<sup>®</sup> (MicroVention Europe S.A.R.L., Saint-Germain-en-Laye, France) 4 × 11 mm occlusion balloon catheter remodeling. The final angiographic checks show the almost complete exclusion of the aneurysm. (Figure 5), no procedural complications were documented. The patient was awakened after two weeks, but unfortunately, he presented looked-in syndrome symptoms due to vasospasm as documented by transcranial Doppler. Then he was addressed to a high-specialized center for rehabilitation for critical neurological patients. A fluid-attenuated inversion recovery (FLAIR) MRI showed large feeding arteries in the context of the lesion and surrounding edema. (Figure 6)

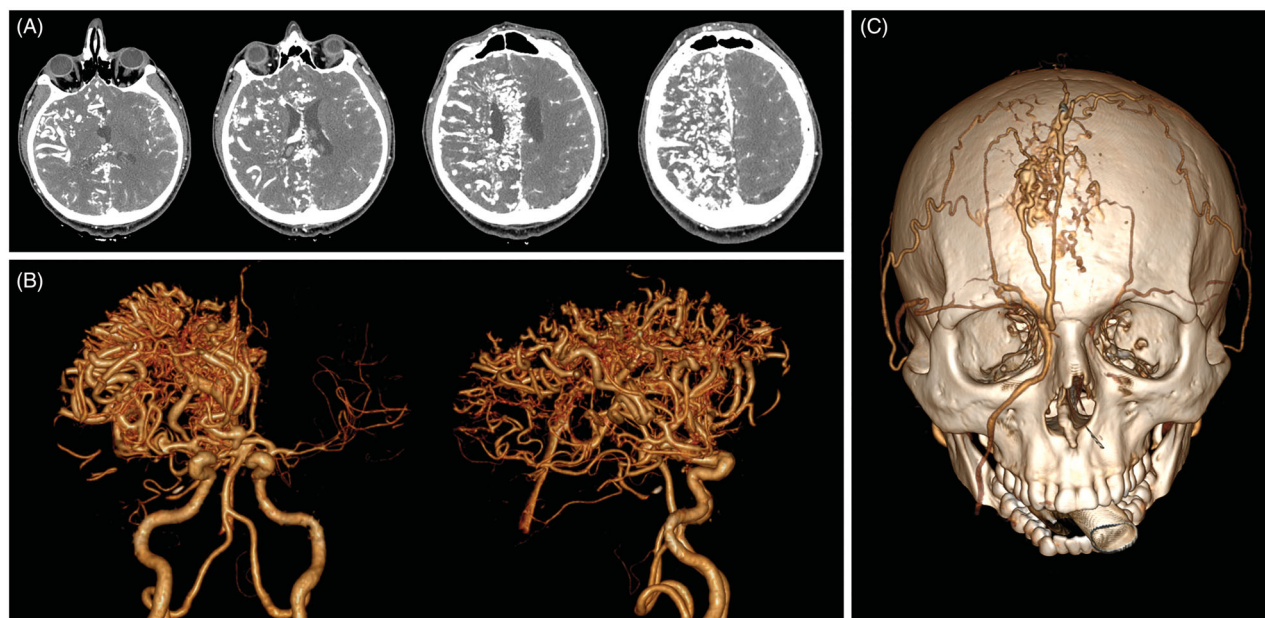


Figure 1. Seriated axial head CT angiography images showed a diffuse network of densely enhancing vascular spaces throughout the right hemisphere (A). Angio-TC 3D reconstruction of the diffuse cerebral angiopathy in lateral and anterior views (B). 3D skull reconstruction with extracranial vascular abnormalities related to diffuse cerebral angiopathy (C).

