ANGIOGRAPHICALLY OCCULT ARTERIOVENOUS MALFORMATION OF THE BRAIN

M.N. Pamir, M.D.* / A.F. Özer, M.D.* / G.E. Keleş, M.D.**

* *Associate Professor, Department of Neurosurgery, Faculty of Medicine, Marmara University, İstanbul, Turkey.*

** *Research Assistant, Department of Neurosurgery, Faculty of Medicine, Marmara University, İstanbul, Turkey.*

SUMMARY

Angiographically occult arteriovenous malformations of the brain have begun to be described more often with the increasing clinical use of new neurodiagnostic imaging techniques. Despite this fact, they are still considered rare. In this paper, a case of histopathologically verified occult arteriovenous malformation is presented and the current literature is discussed.

Key words : Cerebral Vascular Malformation, Cerebral Angiography.

INTRODUCTION

The fact that small diameter vascular malformations may cause fatal brain hemorrhage was first pointed out in 1951 by Morgalis (1). Later, Crawford and Russell (2) named these anomalies "cryptic vascular malformations" because of their radiologically and pathologically latent character. Because they are small and compressed by the surrounding hematoma, these vascular anomalies usually cannot be demonstrated angiographically. For this reason they are called occult arteriovenous malformations (AVM).

Following the introduction of Computerized Tomography (CT) and Magnetic Resonance Imaging (MRI) for clinical use, such angiographically occult anomalies have begun to be described more often (3,4,5,6). It has also been shown that, not only small diameter anomalies but also those of two to three centimeters in diameter or even larger may be occult (4,5). Despite this, occult AVMs are a relatively rare group amongst all cerebral AVMs.

In this report we present a histopathologically verified occult AVM and discuss the current literature.

CASE REPORT

A 43 year old female patient presented to our neurosurgical out-patient clinic complaining of epileptic fits. The first episode had occurred one month previously when she had had a seizure which began with chewing and then spread to her left side and had become generalized. She also had had a similar seizure two weeks previously.

The patients's physical and neurological examinations were normal. The CT scan revealed a hyperdense round shaped lesion within the right temporal pole. The lesion was 2 to 3 centimeters in diameter, with no edema or mass effect around it, and with no contrast (meglumine iothalamate 60 %) enhancement. (Fig. 1a, 1b). Percutaneos right carotid angiography was performed but no pathology was revealed. (Fig. 2).

The patient was taken to theatre with a diagnosis of right temporal space occupying lesion. Under general anesthesia, a right temporal craniotomy was performed. Upon opening the dura, it was observed that the colour of the gyri of the temporal region had changed and had become yellowish. Following a small cortical incision 2 centimeter diameter dark brown mass was encountered at a depth of 3 millimeters and was completely removed. Patchy, dark brown thrombus-like material was aspirated. The surrounding brain tissue was a light yellow-brown colour. There was no bleeding during the excision of the abnormal tissue. The postoperative period was uneventful and she was discharged on the tenth postoperative day.

Histopathological examination of the mass revealed multiple, partially fibrotic, adhered vascular formations embedded in the brain tissue. Some of these had organised thrombi in their lumen. Macrophages con-

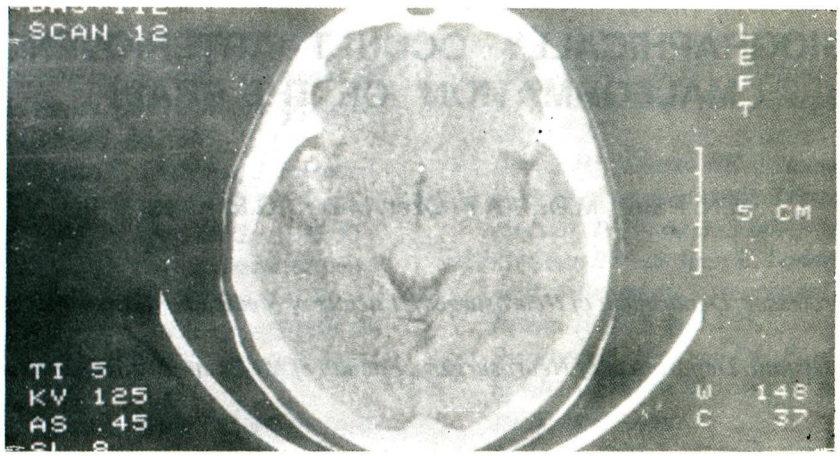


Fig. 1a : Computerized Tomography revealed a hyperdense at the pole of the right temporal lobe.

