



Cerebral Meningioangiomatosis

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Meningioangiomatosis is a rare and benign vascular malformation of the central nervous system brought about by an intracortical perivascular proliferation of meningothelial cells and fibroblasts. These tumors are more common in young people. Clinically, it presents with seizures but may be asymptomatic throughout life. The commonest finding on a computer tomography (CT) scan is a calcified, enhancing lesion with surrounding low density and the commonest finding on a magnetic resonance imaging (MRI) scan is a low or mixed central signal on T1/T2-weighted images with a surrounding high signal on T2-weighted sequences. Surgical treatment is the standard procedure and results in satisfactory control of seizures. Here, we report a case of cerebral meningioangiomatosis. Clinical features, radiological findings, and pathologic findings are mentioned. We removed the tumor lesion in its entirety without any resulting neurological deficiencies. In this report, the diagnostic procedure and distinctive histopathology of the case are discussed, and the relevant literature is reviewed.

Key words: cerebral, histopathology, meningioangiomatosis, surgery

INTRODUCTION

Meningioangiomatosis (MA) is a rare vascular malformation of the central nervous system. It is a benign, focal proliferation of blood vessels and meningothelial cells in the brain and leptomeninges. It is an uncommon entity that occurs either sporadically or, less commonly, in patients with neurofibromatosis type 2 (NF2), in which case multifocality can be seen. Its pathogenesis has not been established; however, it is known to be associated with NF2 in nearly 50% of reported cases.

Grossly, the dense lesion thickens and hardens the affected cortex over a sharply defined plaque-like area. Fibrotic and heavily examples can resist and even dull a scalpel. Microscopical presentation varies from lesion to lesion. Some are proliferations of elongated, perivascular, fibroblast-like cells, whereas other lesions have more

obvious meningothelial features, sometimes even forming nodules or lobules similar to minute meningiomas. Psammoma bodies are present in varying numbers in both types of lesions.

In human medicine, there are two populations of patients with MA: symptomatic children and young adults who present with headaches or seizures, and asymptomatic individuals. The accurate diagnosis of MA is important since MA is a benign, surgically correctable cause of seizures.

CASE REPORT

A 19-year-old girl presented with sudden onset of syncope and seizure attack on January 10, 2005; she was conscious on admission. There was no history of central nervous system infection, head trauma, or febrile seizures. Results of physical and laboratory examinations were normal. Electroencephalographic study was also normal. Computed tomography scanning demonstrated a tumor mass with calcification in the right lateral frontal region (Fig. 1A). Magnetic resonance imaging with and without contrast revealed an abnormal signal intensity lesion of about $3 \times 2.5 \times 2$ cm in the right lateral frontal lobe adjacent to the right sylvian fissure. The mass lesion showed a low signal intensity on T1WI (Fig. 1B) and heterogeneous

Received: April 17, 2008; Revised: July 10, 2008; Accepted: July 31, 2008

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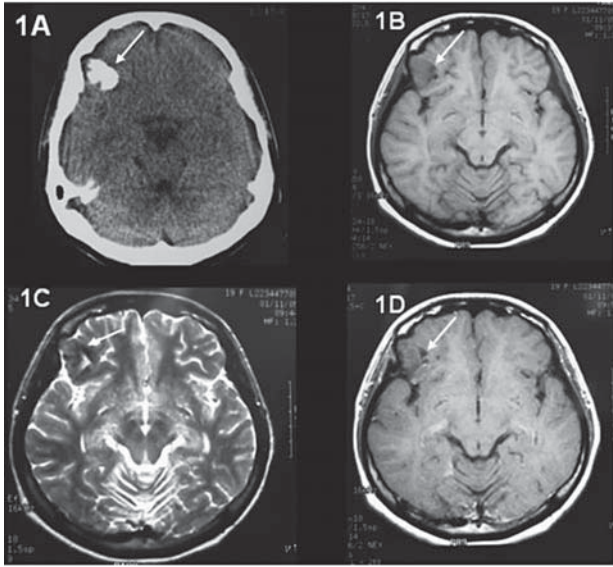


Fig. 1 1A: CT of brain without contrast showed a mass with calcification (arrow) in the right lateral frontal region. 1B: T1WI without contrast showed a low intensity lesion (arrow). 1C: T2WI without contrast revealed a heterogeneous high and low signal intensity mass (arrow). 1D: T1WI with contrast revealed a non-enhancing mass (arrow).

high and low signal intensities on T2WI (Fig. 1C). It had neither significant mass effect nor perifocal edema (Fig. 1D).

