

Diffuse Cerebral Arteriovenous Malformation

Diffüz Serebral Arteriovenöz Malformasyon

ABSTRACT

Diffuse cerebral arteriovenous malformation (AVM) is a rare disorder of the brain and defined as diffuse infiltration of brain by complex vascular structures. It is usually associated with hereditary syndromes and presented with hemorrhage or seizure. We report a 20-year-old male patient who presented with drooping of the left eyelid. He had no skin lesion. The ophthalmological examination was within normal limits except periorbital bruit on auscultation. Radiological examination revealed a diffuse AVM comprising multiple arteriovenous shunts, draining bihemispherically through numerous dilated veins but without a typical arteriovenous malformation nidus. No hereditary disorder was detected. No treatment was performed and the patient is still under follow-up.

KEYWORDS: Diffuse arteriovenous malformation, Hereditary syndromes, Radiology

ÖZ

Diffüz serebral arteriovenöz malformasyon (AVM) beynin nadir görülen bir bozukluğudur ve beynin kompleks vasküler diffüz infiltrasyonu olarak tanımlanır. Genellikle kalıtsal sendromlarla birlikte ve kanama veya nöbet ile ortaya çıkar. Biz, 20 yaşında sol göz kapağı düşüklüğü ile gelen erkek hastayı yazdık. Hastanın cilt lezyonu yoktu. Oftalmolojik muaynesi periorbital oskültasyonda duyulan anormal ses dışında normal sınırlardaydı. Radyolojik tetkiklerde tipik arteriovenöz malformasyon nidusu olmaksızın bihemisferik birden fazla dilate venlerle drene olan multipl arteriovenöz shuntların olduğu diffüz AVM tesbit edildi. Kalıtsal bozukluk tesbit edilmedi. Cerrahi tedavi uygulanmadı ve hastanın takibi devam ediyor.

ANAHTAR SÖZCÜKLER: Diffüz arteriovenöz malformasyon, Kalıtsal sendromlar, Radyoloji

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INTRODUCTION

Angiomatosis is defined as diffuse infiltration of bone or soft tissue by hemangiomatous or lymphangiomatous lesions. Patients are mainly affected within the first 3 decades of life. Angiomatosis can involve predominantly osseous structures or soft tissue (3). There are considerable variations in the size and connections of the arteries and veins of the brain (4) and brain involvement of the angiomatosis, or arteriovenous malformation, is very rare. Only a few case reports in the literature could be found that addressed this lesion in a comprehensive manner (2,7,9,10).

We report a patient with diffuse cerebral arteriovenous malformation (AVM) of the brain, who presented with drooping of the left eyelid. The radiological characteristics of this unique entity are presented and discussed with the literature.

CASE REPORT

A 20-year-old male patient presented with drooping of his left eyelid which had existed since his childhood. He was healthy otherwise. There was no family history. There was no history of seizure or loss of vision. The ophthalmological examination was within normal limits except for periorbital bruit on auscultation. No skin lesion was detected on the body.

Computed tomography of the head after contrast administration revealed diffuse vascular structures throughout the brain and especially on the left orbit.

Magnetic resonance imaging (MRI) (Magnetom Vision, 1.5 Tesla, Siemens, Germany) of the brain showed multiple tortuous pathologic vessel signals all around the brain tissue and in the ventricular system (Figure 1). MRI of the orbits showed large vessels compressing the left orbit, and causing the drooping of eye. These vessels were in continuity with the intracranial vessels (Figure 2). MRA revealed diffuse vascular structures in the brain and tortuous vessels lying in the left orbit and scalp (Figure 3A,B).

Digital subtraction angiography (DSA) demonstrated multiple arteriovenous shunts and diffuse cortical angiomatosis, without a classical AVM nidus, draining into dilated cerebral veins (Figure 4). There was a smoke-like appearance of the cerebral vessels in the arterial phase of DSA.

