

CASE REPORTS

Mixed Venous and Cavernous Malformations of the Brain

Beyinde Birleşik Venöz ve Kavernöz Malformasyon

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Abstract: Vascular malformations with histopathological features of more than a single type of malformation present within the same lesion have been previously described. These lesions are very rare and named as mixed vascular malformations. In this report, we presented a case of mixed vascular malformation with radiological features of cavernous and venous malformations.

Key Words: Arteriovenous malformation, capillary malformation, cavernous malformation, mixed malformation, vascular malformation, venous malformation

Özet: Aynı malformasyonda birden fazla histolojik tip içeren damarsal malformasyonlar daha önce tanımlanmıştır. Nadir görülen bu lezyonlar birleşik damarsal malformasyonlar olarak adlandırılır. Bu makalede kavernöz ve venöz malformasyonların radyolojik özelliklerini gösteren bir birleşik damarsal malformasyon sunuldu.

Anahtar Sözcükler: Arteriyo venöz malformasyon, damarsal malformasyon, kapiller malformasyon, kavernöz malformasyon, venöz malformasyon

INTRODUCTION

Vascular malformations of the brain are classically divided into four groups, each with different pathological and radiological properties. This widely accepted classification includes venous, cavernous, arteriovenous and capillary malformations (4,5,6). However, some authors recently reported cases of vascular malformations exhibiting pathological features of more than one discrete malformation type within the same lesion (1,3,9,10). These lesions are rare and are named as "mixed vascular malformations". In this report, we present a case of vascular malformation which has characteristic magnetic resonance (MR) and angiographic features of cavernous and venous malformations.

CASE REPORT

A 34-year-old woman was admitted to the hospital because of recurrent headaches. Her medical

history was insignificant. Neurological and physical examinations were entirely normal. A computed tomography (CT) scan showed a hyperdense lesion on the roof of the fourth ventricle (Figure 1). The lesion was enhancing and typical appearance of caput medusa adjacent to the lesion was also seen (Figure 2a). An MR scan revealed both the venous structure and the lesion characterized by a core of mixed increased and decreased signal intensities, typical of cavernous malformation (Figure 2b). Angiography demonstrated abnormal venous drainage of the pontomedullary venous circulation (Figure 3). The patient did not receive any surgical treatment.

At follow-up examination, three months later, the patient was neurologically intact.

DISCUSSION

Histopathological criteria for the classification of vascular malformations of the brain have been well described (4,5). An arteriovenous malformation

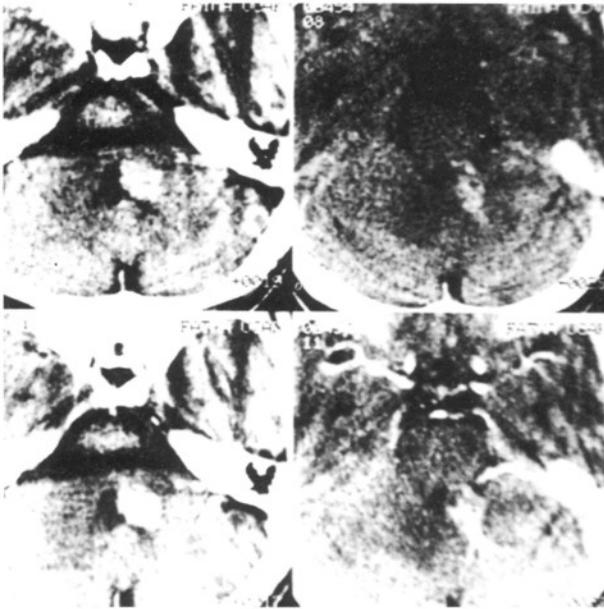


Figure 1: Postcontrast CT scan showing a hyperdense contrast enhancing mass on the left wall of the fourth ventricle and a vascular structure having caput-like appearance.

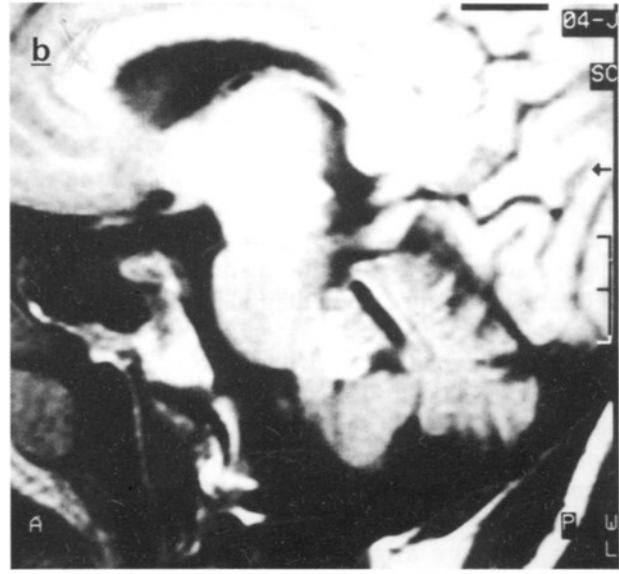
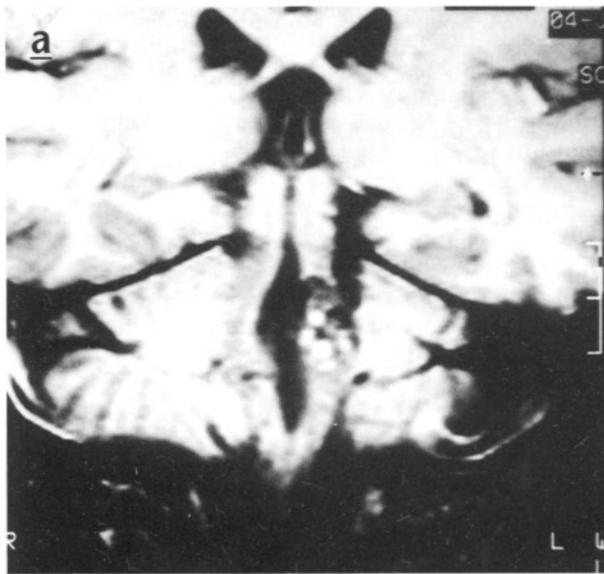