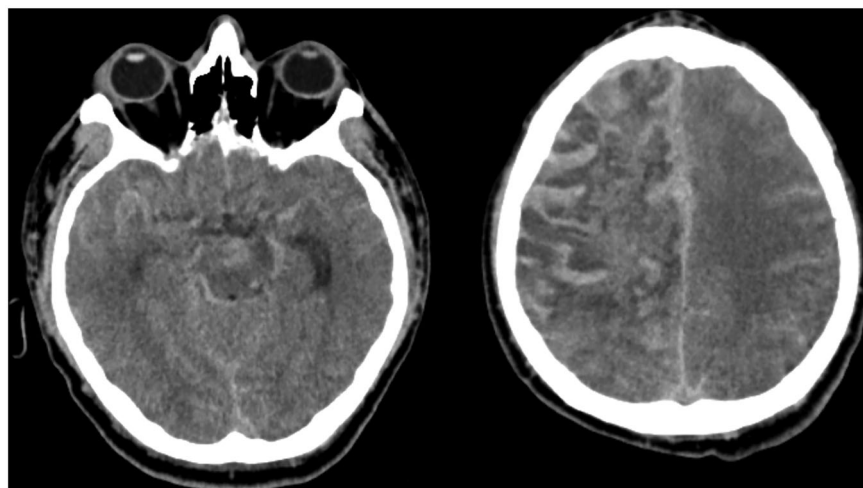


Figure 2. Axial head CT images showed a diffuse and thick subarachnoid hemorrhage (Fisher 3) caused by a ruptured basilar artery aneurysm.

## Discussion

Differential diagnosis for DCA must include hereditary hemorrhagic telangiectasia (HHT), Sturge Weber Syndrome (SWS) and Wyburn Mason Syndrome (WMS). None of these syndrome and their characteristic vascular skin lesions were found in our case.

The pathophysiology of the development of aneurysm associated with AVMs debated. It is reported that 10-20% of AVMs have associated aneurysms.<sup>16-21</sup> Of those, 50% are on the AVM's feeding arteries, 25% are on remote arteries and the remaining 25% are intranidal. It has been reported that feeding artery and intranidal aneurysms are at increased risk of bleeding.<sup>22-25</sup>

We found no other report of DCA and aneurysm. In our case, the basilar tip aneurysm was on an artery that was feeding the DCA, but all cerebral arteries on that side of the brain were also contributing, the flow from the posterior circulation was relatively low and the aneurysm was quite remote from the DCA. These factors blur the distinction between remote and feeding artery aneurysms in this condition.

It is possible that the association in this case is purely coincidental and there is no pathological link between these two conditions. Several terms have been used to describe the condition including DCA, cerebral proliferative angiopathy and diffuse AVM, and between them the number of reported cases is around 50. That means that the association that occurred in our case could be a coincidence because it makes the observed rates of subarachnoid hemorrhage and harboring aneurysms not very different from those risks in the background population. Perhaps more significant is that this DCA seems to be less associated with aneurysms and the more familiar ones.

## Conclusions

The present report shows a possible association between diffuse cerebral angiomas and cerebral aneurysms, but this association appears to be less strong than it is with other AVMs.