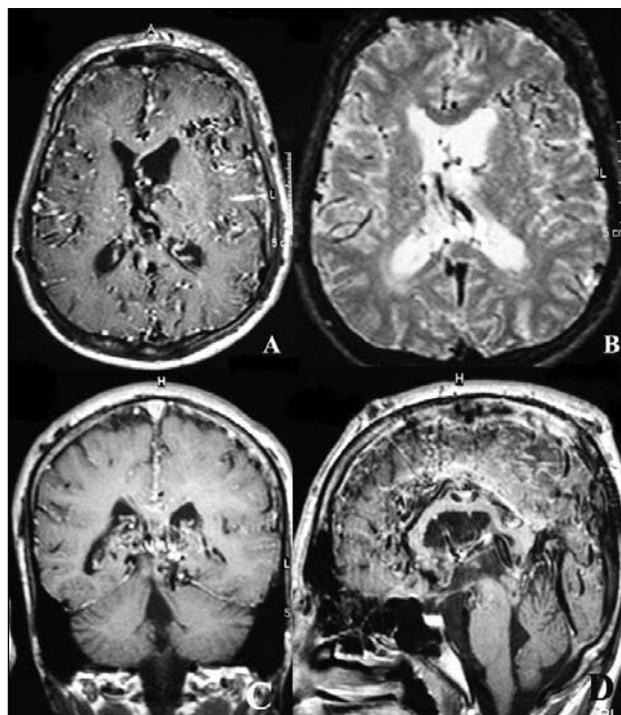


Figure 1: T1-weighted axial (A), coronal (C), and sagittal (D) MRI scans of the brain show multiple dilated vessels in brain parenchyma. T2-weighted axial MRI scan (B) shows large vessels in the lateral ventricles.

No surgical intervention was planned for the patient because of the risk of death or severe neurological deficits. Embolization or radiotherapy was also not scheduled. He is under follow-up for the AVM.

DISCUSSION

We report a case of diffuse cerebral AVM that developed in the brain of a young man. This location of the lesion was unusual because this entity mostly involves the other parts of the body and is associated with some hereditary disorders. However, the imaging features of our patient closely resembled the case reported by Schreiber et al. (9). This patient suffered from a bilateral retinal angiomatosis in combination with a diffuse cerebral AVM comprising multiple small cortico-leptomeningeal arteriovenous shunts, draining bihemispherically through numerous dilated veins but without a typical AVM nidus. The retina was not involved in our patient.

AVM is a localized or generalized congenital vascular abnormality composed of microscopic direct connections between arteries and veins, without the normal intervening capillary bed. Furthermore, the parenchyma found among the abnormal vasculature

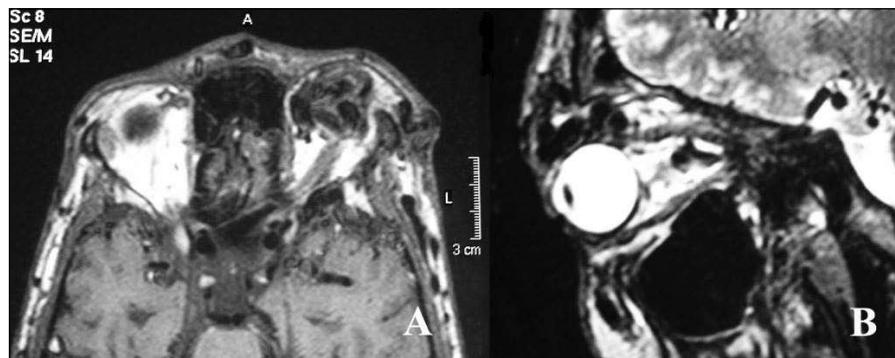


Figure 2: T1-weighted axial (A), and T2-weighted sagittal (B) MRI scans of the orbits show dilated tortuous vessels lying into the left orbit and compressing the left eye.

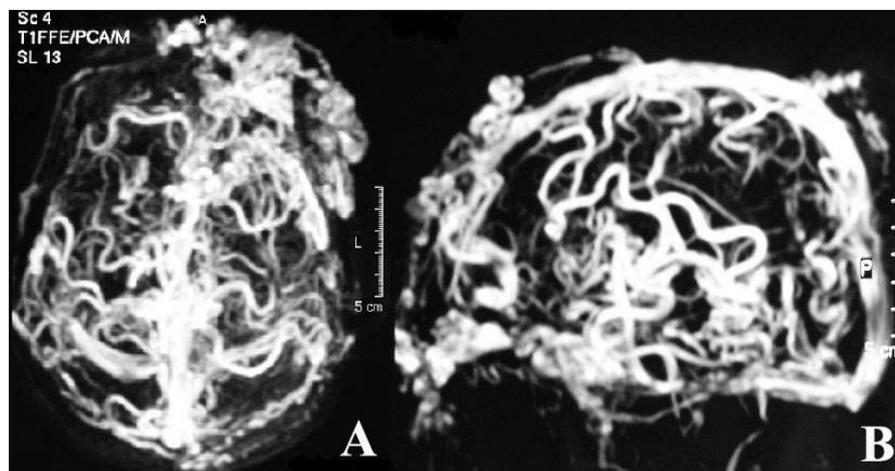


Figure 3: MRA of the patient in (A) axial plane, and in (B) sagittal plane show diffuse and multiple vascular structures in all around the brain.

