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Meningioangiomatosis

— A Case Report —

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Meningioangiomatosis is a rare benign hamartomatous lesion. We describe a case of meningioangiomatosis in an 18-year-old boy with a 15 year history of seizures. Computed tomography reveals an irregular calcification density along the right temporal gyrus. Microscopically, irregularly branched blood vessels, surrounded by a concentric arrangement of proliferating spindle cells, are extending into the gray matter from the meningeal surface. Ultrastructural and immunohistochemical examination failed to demonstrate features of meningothelial cell origin in this case.

This is the first case of meningioangiomatosis published in Korea along with immunohistochemical and electron microscopic studies. The pathogenesis and previous reports of this lesion will be discussed.

Key Words: *Meningioangiomatosis, Hamartoma, Brain*

INTRODUCTION

Meningioangiomatosis (MA) is a rare benign lesion which affects the cerebral cortex and occasionally the thalamus or cerebral peduncle, characterized by histologically cortical meningiovascular proliferation and calcification that may extend to involve the overlying meninges (Rubinstein, 1972). It was described by Bassoe and Nuzum in 1915 and later named by Worster-Drought and associates in 1937, so far 28 additional cases have been reported (Forster and Gagel, 1932; Hozay, 1953; Rubinstein, 1963; Rubinstein, 1972; Kasantikul and Brown, 1981; Halper et al., 1986; Sasaki et al., 1987; Kunishio et al., 1987; Kuzniecky et al., 1988; Huson et al., 1988; Liu et al., 1989; Paulus et al., 1989; Ogilvy et al., 1989; Partington et al., 1991; Goates et al., 1991; Aizpuru et al., 1991).

Previously it was commonly seen in association with von Recklinghausen's disease, but nearly two thirds of the reported cases showed no other stigmata of family history of neurofibromatosis (Kasantikul and Brown, 1981; Halper et al., 1986; Sasaki et al., 1987; Kunishio et al., 1987; Liu et al., 1989; Paulus et al., 1989; Ogilvy et al., 1989; Partington et al., 1991; Goates et al., 1991; Aizpuru et al., 1991).

We will report a recent case of MA with no evidence of von Recklinghausen's disease in which we undertook pathological, immunohistochemical and electron microscopic studies, to better elucidate the nature of this lesion. We will also review the previous reports of MA in terms of its histogenesis.

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CASE REPORT

This 18-year-old boy was admitted after suffering headaches for two weeks. At three years of age, the patient initially developed seizures characterized by 2-3 minutes episodes. He had no history of meningitis or significant head trauma. He had gone untreated until 3 years ago and since that time some anticonvulsants had been prescribed. Upon admission, he was mentally alert. Orientation and verbal communication were intact. The extraocular muscle showed a full range of movement and there was no facial palsy. And there was no evidence of pathologic reflexes. Past medical history was unremarkable. There was no family history of mental and hereditary disorders. The general physical examination was unremakable and did not

reveal ocular abnormalities. No cafe au lait patches or cutaneous nodules were found. Neurologic examination revealed a mild degree of left hemiparesis. Electroencephalograms revealed a tight temporoparietal wave. Roentgenograms of the skull were normal. Six-vessel angiography was normal, without evidence of mass, arteriovenous malformation, or tumor stain. Computed tomography revealed a lesion with irregular high density and spotty calcification with low density of edema at the right temporoparietal area (Figure 1).

A right temporoparietal craniotomy was performed. Upon opening, it was discovered that a large area of the temporal lobe was discolored a grayish-yellow. Numerous calcified granules of various sizes were found on the surface of this lesion. The lesion was relatively well demarcated from the surrounding soft brain parenchyme and there was no involvement of the overlying dura. A partial temporal lobectomy was done. The patient has remained seizure-free in the six months since surgery.

The resected specimen consisted of several irregular fragments of gray white firm brain tissue which measured approximately 1.5cc in aggregate and contained focal areas of calcification. Microscopically, irregular 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 cells in association with intervening glial tissue. Scattered psammoma bodies in the hyalinzed vascular walls were occasionally found (Figure 2). The number of adjacent neurons were reduced and neurofibrillary tangle was not seen. No proliferation of astroglial cells was discernible.

