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Case Report

Massive Arteriovenous Malformation with Stroke-Like Presentation

Mathias Bürki[a] Liliane Kappeler[b] Robert Bühler[b] Dirk Springe[a] Lukas Ebnöther[a] Marcos Delgado[a]

[a]Intensive Care Unit, Department of Anaesthesiology, Bürgerspital Solothurn, Solothurn, Switzerland; [b]Department of Neurology, Bürgerspital Solothurn, Solothurn, Switzerland

Keywords
Arteriovenous malformation · Stroke mimics · Age

Abstract
We report of a 75-year-old patient with stroke-like presentation, where cerebral imaging led to the diagnosis of a massive arteriovenous malformation (AVM) of the whole left hemisphere. We suggest considering AVM as a differential diagnosis in patients with symptoms of acute stroke despite age and, in the absence of contraindications, in this setting to obtain MRI or CT angiography of the brain.

Introduction

In patients with symptoms of acute stroke, diagnostic measures do not have to be taken only to exclude hemorrhagic stroke. Arteriovenous malformation (AVM) is an etiology that can mimic acute ischemic or hemorrhagic stroke. In view of that, computed tomography (CT) angiography of the brain has to be obtained, as cranial CT without angiography could lead to intravenous fibrinolysis. In patients with AVM, who have an elevated risk of intracerebral bleeding, this could lead to fatal outcome.

KARGER

Marcos Delgado
Department of Intensive Care Medicine
Bürgerspital Solothurn
4500 Solothurn (Switzerland)
E-Mail marcos.delgado@spital.so.ch

Case Presentation

A 75-year-old patient was assigned to our emergency department with acute onset aphasia and reduced strength in the right arm and leg. Clinical examination showed a right sided hemiparesis and hemihypesthesia, a neglect to the right and global aphasia. The tendon reflexes were brisk. Furthermore, orientation was impaired. Suspected diagnosis was ischemic stroke with possible epileptic component due to myoclonia in the upper extremities of the patient. Intravenous diazepam was administered.

His medical history revealed resection of urothelial carcinoma without relapse, chronic renal insufficiency, diabetes mellitus type 2, and peripheral and coronary artery disease with quadruple aorto-coronary bypass several years previously.

CT angiography of the brain and neck was performed, showing unexpected, in consideration of the patient's age, massive AVM of the left hemisphere with drainage to the superior sagittal sinus (Fig. 1). There were no signs of cerebral ischemia or bleeding. According to these findings, the planned fibrinolysis was cancelled due to the high risk of intracerebral bleeding.

The patient was transferred to the ICU. An electroencephalogram (Fig. 2) showed alpha activity on the right, epileptic focus with rhythmic delta activity, and several suspected epileptic discharges frontotemporal and frontocentral on the left. No status epilepticus could be detected. An anti-epileptic therapy with levetiracetam was started and subsequently sensory as well as motor deficits and aphasia disappeared.

The next day, MRI was carried out, confirming a huge AVM of the whole left hemisphere. The nidus was suspected in the left visual cortex with perifocal gliosis as an expression of a chronic steal phenomenon and eventual pseudoaneurysm in the left-sided basal ganglia (Fig. 3, 4). Blood supply was provided by the posterior cerebral artery and M1 branch of the middle cerebral artery. No tumors were detected. Progressive aneurismatic dilatation within the AVM was postulated as the cause of the acute onset of symptoms.

The remainder of his stay in the ICU was unremarkable, and the patient was discharged to the neurological ward with permanent anti-epileptic therapy. Later, the patient was assigned to the hospital reference center for evaluation of further treatment.

Discussion

AVM are congenital vascular lesions, characterized by an atypical aggregation of dilated arteries and veins. These malformations arise from a deficient structure of blood vessels and a lack of capillaries. The exact pathological mechanisms are not yet fully understood. Severe clinical manifestations only occur in the brain [1].

The cause of AVM is most likely a high variability of hemodynamic forces. Furthermore, it is assumed that within this variability epigenetic factors in endothelial cells are being triggered. This causes a change in development and finally an aberration of the phenotype of blood vessels [1]. The average age for the diagnosis is 31.2 years, with a similar spread to both sexes. The symptoms that lead to further testing for AVM are most commonly epileptic seizures, focal-neurologic deficits, or headaches [2].