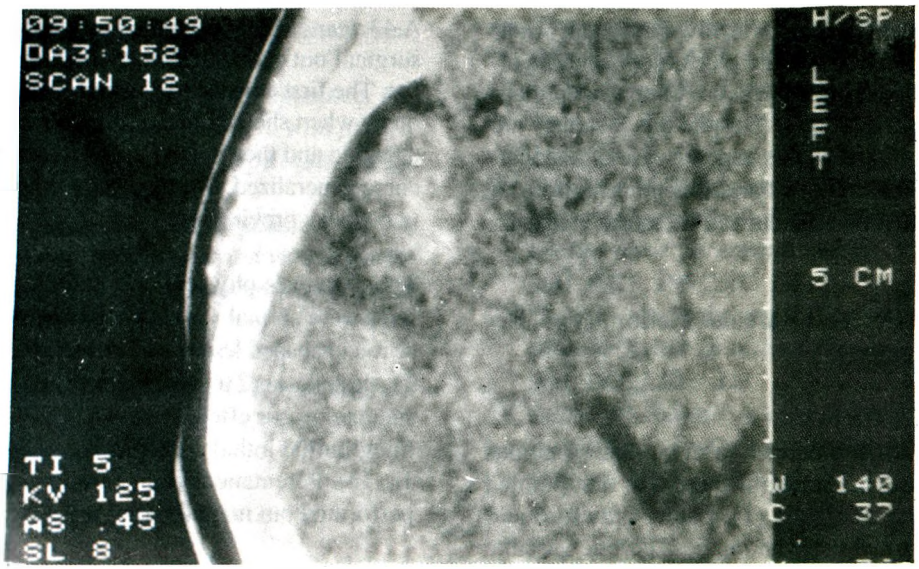


Fig. 1b : Magnification showed that the mass was non-homogenous, irregular, with slight surrounding edema No enhancement was detected after injection of intravenous contrast material.

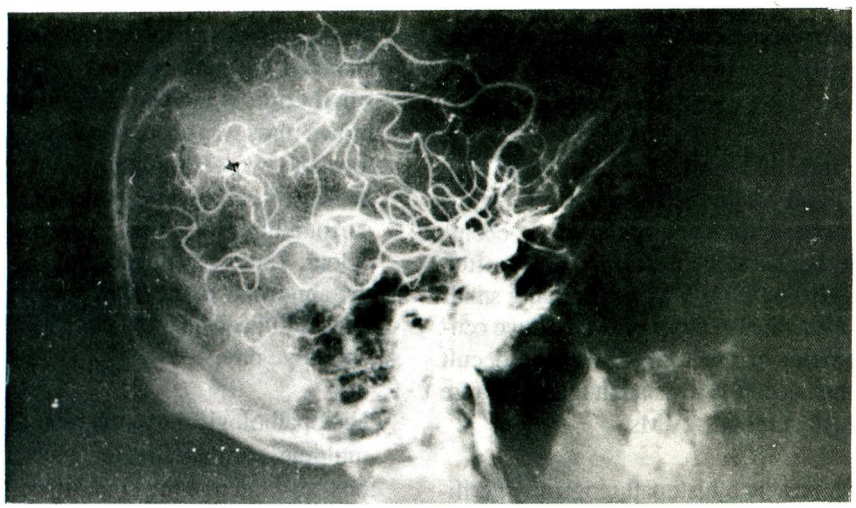


Fig. 2: Right carotid angiography was normal, no vascular abnormality was detected.

taining very dense hemosiderin deposits, areas of hemorrhage, areas containing inflammatory cell infiltrates, and areas of organisation were also observed. There were gliosis and small calcified fields in the surrounding brain tissue (Fig.3).