Immunostains for glial fibrillary acidic protein (GFAP) were present in subpial gliosis occuring beneath the leptomeningeal proliferations. Immunostaining for epithelial membrane antigen (EMA), S-100, desmin, and factor VIII related antigen failed to stain the main spindle cells forming the lesion in the perivascular spaces. The only positive, albeit weak, immunostaining of the spindle cells was for vimentin. Electron microscopy showed that the cells surrounding the vessels were invested by incomplete basal laminae and collagen. Proliferating cells were long and spindle-shaped, contained elongated heterochromatin-rich nuclei and were arranged concentrically (Figure 3). However, no distinct desmosomal junction or interdigitating cell membrane was found.

DISCUSSION

The pathogenesis of MA is uncertain. Russell and

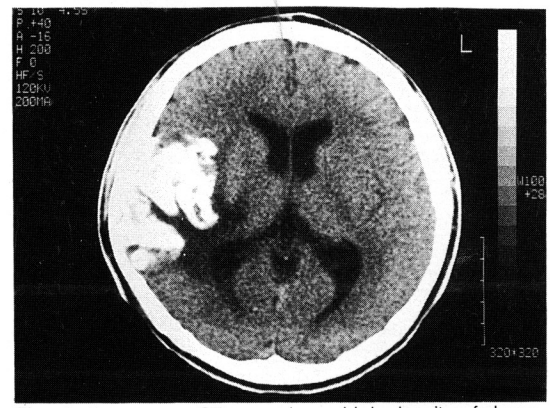

Fig. 1. Precontrast CT scan shows high density of dense cortical calcifications with low density of edema in the right frontoparietal lobe.

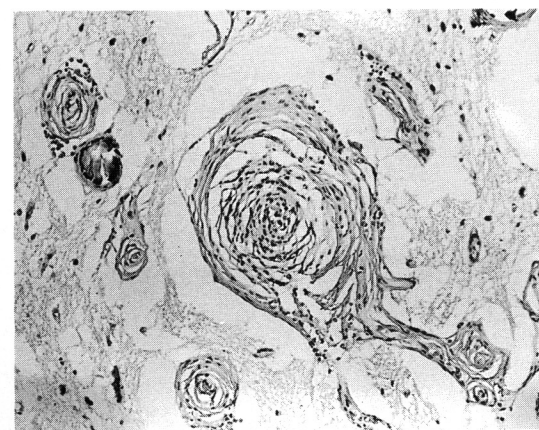

Fig. 2. Proliferated blood vessels surrounded by concentric arrangement of spindle-shaped cells in association with intervening neural tissue and multiple psammomatous calcifications (H&E, ×100).

Rubinstein (1989) thought that the pathogenesis of MA was a meningioma "en plaque" which infiltrated the brain mainly by way of the proliferating blood vessels. But, there is little to suggest pure neoplastic activity. The second hypothesis is a congenital faulty development of angiomatous and meningeal tissues (Kasantikul and Brown, 1981: Partington et al., 1991). Early onset of symptoms would suggest that the lesions are congenital or could arise as a fault of development. These lesions suffer from chronicity, as evidenced by the presence or extent of dense collagen and hyalinization of the vascular component. The third, favored possibility is that angiomatous tissue could have occurred initially with meningiomatous components arising secondarily from the perivascular elements