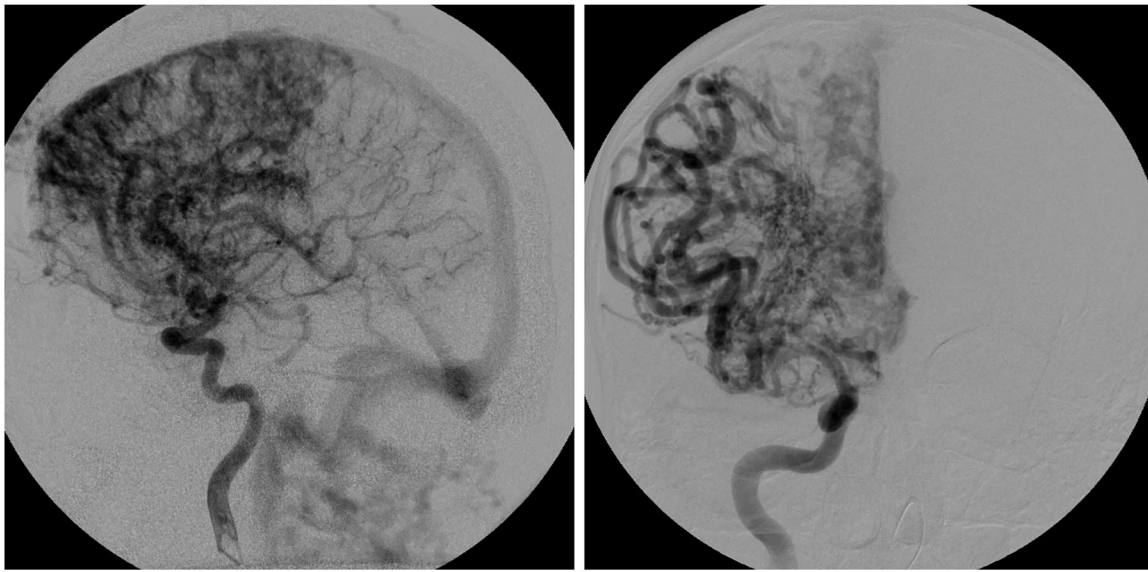


Figure 3. Cerebral angiography in lateral (left) and anterior (right) views showed the right hemispheric diffuse cerebral angiopathy.



Figure 4. Cerebral angiography in lateral view (up) demonstrated the presence of a basilar apex aneurysm. DSA volumetric 3D reconstruction of the basilar aneurysm (down).

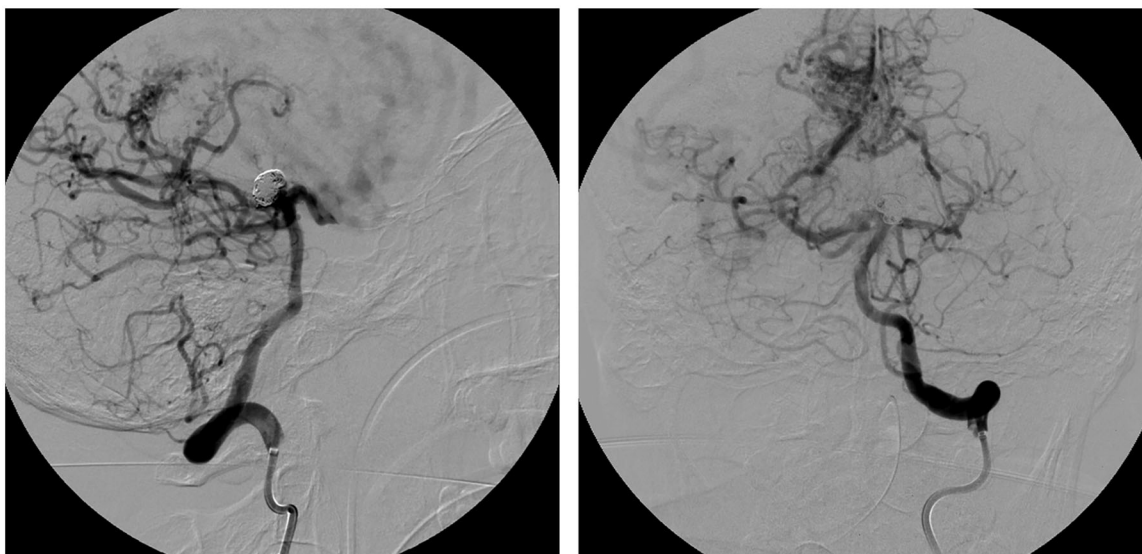


Figure 5. Cerebral angiography in lateral (left) and anterior (right) documented the embolization of the basilar apex aneurysm.