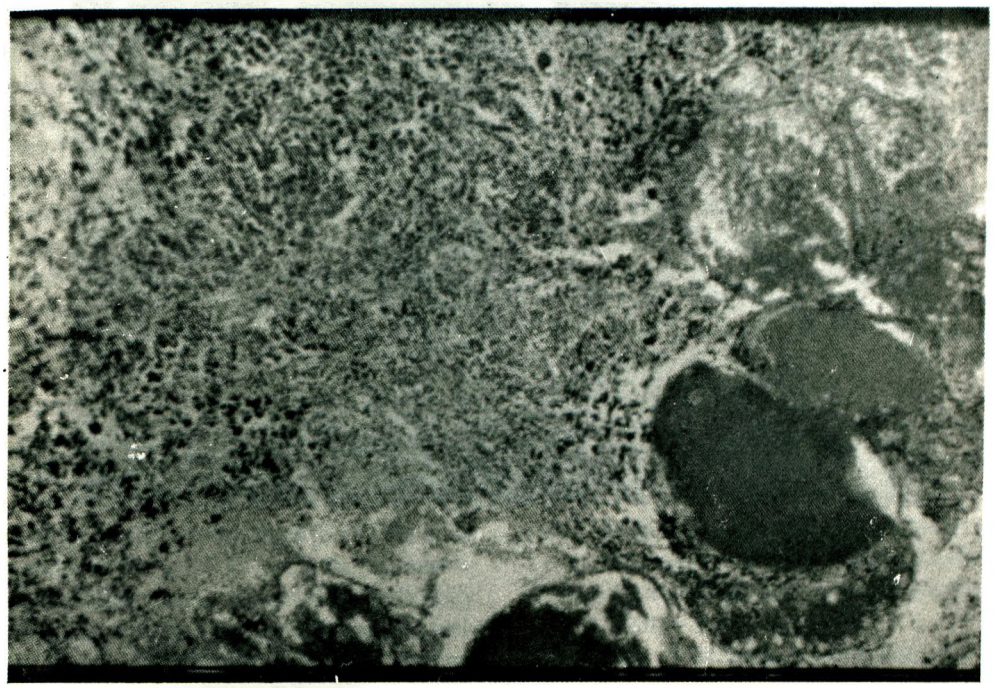


Fig. 3 : Histopathological examination of the mass revealed sections of several abnormal vessels, macrophages containing very dense hemosiderin deposits, organisation areas and inflammatory cell infiltrates.

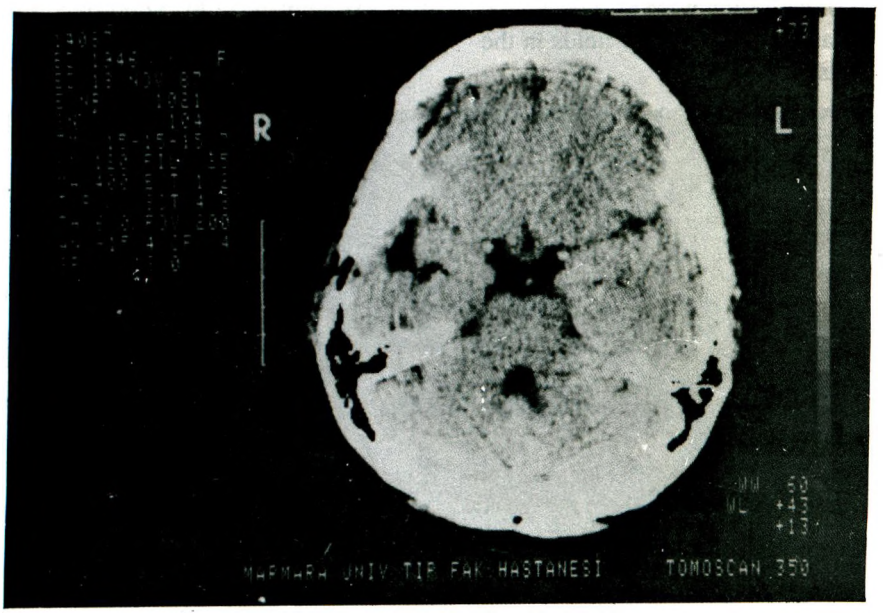
Six months later, the patient had no complaints of seizure, and her physical and neurological examinations were normal. The control CT scan showed right temporal atrophy at the localization of the excised pathological tissue (Fig. 4a, 4b).

DISCUSSION

Cerebral vascular malformations (CVM) are the result of persisting primitive vascular formations of the embryological period which should normally disappear (7). CVMs, according to McCormick (8), can be classified as venous angiomas, telengiectasia, cavernous angiomas and AVMs. This classification is based upon the histopathology of the vessels forming the pathology.