AVM are often graded after the Spetzler-Martin classification that separates six degrees of severity, with grade I being small, superficial, and located in a noneloquent part of the cortex and grade VI being inoperable [3]. This grading system evaluates the risk of surgical resection. In our case, Spetzler-Martin grade II in an eloquent area of the brain was diagnosed.

With microsurgery, a complete obliteration can be achieved in 96% of all cases. If surgical resection is not feasible or contraindicated, radiotherapy is a safe and effective alternative [4]. Radiotherapy can achieve complete obliteration in 40% of all cases [5]. A third option is endovascular embolization. On its own it often does not provide complete obliteration. Therefore, endovascular obliteration is mainly used to support and facilitate microsurgical resection [5]. In conclusion, microsurgical resection remains the gold standard in the treatment of AVM with increasing evidence that radiotherapy is an efficient alternative [6].

Concerning prognosis, a meta-analysis reported a case fatality of 1.1 per 100 patient-years with similar mortality rates after microsurgical resections and endovascular embolization. Mortality among patients that were treated with radiotherapy was only half as high. Male sex, drainage solely through the deep veins, and smaller AVM seemed to be protective factors [5].

Though most of the patients with AVM are young when diagnosis is made there are also patients becoming symptomatic only in older age, as in our case. A multicenter study [7] showed that 65% of all AVM of patients over 60 years were treated with microsurgery, supported by preoperative embolization in 1 out of 3 cases. Radiosurgery, embolization only, or a conservative approach was each chosen as therapy regimen in 10% of all cases. Patients who received microsurgical resection showed (distinct) best outcome. In 2 out of 3 cases, regardless of treatment regimen, a satisfactory outcome was achieved. After interventional treatment of AVM, age is not a risk factor for subsequent hemorrhage [8]. In untreated AVM age has been proposed as a risk factor for hemorrhage, with some studies showing older age [9] and others showing younger age [10] to be associated with higher risk of hemorrhage.

In the literature, there are several descriptions of first diagnosis of AVM in younger patients. Surprisingly, in our case the patient did not show any symptoms until an advanced age despite the huge size of AVM. To our knowledge, the age of our patient combined with the expanse of the AVM are unique.

Given the symptoms the patient presented, the main suspected diagnoses at this age were ischemic or hemorrhagic stroke, epileptic seizures, hypoglycemia, abscess, or tumor of the central nervous system [11]. Neuroimaging plays an important role in the differential diagnosis of neurological events with regard to treatment and improvement of outcome. Plain cranial CT scan (without contrast) is the most commonly used form of neuroimaging carried out in the emergency unit in the case of acute neurological symptoms resembling stroke. Although this technique is sufficient to recognize contraindications for fibrinolysis [11], it is essential to obtain CT angiography to detect other etiologies, such as AVM, and to avoid fibrinolysis which would increase the risk for intracerebral bleeding and fatal outcome.

The case described above shows that AVM should be considered as a differential diagnosis in patients with symptoms of acute stroke despite age. The performance of CT angiography is an essential first diagnostic tool to improve diagnostic accuracy and outcome.

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Statement of Ethics

The trial was conducted in accordance with the Declaration of Helsinki and its amendments. For the actual case it was not necessary to involve the ethics committees, and the patient provided written informed consent before inclusion in the study.

Disclosure Statement

The authors declare that they have no financial or nonfinancial competing interests regarding this paper. No funding was received for this study.

Author Contributions

Conception and design of study: Mathias Bürki and Dr. Marcos Delgado. Acquisition of data: Mathias Bürki, Dr. Marcos Delgado, and Dr. Robert Bühler. Analysis and/or interpretation of data: Mathias Bürki, Dr. Marcos Delgado, and Dr. Liliane Kappeler. Drafting the manuscript: Mathias Bürki. Revising the manuscript critically for important intellectual content: Dr. Marcos Delgado and Dr. Liliane Kappeler. All authors gave their approval of the version of the manuscript to be published.