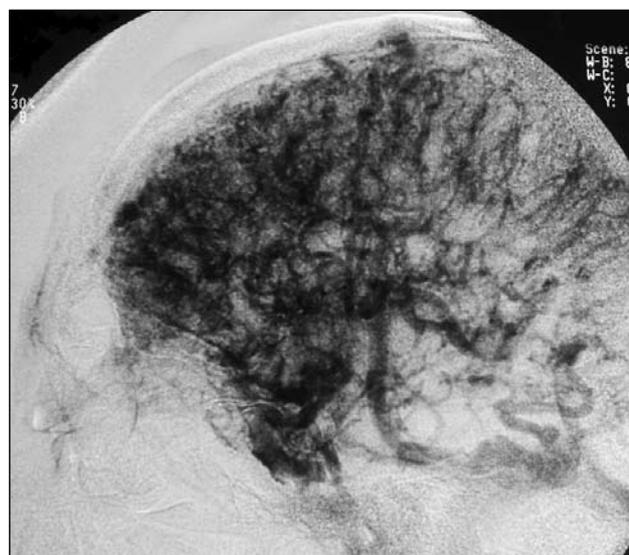


Figure 4: DSA of the patient from the right carotid artery shows diffuse cerebral vessels in smoke-like fashion without a nidus formation.

regions (4,5). The precise cause(s) are unknown. The AVM that appears diffuse on cerebral angiography has been studied far less. In fact, only a few case reports and one series of diffuse AVMs have been reported in the literature. This subtype of AVM has a distinct angiographic appearance and implies the existence of neural tissue within the malformation (11,12,13). Internal carotid arteries are the main vascular source of the brain (6). Diffuse AVMs often have an arterial supply from multiple vascular distributions, including the anterior as well as the posterior circulation, and in many cases, the arterial supply is bilateral. In our case, the arterial supply of the AVM was bilateral from the internal carotid arteries.

MRI and MRA are generally used in the diagnosis of diffuse AVMs. DSA, the “gold standard” for AVM diagnosis, allows direct vessel visualization, and shunt estimation via the assessment of regional or global arteriovenous circulation times (11,12,13).

The AVMs were defined as MRI features: a collection of enlarged vessels easily seen on both T2- and T1-weighted (without and with contrast

is usually reported as absent or highly gliotic and nonfunctional (2,13). Because of the significant variability in the vascular pattern of the human brain, an AVM is more common in the brain than in other

enhancement) sequences; associated enlargement or ectasia of adjacent pial arteries and draining veins; a well-defined, relatively large nidus (0.15 mm); and such secondary features as dilated proximal intracranial or extracranial arteries (angiomatic change). The brain tissue adjacent to AVM might show old hemorrhage, gliosis, or both (2). In our case, MRI of the brain shows enlarged vessels in the parenchyma and also in the ventricles. No nidus formation was observed.

Four-vessel cerebral DSA findings were diagnostic of patients with diffuse AVM. Diagnostic findings consisted of a large and multiple feeding arteries, no classical AVM nidus, and large, tortuous draining veins to superficial or deep venous structures. The diffuse AVM opacified late in the arterial phase before filling in the capillary phase (2,11,12,13). Smoke-like diffuse vascular staining was observed in the arterial phase of our patient's DSA. Dilated multiple draining veins were demonstrated in the venous phase. These findings are typical for a diffuse AVM of the brain.

Although conventional radiographic DSA provides higher spatial and temporal resolution, the noninvasive nature of MRA may justify its use over conventional radiography in certain cases, particularly in screening and follow-up situations. In addition, time-resolved ^{MRA} could become part of a comprehensive neurological MR examination including both anatomic and functional (diffusion, perfusion, and fMRI) imaging (1). We used MRA in the diagnosis and follow-up of our patient.

The pathogenesis of angiomatosis remains unclear, but it is currently believed to result from vascular malformations of congenital origin. Although many theories have been proposed as to how these malformations actually occur in the fetus, there still remains considerable debate. However many cases of angiomatosis appear in familial genetic disorders such as hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber syndrome), Sturge-Weber syndrome, Wyburn-Mason-Syndrome and Divry-van Bogaert syndrome (2,7,8,9). None of these syndromes were detected and no cutaneous sign was observed in our patient.

Blood vessels within the AVM carry a higher risk of rupture and up to one half of affected patients primarily present with an intracranial hemorrhage. Other common symptoms are epileptic seizures,

focal-neurological deficits, and pulsatile tinnitus or headaches, due to the effects of the altered arterial and venous hemodynamic status. However, a considerable number of patients remain asymptomatic (2,11,13). Our patient is neurologically intact and presented with the drooping of the left eyelid. He had no history of hemorrhage or seizure.

In conclusion; diffuse cerebral AVM is a rare vascular disorder of the brain and usually manifests together with a hereditary syndrome. DSA is still the imaging mode of choice. A detailed study of the arterial feeders, the nidus and venous drainage is mandatory.

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