Right frontal craniotomy with removal of the tumor was performed on January 14, 2005. The operative findings (Figs 2A and 2B) were: (1) the surface of the brain was boggy and nonpulsatile, (2) no invasion of the tumor could be observed on the brain surface, (3) the tumor was lying under the cortex and was composed of stony-hard and fibrous components, and (4) the gyral pattern was maintained.

Histopathology revealed perivascular proliferations of elongated fibroblast-like cells and meningothelial cells, which ensheathed and thickened vessels. Irregularly branched thick-walled blood vessels extended into the gray matter from the meningeal surface, and proliferated blood vessels were surrounded by a wavy or concentric arrangement of spindle-formed cells in association with intervening neural tissue. Calcified psammoma bodies were identified (Fig. 3A). The connective tissue element became more prominent and large bands of dense collagen, associated with meningothelial cells, entrapped and encoached upon islands of distorted parenchyma (Fig. 3B).

The pathologic diagnosis was meningioangiomatosis.

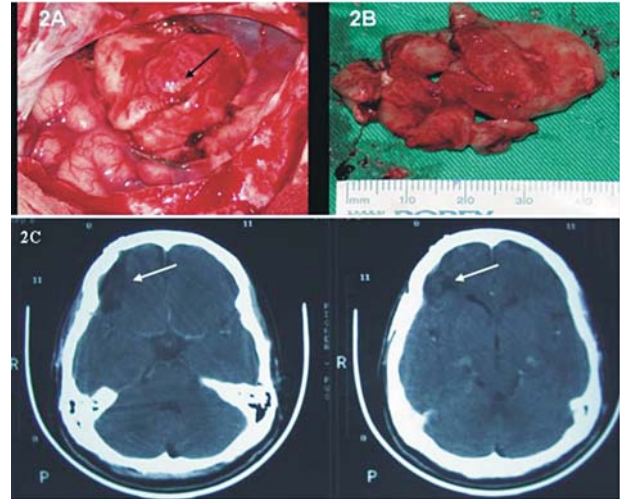


Fig. 2 2A & 2B: intra-operative findings: The surface of the brain was boggy and non-pulsatile. No proliferation and invasion of the mass could be observed over the brain surface. This mass (arrow) was lying under the cortex, and was composed of stony-hard and fibrous components. 2C: CT of brain with contrast one year after surgery: Mild brain tissue loss with encephalomalacic change (arrow) in the right lateral and inferior frontal lobe. There was no mass like lesion.

One year after operation, a brain CT scan demonstrated no evidence of residual or recurrent tumor (Fig. 2C) and the patient was free of seizures.

DISCUSSION

Meningioangiomatosis is a relatively rare, benign, and hamartomatous lesion and its pathogenesis is unclear. It is characterized by cortical meningovascular proliferation and leptomeningeal calcification^{1,2}. It occurs twice as frequently in males as in females³. Lesions may be isolated or diffuse. The sporadic lesions are single and four times more common than the diffuse forms³. The multiple diffuse forms tend to occur in association with neurofibromatosis (NF, more commonly in type 2 than in type 1) and are almost 10 times more frequent in the presence of NF.

Meningioangiomatosis involves the cortex in 90% of patients. It involves the cortical gray matter only, although imaging studies may give the impression of white matter involvement. Extracortical lesions occur three times more frequently with NF³. Seventy percent of all lesions occur in the frontotemporal region, and the single most frequent lesion location is the temporal lobe (40%). The right hemisphere is affected twice as frequently as the left