Figure 2 a, and b: T1-weighted MR images revealed the lesion characterized by a core of mixed signal intensity and a rim of decreased signal intensity. This appearance is highly characteristic for cavernous malformations. The linear signal void adjacent to the lesion outlines the venous structure.

includes clusters of abnormal arteries and veins with typically thickened vessel walls and minimal intervening gliotic brain tissue. A cavernous malformation is composed of large sinusoidal vascular spaces without intervening brain parenchyma, while a venous malformation entirely consists of veins interspersed in neural

parenchyma. These veins converge into a large centrally located vein making the feature of caput medusa. A capillary malformation consists of a compilation of dilated capillaries with intervening normal brain tissue. The category of mixed vascular malformation has recently been added to this classification (1,9). However, the real prevalence of mixed vascular malformations is not known. Although almost all cases have been reported in the MR era, most of the reports have emphasized the rarity of such lesions. According to Awad et al. 5 % of the all vascular malformations are mixed lesions (1). However, increasing availability of modern diagnostic techniques will allow better recognition of such lesions.

Mixed vascular malformations consist of three major subgroups including mixed cavernous and venous malformations, mixed arteriovenous and venous malformations and mixed cavernous and arteriovenous or capillary malformations. Most of the mixed vascular malformations are mixed cavernous and venous malformations, and can be easily

diagnosed on imaging studies because MR features of cavernous malformations are well known although they are usually occult on angiography (8).

A mixed arteriovenous and venous malformation is more difficult to recognize. MR and CT studies are usually insufficient for diagnosis. Only

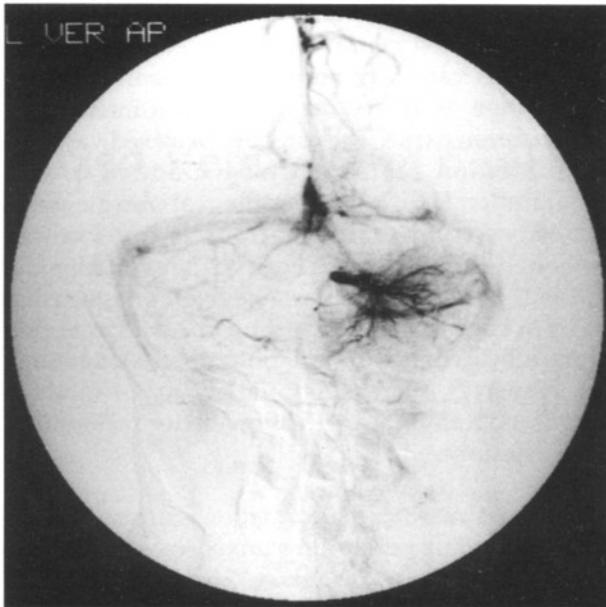


Figure 3: Angiography showing venous angioma in the posterior fossa.

comprehensive angiographic studies can visualize the arteriovenous and venous components of the lesion.

Cavernous malformations with mixed arteriovenous or capillary malformation are most difficult to recognize on diagnostic radiological studies. Only careful histopathological examination of the lesion and the surrounding brain can be helpful in establishing the correct diagnosis.

Most vascular malformations have been thought of as developmental anomalies or hamartomas, while enlargement of a cavernous malformation following repetitive hemorrhages and thrombosis has been shown (7). Therefore, microhemorrhages within an occult malformation cause reactive angiogenesis with new vessel formation. This process is named as hemorrhagic angiogenic proliferation and explains the occurrence of mixed vascular malformations (1).

Patients with mixed vascular malformations may present with seizures, recurrent hemorrhages or progressive neurological deficits (4). These clinical manifestations are not different from those encountered in patients with a single-type vascular malformation. However, venous angioma is the most common form of intracranial vascular malformation and has a risk of hemorrhage of only 0.22 % per year (2). Thus, venous angiomas are considered to be

relatively benign lesions and clinical findings are usually related to the other component of the mixed vascular malformation.

Management of mixed vascular malformations is difficult and complicated, especially if the lesion has a venous component. Since the venous malformation is frequently the only venous drainage pathway for the surrounding brain tissue, preserving the venous malformation during surgery is essential (10). However, if venous hypertension causes recurrent hemorrhages which subsequently stimulate the growth of new vascular malformations, only total removal of the mixed vascular malformation will be curative.

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