Yaşargil (9) pointed out that all of these anomalies may be present as occult lesions. There are several proposed theories which attempt to explain how lesions may be angiographically occult. Telengiectasias and cavernous angiomas may not be demonstrated angiographically because they are away from the major circulatory system and they have a slow circulation or a prolonged circulatory time (10,11,12). There are some reports of angiographically occult thrombosed venous angiomas with hemorrhagic findings (5,13). Leblanc et al (4) reported that 11 % of real AVMs are occult angiographically. According to Sengupta (14) the prevalence is 12-15 % in the pediatric age group. It has been claimed that, spontaneous intramural thrombus or compression by the hematoma is the cause of the lesions being occult in these cases. Spontaneous intramural thrombus may be due to flow changes, hormonal variations, changes in vascular structure, extraluminal fibrosis, vasospasm due to hemorrhage, atherosclerosis, the elongation of the feeding artery or its kinking, or intraluminal thrombosis in the abnormal vascular structure (15,16,17,18,19).

In their literature review, Wakai et al (20) report that the temporal lobe was one of the most frequent places where occult AVMs were encountered and they also describe a clinical picture of three different types. These are sudden headache or focal neurological deficit with or without unconciousness, and progressive neurological deficit and epileptic fits. The cause of the epileptic seizures is irritation due to successive subclinical hemorrhages. In our patient, the presence of a thrombus conglobated with the brain tissue, and hemosiderin present at the brain tissue adjacent to the lesion support this theory.



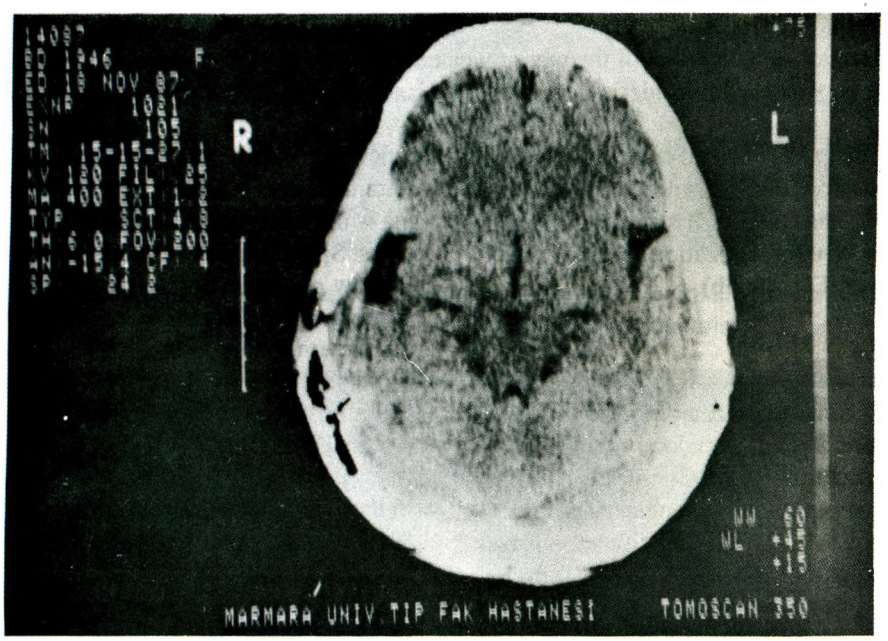


Fig. 4a and 4b : Six months later, CT scan revealed right temporal atrophy at the resection site of the mass.

The CT scan is of vital importance in the diagnosis. Although they cannot be demonstrated by conventional angiograms, with the increasing clinical use of CT and MRI scans, their chance of diagnosis has become much improved and it has been shown that many unexplained headaches and especially seizures are due to angiographically occult malformations (4,5,6,7,8,14,21,22,23). Their CT appearance is that of a uniform, hyperdense lesion. They may have no or slight contrast enhancement. The hyperdensity is explained by focal hemosiderin accumulation or by the presence of a new or old hematoma. Kucharczyk (21) and Gomori et al (22) reported that MRI is more specific in the non-calcified cases. In our case, the lesion was hyperdense in the CT scan and during surgery, hemosiderin due to recent hemorrhage and thrombus were seen.

Surgical intervention is the therapeutical method of choice. It is generally accepted that the patient must undergo surgery if there is no physical contrindication. The rational behind surgical intervention is the prevention of recurrent hemorrhages (3,24,25), ablation of the epileptic focus (26,27), and the histopathological confirmation of the underlying pathology (28,29). We share this view and believe that the patient must go to surgery unless he/she has a medical problem.

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