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Diffuse cerebral angiomas: a case report with fatal outcome

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ABSTRACT

Diffuse cerebral angiomas is a rare disorder of the brain and only 5 cases have been reported to date. We report a 16-year-old female patient who presented to the emergency department with seizures. The patient had no hereditary syndromes, no epistaxis, no skin lesions and no telangiectasia. There was no livedo reticularis. The ophthalmologic examination was within normal limits. Magnetic resonance angiography (MRA) showed multiple diffuse tortuous vessels all around the brain tissue and ventricular system. Brain CT showed multiple calcifications. Digital subtraction angiography (DSA) demonstrated multiple arteriovenous shunts and diffuse arteriovenous malformations (AVM) without a typical nidus, draining into dilated cerebral veins. Multimodal ultrasound showed increased flow velocities in all cerebral arteries. We report the case of a patient with diffuse cerebral angiomas. This is the third study in which hemodynamic indices were evaluated through transcranial Doppler. Unlike all other case reports our patient had a bad outcome with important disability, dementia and drug resistant epilepsy that lead to the patient's death.

HIGHLIGHTS

- Diffuse cerebral angiomas is a rare disorder of the brain and only 5 cases have been reported to date.
- Unlike all other case reports our patient had a bad outcome with disability, dementia and drug resistant epilepsy that lead to the patient's death.

ARTICLE HISTORY

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KEYWORDS

Arteriovenous malformation; Magnetic resonance imaging; cerebral angiography; transcranial Doppler

Introduction

Diffuse cerebral angiomas is defined as diffuse infiltration of the brain by complex vascular structures. Cerebral arteriovenous malformations (AVM) are a common subgroup of intracranial vascular malformation composed of a nidus of vessels through which arteriovenous shunting occurs. AVM are thought to represent a congenital abnormality. Most cases are solitary (98%). They are comprised of feeding arteries, a nidus (shunting arterioles and interconnected venous loops) and draining veins. There are two types: A compact or glomerular nidus with abnormal vessels without any interposed brain tissue and a diffuse proliferative with no well-formed nidus, with functional neuronal tissue interspread amongst the anomalous vessels.¹

Multiple AVM are uncommon and usually a syndromic association is considered like Osler-Weber-Rendu, Divry-Van Bogaert or Sneddon's syndrome.^{2,3} When no association is found a diffuse cerebral arteriovenous malformation is considered. Diffuse cerebral arteriovenous malformation is a very rare disorder of the brain defined as diffuse infiltration of brain by complex vascular structures.⁴ Very few case reports have been reported and no fatality has been reported to date.^{4,5} We present a 16-year-old female patient with diffuse cerebral angiomas.

Case report

A 16-year-old Hispanic middle school female student arrived at the emergency department do to seizures. There was family

history for type 2 diabetes (grandmother). The patient had penicillin allergy.

The medical history was irrelevant until age 16 when she experienced her first focal seizures. She had transient episodes of tingling (crawling ants) on the left lower facial area. At age 18 the tingling was followed by myoclonic jerks of the left lower face and clonic seizures of the left arm with a duration of 5 minutes. She was sent to a neurology outpatient consult where the medical doctor prescribed Carbamazepine 600 mg/day in tid which achieved seizure control.

The patient's physical examination revealed no skin lesions and no telangiectasia. There was no livedo reticularis. The patient had a normal cognitive function. There was no cranial nerve involvement. The ophthalmologic examination was within normal limits. The patient's motor and sensory exploration was normal.

The laboratory test reports were normal. The Magnetic Resonance Imaging (MRI) of the brain showed multiple tortuous pathological vessel signals on the brain tissue. The Magnetic Resonance Angiography (MRA) showed diffuse vascular structures in the brain and tortuous vessels. The Brain CT revealed extensive calcifications (Figure 1).