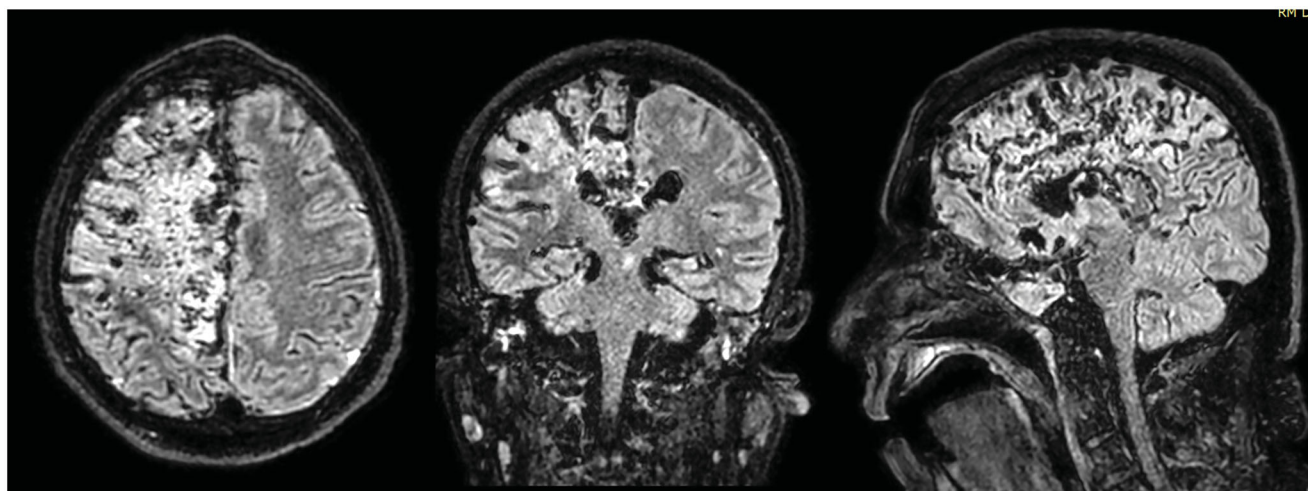


Figure 6. Axial (left), coronal (center) and sagittal (right) FLAIR MRI sequences showed large feeding arteries in the context of the lesion and surrounding edema.

### Ethical approval

There is no ethical issue in this paper. Note: all the images and patient's data in question have been anonymized, therefore not attributable to any person.

### Disclosure statement

No potential conflict of interest was reported by the author(s).

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### ORCID

Giuseppe Emmanuele Umana  <http://orcid.org/0000-0002-1573-431X>  
 Giovanni Federico Nicoletti  <http://orcid.org/0000-0003-1182-9028>  
 Gianluca Scalia  <http://orcid.org/0000-0002-9465-2506>

### References

1. Albright AL, Pollack IF, Adelson PD, *Principles and practice of pediatric neurosurgery*. 3rd ed. New York: Thieme Medical Publishers; 2014.
2. Montes-Ramirez J, Gomez-Brockmann P, Carrillo-Mezo R, Flores-Silva F, Cantu-Brito C. Diffuse cerebral angiomas: a case report with fatal outcome. *Br J Neurosurg* 2019; 1–4. doi: [10.1080/02688697.2019.1567682](https://doi.org/10.1080/02688697.2019.1567682). Epub ahead of print. PMID: 30829073
3. Schreiber SJ, Doepf F, Bender A, Schmierer K, Valdeuza JM. Diffuse cerebral angiomas. *Neurology* 2003;60:1216–8.
4. Wagle V, McCutcheon I, Melanson D, Ethier R, Roy LE. MR in diffuse angiomas. *AJNR Am J Neuroradiol* 1987;8:382–3.
5. Divry P, Bogaert V. L. Une maladie familiale caractérisée par une angiomasose diffuse corticoméningée non calcifiante et une démyélinisation progressive de la substance blanche. *J Neurol Neurosurg Psychiatry* 1946;9:41–54.
6. Solmaz I, Secer HI, Tehli O, Temiz C, Izci Y, Gonul E. Diffuse cerebral arteriovenous malformation. *Turk Neurosurg* 2010;20:536–9.
7. Lasjaunias PL, Landrieu P, Rodesch G, et al. Cerebral proliferative angiopathy: clinical and angiographic description of an entity different from cerebral AVMs. *Stroke* 2008;39:878–85.
8. Hong KS, Lee JJ, Hong SC. Neurological picture. Cerebral proliferative angiopathy associated with haemangioma of the face and tongue. *J Neurol Neurosurg Psychiatry* 2010;81:36–7.