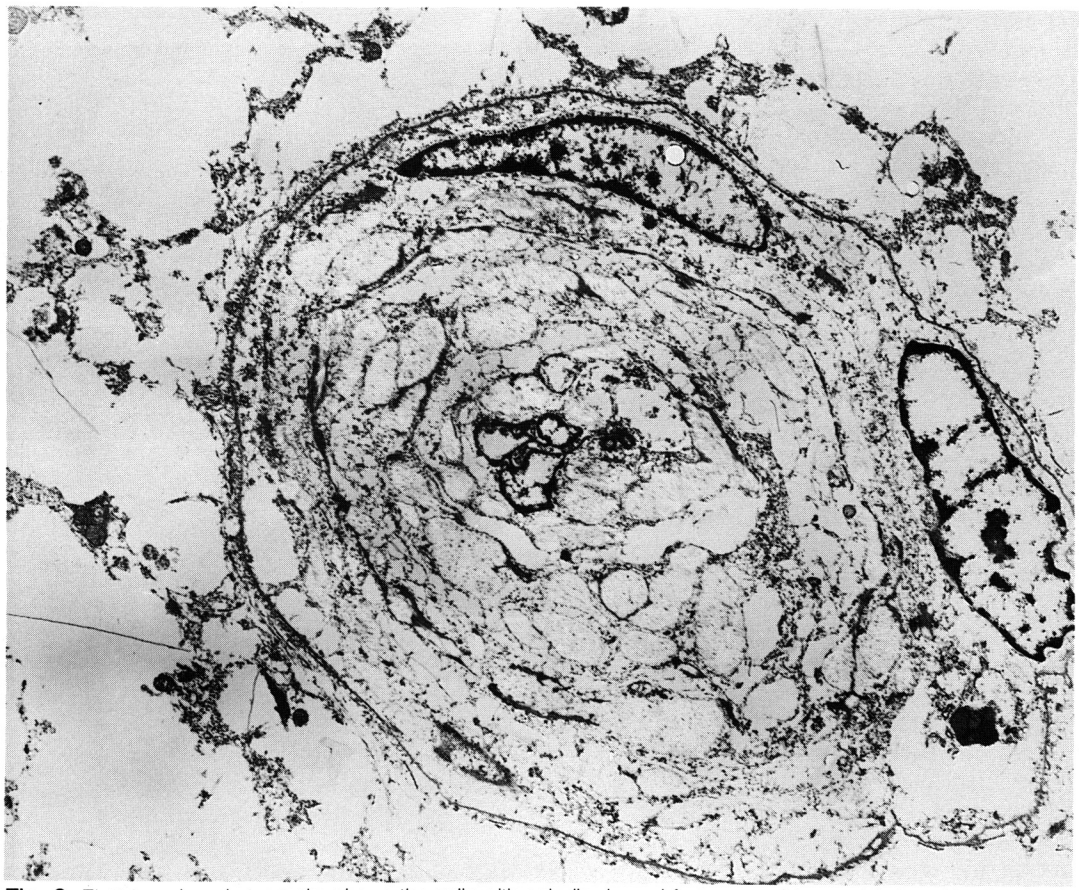

Fig. 3. Electron microphotography shows the cells with spindle-shaped features and incomplete basal lamina (×4000).

(Kasantikul and Brown, 1981). The association of MA with neurofibromatosis further supports the view that MA is a hamartoma or developmental abnormality.

The perivascular fibroblast-like cells present in MA are generally regarded as being of meningothelial origin for the following reasons: 1) direct connection with the leptomeningeal part, 2) a similarity to the cells of fibroblastic meningioma, 3) the existence of scattered psammoma bodies, 4) the electron-microscopical demonstration of desmosomal junctions and interdigitating cell membranes and 5) the negative immunohistochemical staining of glial fibrillary acidic protein, S-100 protein, factor VIII related antigen, and desmin (Paulus et al., 1989). In this case epithelial membrane antigen, which is an excellent marker of meningothelial cells (arachnoid cap cells), was not expressed by these spindle cells. The only positive immunostaining was vimentin. Ultrastructually we did not find features of meningothelial cells with interdigitating cell membrane and desmosome. Our results from immunostains and electron microscopy did not support the view of a meningothelial origin of this lesion. Rather, the main spindle cells in MA may be fibroblasts, likely derived from vessel walls. Our findings are very similar to those of an immunocytochemical study of MA (Goates et al., 1991). Goates et al. (1991) could be argued that these fibroblast-like cells might have originated from pluripotent arachnoid cap cells that selectively differentiated into a fibroblastic cell lineage. The possibility that these fibroblast like cells might have originated from pluripotent meningothelial cells, cannot be completely ruled out.

The clinicopathologic features of 20 cases of MA without neurofibromatosis including our case are represented by a history of seizure that has been either focal or generalized. Nineteen of these lesions were located in the temporal, parietal or frontal area and one case was located mulfifocally (Paulus et al., 1989). Calcifications were noted in 18 cases, but two cases lacked any leptomeningeal calcification (Kasantikul

and Brown, 1981; Paulus et al., 1989).