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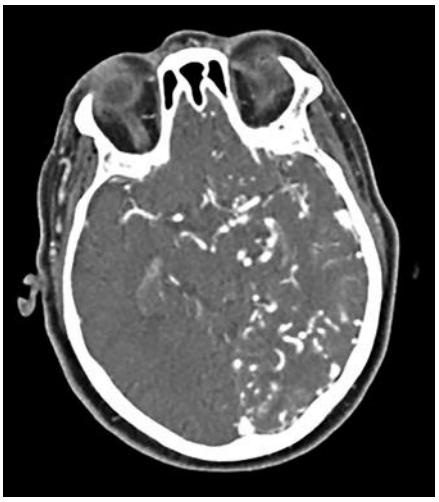

Fig. 1. Extensive arteriovenous malformation of left hemisphere with multiple aneurismatic extensions and expansion from the brainstem to the whole left hemisphere; accentuation in the left visual cortex.

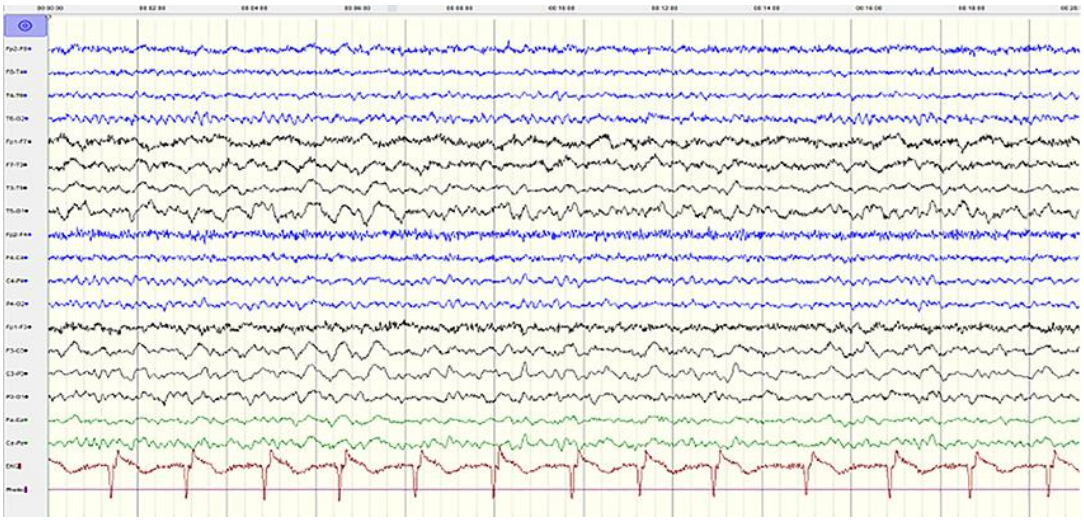

Fig. 2. EEG: normal alpha rhythm on the right; focal slowing on the left.

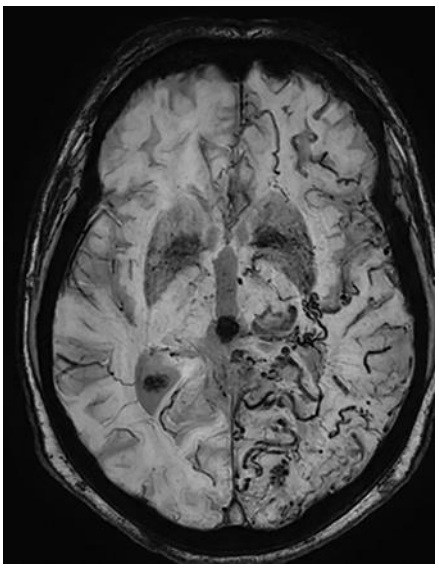

Fig. 3. MRI with depiction of extensive arteriovenous malformation of the left hemisphere.

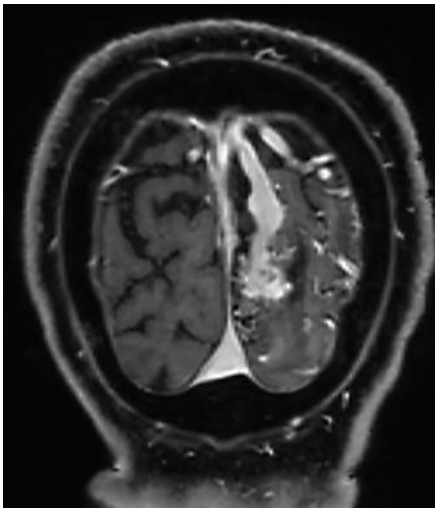

Fig. 4. MRI with depiction of extensive arteriovenous malformation of the nidus in the left visual cortex.