

Meningioma Arising from Meningioangiomas Without Neurofibromatosis

- A Case Report -

Jae Hong Park • Seung-Yeon Ha
Na Rae Kim

Department of Pathology, Gachon
University of Medicine and Science,
Gil Medical Center, Incheon, Korea

Received : January 17, 2007
Accepted : March 2, 2007

Corresponding Author

Na Rae Kim, M.D.
Department of Pathology, Gachon University of
Medicine and Science, Gil Medical Center, 1198
Guwol-dong, Namdong-gu, Incheon 405-760, Korea
Tel: 032-460-3847
Fax: 032-460-3073
E-mail: naraech@empal.com

We report a rare case of meningioma associated with meningioangiomas in a 9-year-old male patient who showed none of the stigmata of neurofibromatosis 2. Brain magnetic resonance images showed marked cortical calcification with slight contrast-enhancement in the parieto-occipital lobe. The resected mass showed that the lesion was mainly composed of meningioangiomas and a small focus was transformed into meningioma. To date, only 17 cases of such combined lesions have been reported in English medical literature. We report a rare case of meningioma that arose from meningioangiomas.

Key Words : Meningioangiomas; Meningioma

A 9-year-old boy visited the hospital for evaluation of head trauma because he had slipped down two weeks before. The results of detailed head and neck, cranial nerve, motor, sensory, and cerebellar examinations were all within normal limits. He had experienced one brief medically intractable seizure five years ago. He had no stigmata of neurofibromatosis 2. T2-weighted magnetic resonance (MR) images revealed that the lesion was located in the leptomeninges and there was cortical calcification with contrast-enhancement in the left parieto-occipital lobe (Fig. 1). The lesion was completely excised. The resected specimen measured $3.0 \times 2.9 \times 2.0$ cm and it weighed 8 grams. The cut surface was uniformly gray white with focal calcification. Hemorrhage or necrosis was not found. Microscopically, the majority of the mass showed proliferation of small blood vessels and vasocentric spindle cell proliferation, which were compatible with meningioangiomas (MA). Further, small foci of the mass that measured about $0.8 \times 0.5 \times 0.5$ cm were transformed to a meningotheiomatous meningioma (Fig. 2, 3). Immunohistochemically, both the MA and the meningioma were positive for vimentin and focally positive for EMA. The Ki-67 pro-

liferation index, GFAP and p53 in the MA and meningioma were all negative.

MA was first described by Basso and Nuzum in 1915 as an incidental autopsy finding in a 15-year-old boy with neurofibromatosis 2.¹ MA is a rare hamartomatous lesion, which is accompanied by neurofibromatosis 2 or occurs sporadically.² Its histogenesis remains unclear, and it has variously been considered as a vascular malformation, a hamartoma, a neoplasm or as metaplasia. It is histologically characterized by a proliferation of cortical capillary-sized vessels and perivascular cells with the leptomeningeal calcifications. The involved cells are thought to be fibroblastic or meningotheiomatous origin.³ Clinically, these MA patients usually present with seizures or persistent headaches, and they can have a benign course through life. However, MA has been rarely described coexisting with meningiomas, arteriovenous malformations, encephaloceles, oligodendrogliomas, meningeal hemangiopericytomas and orbital erosion. Among them, meningioangiomas with meningioma is the most frequent combination. To date, only 17 cases of meningiomas derived from MA have been reported.⁴ Among them, the largest

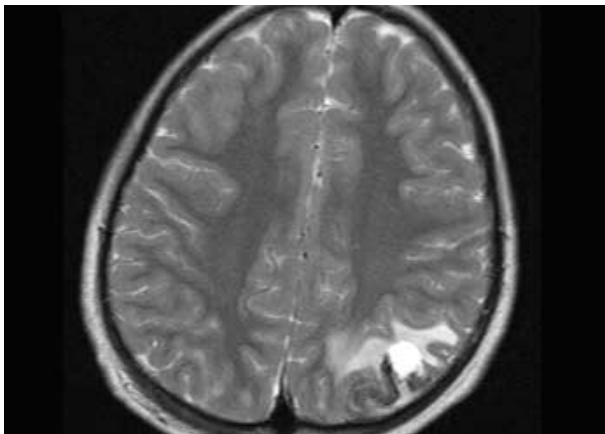


Fig. 1. MRI reveals that the lesion shows irregular peripheral rim enhancement pattern, focal meningeal enhancement adjacent to the mass and surrounding edema.