9. Maekawa H, Tanaka M, Hadeishi H. Fatal hemorrhage in cerebral proliferative angiopathy. *Interv Neuroradiol* 2012;18:309–13.
10. Marks MP, Steinberg GK. Cerebral proliferative angiopathy. *J Neurointerv Surg* 2012;4:e25.
11. Liu P, Lv X, Lv M, Li Y. Cerebral proliferative angiopathy: clinical, angiographic features and literature review. *Interv Neuroradiol* 2016;22:101–7.
12. Ducreux D, Petit-Lacour MC, Marsot-Dupuch K, et al. MR perfusion imaging in a case of cerebral proliferative angiopathy. *Eur Radiol* 2002;12:2717–22.
13. Gold JJ, Crawford JR. Acute hemiparesis in a child as a presenting symptom of hemispheric cerebral proliferative angiopathy. *Case Rep Neurol Med* 2013;2013:1–3.
14. Ellis MJ, Armstrong D, Dirks PB. Large vascular malformation in a child presenting with vascular steal phenomenon managed with pial synangiosis. *PED* 2011;7:15–21.
15. Fierstra J, Spieth S, Tran L, et al. Severely impaired cerebrovascular reserve in patients with cerebral proliferative angiopathy. *PED* 2011;8:310–5.
16. Stapf C, Mohr JP, Pile-Spellman J, et al. Concurrent arterial aneurysms in brain arteriovenous malformations with haemorrhagic presentation. *J Neurol Neurosurg Psychiatry* 2002;73:294–8.
17. Turjman F, Massoud TF, Sayre JW, et al. Epilepsy associated with cerebral arteriovenous malformations: a multivariate analysis of angioarchitectural characteristics. *AJNR Am J Neuroradiol* 1995;16:345–50.
18. Turjman F, Massoud TF, Vinuela F, et al. Correlation of the angioarchitectural features of cerebral arteriovenous malformations with clinical presentation of hemorrhage. *Neurosurgery* 1995;37:856–60.
19. Gross BA, Lai PM, Du R. Hydrocephalus after arteriovenous malformation rupture. *FOC* 2013;34:E11.
20. Gross BA, Du R. Natural history of cerebral arteriovenous malformations: a meta-analysis. *J Neurosurg* 2013;118:437–43.
21. Marks MP, Lane B, Steinberg GK, et al. Hemorrhage in intracerebral arteriovenous malformations: angiographic determinants. *Radiology* 1990;176:807–13.
22. Chin LS, Raffel C, Gonzalez-Gomez I, et al. Diffuse arteriovenous malformations: a clinical, radiological, and pathological description. *Neurosurgery* 1992;31:863–9.
23. Yaşargil MG. *Microneurosurgery*. Vol. IIIA. Stuttgart: Georg Thieme Verlag; 1987.
24. Kimiwada T, Hayashi T, Shirane R, et al. 123I-IMP-SPECT in a patient with cerebral proliferative angiopathy: a case report. *J Stroke Cerebrovasc Dis* 2013;22:1432–5.
25. Kono K, Terada T. Encephaloduroarteriosynangiosis for cerebral proliferative angiopathy with cerebral ischemia. *J Neurosurg* 2014;121:1411–5.