At age 20 the patient experienced a decrease on strength on the left upper limb as well as breakthrough seizures. Levetiracetam 2000 mg/day in two divided doses was added to the treatment. A Digital subtraction angiography (DSA) demonstrated a right and left fronto-temporo-parieto-occipital vascular

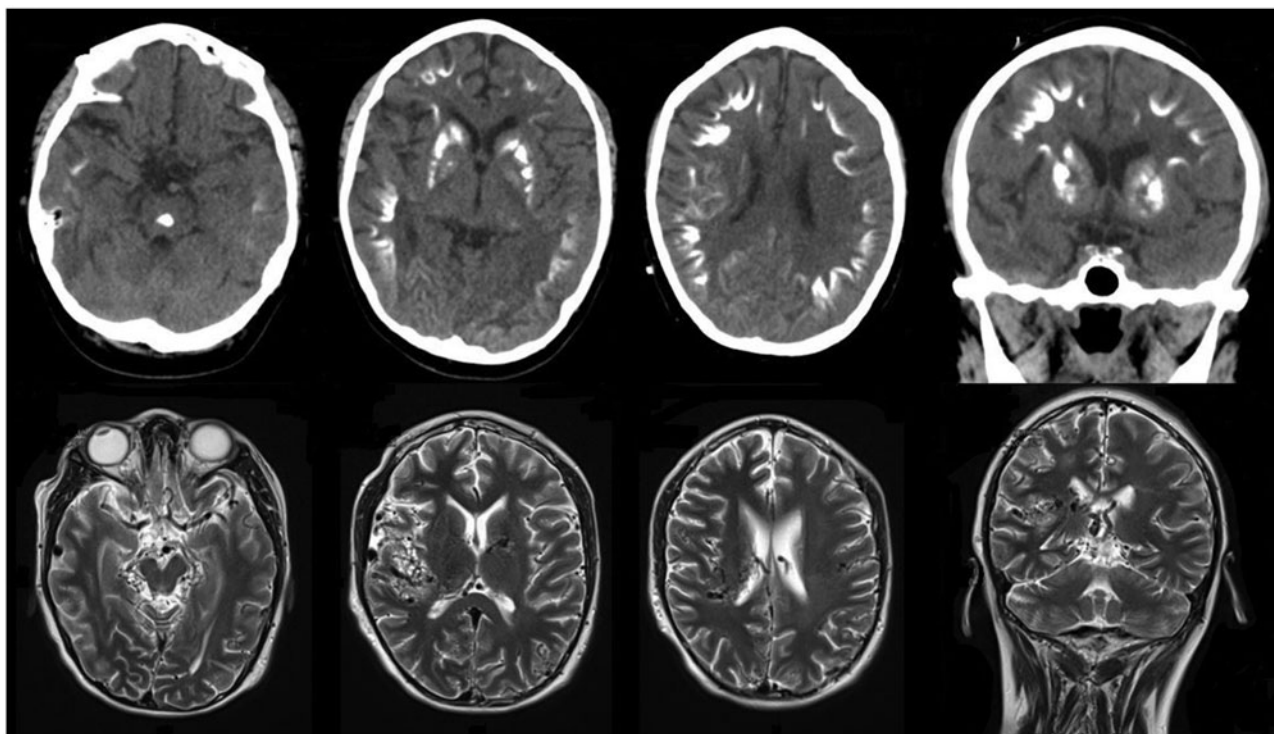


Figure 1. (a) Axial and coronal brain CT shows calcifications on pons, thalami, basal ganglia and subcortical white matter. (b) T2 weighed axial and coronal MRI shows multiple pathologic dilated vessels in the brain parenchyma with a right predominance.



Figure 2. DSA, selective right ICA demonstrates a 5.0 × 3.6 cm vascular conglomerate on the right fronto-temporo-parieto-occipital area. It was fed by multiple arteries (Medial Meningeal Artery, Occipital artery and Temporal artery) through multiple dural shunts. It's dominant feeding artery was the medial cerebral artery. The early venous drainage was through the superior longitudinal sinus.

conglomerate (Figures 2–4). Increased cerebral blood flow velocities (CBF) and low pulsatility were found in all basal cerebral arteries by transcranial Doppler (TCD) (Table 1).