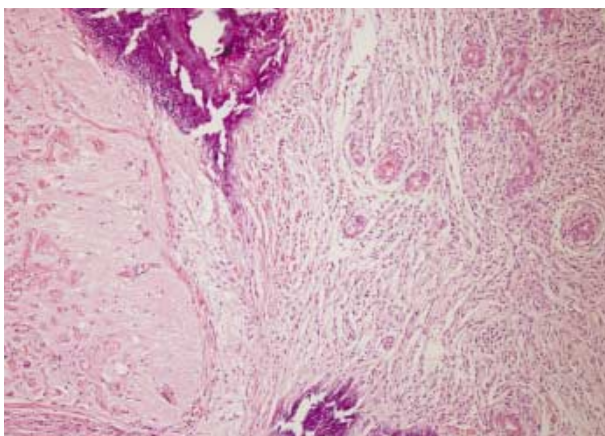


Fig. 2. Transition from meningoangiomatosis (left side) to meningioma (right side).

series is a reported collection of Korean cases.⁵ The combination of MA and meningioma, like our case, generally occurs in children and young adults. This stands in contrast with ordinary meningiomas, for which the peak occurrence is in the fifth and sixth decades of life. Although there have been many debates, MA may primarily be a meningotheelial lesion that occasionally proliferates to produce a solid mass of a meningioma. Moreover, recent molecular reports have shown the loss of heterozygosity of the neurofibromatosis 2 gene locus (22q12) chromosome in both meningioma and MA; this raises doubts about the presumed hamartomatous nature of MA and it supports a neoplastic relationship.^{2,6} In contrast, the perivascular spindle cells of pure MA without meningioma are genetically and immunohistochemically similar to the non-neoplastic meningotheelial cells in a study of Perry *et al.*⁴ That is, there is a very low Ki-67 pro-

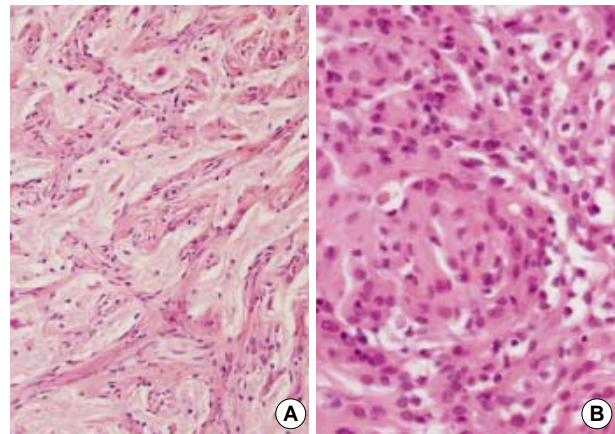


Fig. 3. (A) The majority of the calcific mass is meningoangiomatosis portion which shows proliferation of small blood vessels and perivascular spindle cells. (B) Note high magnification of meningioma portion.

liferation index or it is absent altogether, and there is no progesterone receptor-immunoreactivities or gene deletions (NF2/4.1B), and there are occasional immunonegativities for merlin or protein 4.1B in pure MA, while most of MAs with meningioma have gene deletions (NF2/4.1B), protein losses (merlin/protein 4.1B), a slightly high Ki-67 proliferation index and/or progesterone receptor-immunoreactivity. In addition, both MA and meningioma have similar or identical phenotypes. Their study supports the neoplastic theory of MA: in MA meningiomas, the MA component is neoplastic, and it even represents an exuberant perivascular pattern of spread from the leptomeningeal meningioma, rather than an underlying hamartoma. Despite the lack of a Ki-67 proliferation index in both the MA and the meningioma component, it is difficult to determine whether our present case is applicable to the neoplastic theory or hamartomatous theory. In our opinion, further molecular studies focusing on a large series of MA-meningiomas are necessary to evaluate the pathogenesis of both MA and its derived meningioma.

REFERENCES

1. Bassoe P, Nuzum F. Report of a case of central and peripheral neurofibromatosis. *J Nerv Ment Dis* 1915; 42: 785-96.
2. Takeshima Y, Amatya VJ, Nakayori F, Nakano T, Sugiyama K, Inai K. Meningoangiomatosis occurring in a young male without neurofibromatosis: with special reference to its histogenesis and loss of heterozygosity in the NF2 gene region. *Am J Surg Pathol* 2002; 26: 125-9.

3. Fuller GN. Central nervous system tumors. In: Parham DM, ed. *Pediatric neoplasia: morphology and biology*. Philadelphia: Lippincott-Raven, 1996; 153-204.
4. Perry A, Kurtkaya-Yapicier O, Scheithauer BW, *et al*. Insights into meningioangiomatosis with and without meningioma: a clinicopathologic and genetic series of 24 cases with review of the literature. *Brain Pathol* 2005; 15: 55-65.
5. Kim NR, Choe G, Shin SH, *et al*. Childhood meningiomas associated with meningioangiomatosis: report of five cases and literature review. *Neuropathol Appl Neurobiol* 2002; 28: 48-56.
6. Sinkre P, Perry A, Cai D, *et al*. Deletion of the NF2 region in both meningioma and juxtaposed meningioangiomatosis: case report supporting a neoplastic relationship. *Pediatr Dev Pathol* 2001; 4: 568-72.