The findings of proliferated vascular mass with calcification in the brain suggest many possibilities, including calcified hemangioma and Sturge-Weber syndrome (Halper et al., 1986). But, the two main criteria of MA, cortical meningovascular fibroblastic proliferation and leptomeningeal calcification, permit a demarcation of MA from similar lesions. An accurate diagnosis of MA is important since most MA causes seizures and yet it is usually peripheral, benign and surgically correctable.

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REFERENCES

Aizpuru RN, Quencer RM, Norenberg M, Altman N, Smirniotopoulos J: *Meningioangiomatosis: clinical, radiologic, and histopathologic correlation. Radiology 179:819-821, 1991.*

Bassoe P, Nuzum F: *Report of a case of central and peripheral neurofibromatosis. J Nerv Ment Dis 42:785-796, 1915 (Cited from Halper et al., 1986)*

Forster O, Gagel O: *Ein Fall von Recklinghausenscher Krankheit mit funf nebeneinander bestehenden verschiedenartigen Tumorbildungen. Ztschr f d Ges Neurol u Psychiat 138:339-360, 1932 (Cited from Halper et al., 1986)*

Goates JJ, Dickson DW, Horoupian DS: *Meningioangiomatosis: an immunocytochemical study. Acta Neuropathol 82:527-532, 1991.*

Halper J, Scheithauer BW, Okazaki H, Laws ER: *Meningioangiomatosis: a report of six cases with special reference to the occurrence of neurofibrillary tangles. J Neuropath Exp Neurol 45:426-446, 1986.*

Hozay J: *Une angioneuromatoses meningo-encephalique diffuse. Rev Neurol (Paris) 89:222-236, 1953 (Cited from halper at al., 1986).*

Huson SM, Harper PS, Compston DAS: *Von Recklinghausen neurofibromatosis: a clinical and population study in South-West Wales. Brown 111:1355-1381, 1988.*

Kasantikul V, Brown WJ: *Meningioangiomatosis in the absence of von Recklinghausen's disease. Surg Neurol 15:71-75, 1981.*

Kunishio K, Yamamoto Y, Sunami N, Satoh T, Asari S, Yoshino T, Ohtuki Y: *Histopathologic investigation of a case of meningioangiomatosis not associated with von Recklinghausen's disease. Surg Neurol 27:575-579, 1987.*

Kuzniecky R, Melanson D, Robitaille Y, Oliver A: *Magnetic resonance imaging of meningio-angiomatosis, Can J Nurol Sci 15:161-164, 1988 (Cited from Partington et al., 19991).*

Liu SS, Johnson PC, Sonntag VKH: *Meningioangiomatosis: a case report. Surg Neurol 31:376-380, 1989.*

Ogilvy CS, Chapman PH, Gray M, Monte SM: *Meningioangiomatosis in a patient without von Recklinghausen's disease. J Neurosurg 79:483-485, 1989.*

Partington CR, Graves VB, Hegstrand LR: *Meningioangiomatosis. Am J Neuroradiol 12:549-552, 1991.*

Paulus W, Peiffer J, Roggendorf W, Schuppan D: *Meningio-Angiomatosis, Path Res Pract 184:446-452, 1989.*

Rubinstein LJ: *Tumeurs et hamartomes dans la neurofibromatose centrales. In: Michaux L, Feld M, eds. Les phakomatoses cerebrales. Paris: S.P.E.I. B: 426-451, 1963 (Cited from Halper et al., 1986).*

Rubinstein LJ: *Tumors of the central nervous system. In: Atlas of tumor pathology, 2nd series, Fascicle 6, Washington DC: Armed Forces Institute of Pathology, pp252, 1972.*

Russell DS and Rubinstein LJ: *Pathology of tumors of the nervous system. Edward Arnold, London, pp777-779, 1989.*

Sakaki S, Nakagawa Nakamura K, Takeda S: *Meningioangiomatosis not associated with von Recklinghausen's disease. Neurosurgery 20:797-801, 1987.*

Worster-Drought C, Dickson WEC, McMenemey WH: *Multiple meningeal and perineural tumours with analogous changes in the glia and ependymoma (neurofibromatosis). Brain 60:85-117, 1937 (Cited from Halper et al., 1986)*