Two months later the patient came to the emergency department with convulsive status epilepticus. She was transferred to the intensive care unit (ICU) due to super refractory status epilepticus. She remained on ICU for 21 days where she developed sepsis due to aspiration of oral content into the lungs. She was

transferred to the neurology department without sepsis. The patient had 3 seizures a day. Levetiracetam was increased to 3000 mg/day in two divided doses and CBZ was added at a dose of 600 mg/day divided in three doses. The patient had no seizures and she was discharged to home. However, the patient was discharged with disability, on a wheel chair and with loss of executive functions. The patient died 1 year later due to pneumonia.

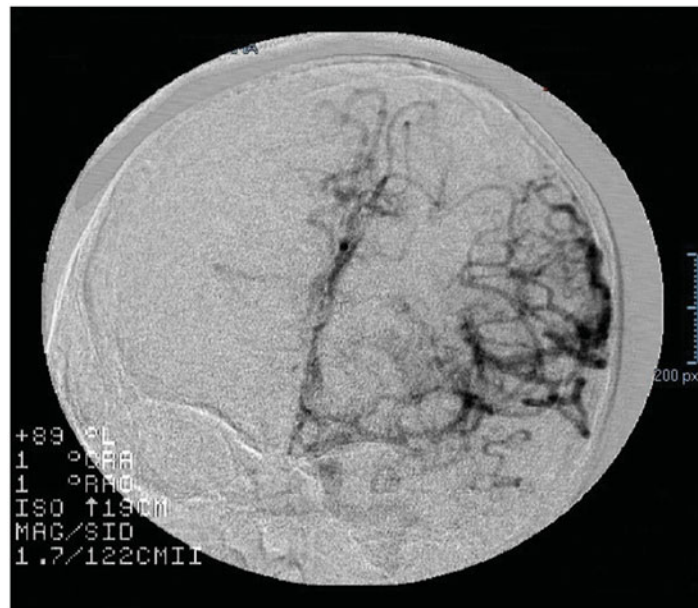


Figure 3. DSA, selective left ICA shows a fronto-temporo-parieto-occipital vascular conglomerate. It was fed by multiple arteries (Medial Meningeal Artery, Occipital artery and Temporal artery). It's dominant feeding artery is the medial cerebral artery (M3-M4) and the anterior cerebral artery (A3). The early venous drainage was through the superior longitudinal sinus and the transverse sinus.



Figure 4. DSA, selective vertebral artery shows multiple arteriovenous shunts.

Table 1. Mean blood flow velocities (MV) of the patient (cm/s).

	Right	Left
Middle Cerebral Artery		
MV	168.37	162.2
Pi*	0.58	0.45
Ri**	0.42	0.35
Posterior Cerebral Artery		
MV	130.27	
Pi*	0.67	
Ri**	0.46	
Extracranial Internal Carotid Artery		
MV	81.96	87.4
Pi*	0.58	0.33
Ri**	0.42	0.27

*Pulsatility Index.

**Resistance Index.

Discussion

We report the case of a patient with diffuse cerebral angiomas, a rare disorder with only 5 cases reported to date.^{4,5} This is the third study in which hemodynamic indices were evaluated through TCD.² Unlike all other case reports our patient had a fatal outcome. Her MRA showed multiple diffuse tortuous vessels all around the brain tissue and ventricular system. Brain CT showed multiple calcifications. The DSA demonstrated multiple arteriovenous shunts and diffuse AVM without a typical nidus, draining into dilated cerebral veins. Multimodal ultrasound showed increased flow velocities in all cerebral arteries.

This patient had no Rendu-Osler-Weber, Divry van Bogaert or Sneddon syndrome since she didn't have livedo reticularis, there was no telangiectasia, her ophthalmologic examination was

normal and there was no hereditary trait. So, this patient has unique characteristics of diffuse cerebral angiomas and another syndrome should not be considered.³

Unlike all other case reports our patient had a bad outcome with important disability, dementia and drug resistant epilepsy that lead to the patient's death. Although it is hard to determine if the dementia was secondary to sequela from status epilepticus or if it was due to ischemia and calcification of the brain parenchyma. We present the case of this rare vascular malformation and expect this will help our understanding of this disease and its hemodynamics.

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Disclosure statement

No potential conflict of interest was reported by the authors.

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Ethical statement

This study was approved by the appropriate ethics committee and have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments. The patient's tutors gave their written informed consent.

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