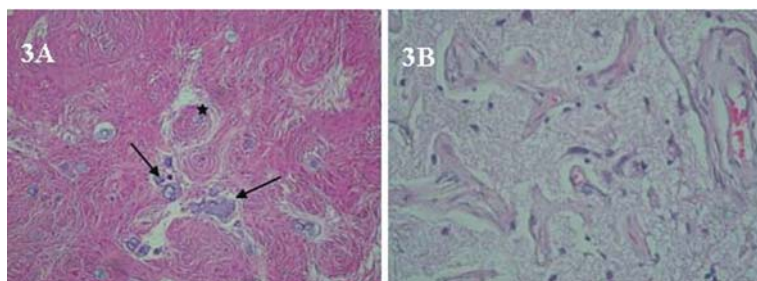


Fig. 3 3A: Proliferated blood vessels surrounded by wavy or concentric arrangement of spindle-formed cells in associated with intervening neural tissue (star). Calcified psammoma bodies (arrow) in the meninges (H & E, x 100). 3B: Irregular branched thick-walled blood vessels extending into the gray matter. (H & E, x 400).

hemisphere³. The tumor grows slowly and is not associated with features of increased intracranial pressure (IICP)⁴.

Most cases are asymptomatic; the symptomatic patient tends to present onset during childhood and early adulthood, usually with seizures. Seizures are refractory to antiepileptic drugs³. Most cases have only partial seizures without secondary generalization. Localization of seizure onset assessed by clinical features corresponds to tumor location. There is no association between location and tendency to generalized seizures. The patients who do not have seizures present with headache, facial pain, lower cranial nerve palsies, or are asymptomatic (most common).

The radiographic findings of meningioangiomatosis may show a variety of features. The commonest finding on a CT scan is a calcified, enhancing lesion with surrounding low density. The commonest finding on an MRI scan is a low or mixed central signal on T1/T2-weighted images and a surrounding high signal on T2-weighted sequences³. Contrast enhancement is common but is not the rule. In view of the wide spectrum of findings, radiographic imaging does not permit a precise preoperative diagnosis.

The electrophysiological studies of meningioangiomatosis are complex and do not reveal significant features.

Grossly, lesions are composed of a well-demarcated thickening of cortex with an overlying meningeal plaque. The gyral pattern is maintained but the brain surface may appear abnormal⁴. The dense lesion thickens and hardens the affected cortex over a sharply defined plaque-like area, with serpentine vessels on the surface, often creating the impression of a vascular malformation.

Microscopy reveals characteristic proliferation of small caliber vessels in the cortex, accompanied by a cuff of meningotheial cells and fibroblasts. Between the vascular and meningotheial proliferations, the cortex either remains normal or shows reactive astrocytic changes. Prolif-

erated blood vessels are surrounded by a wavy or concentric arrangement of spindle-formed cells⁵. Calcification is usually present in the leptomeningeal and cortical areas, both in the vessels and in the surrounding cortex⁴. The connective tissue element becomes more prominent and large bands of dense collagen, associated with meningotheial cells, entrap and encroach upon islands of distorted parenchyma.

The results of immunostaining studies vary for each meningioangiomatosis case, but the proliferating cells in this disease do not correspond to a known, normally occurring cell type. Thus, immunohistochemistry has limited diagnostic value in this disease³.

Genetic differences exist between sporadic meningioangiomatosis and meningioangiomatosis with NF. This suggests that the genetic makeup of the individual may play a role in determining the dissimilar clinical expression of histopathologically identical lesions³; additional evidence is required to clarify this issue.

Surgical treatment is the standard procedure for the management of meningioangiomatosis. The seizure outcome after surgery is variable and resection of the tumor lesion and epileptogenic cortex may be required. Seizures persist in a significant proportion of patients, despite removal of the tumor lesion and apparent confinement of epileptogenicity to one focus. No single factor emerges as the determinant of seizure outcome following resection of tumor lesions³.

In conclusion, in the case described here, the patient presented with symptoms of syncope and seizures and was diagnosed accidentally by histopathology as a case of meningioangiomatosis. The tumor was isolated and located in the gray matter of the right frontal cortex, and it was not associated with NF. It is interesting to note that, in our case, the patient presented with IICP, which is unusual in this disease. The definite reason for this was unknown, but one can speculate that it may have been caused by perifocal edema.

Preoperative diagnosis of meningioangiomatosis remains difficult because diagnostic tools lack specificity. Total surgical resection is the treatment of choice for meningioangiomatosis and the prognosis following surgery is excellent since patients are free of seizures in almost all cases (as for the case reported here). Although this type of tumor occurs infrequently, it is important to establish a correct diagnosis because of its benign histopathological features and nonrecurrent nature.

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