Meningioangiomatosis Not Associated with 
Von Recklinghausen’s Disease
—Case Report—

Kenta FUJIMOTO, Yuji NIKAIDOH, Takashi YUASA, Kiyoshi NAGATA,
Yuhki IDA, Masayuki FUJIOKA, Hideyuki OHNISHI*
and Shinichiroh KUROKAWA**

Department of Neurological Surgery, Osaka Minami National Hospital, Kawachinagano,
Osaka; *Department of Neurological Surgery, Osaka Neurological Institute, Toyonaka,
Osaka; **Department of Neurological Surgery, Nara Medical University, Kashihara, Nara

Abstract
A 7-year-old girl with meningioangiomatosis not associated with von Recklinghausen’s disease is de-
scribed. The radiological findings were similar to meningioma, but intraoperatively, a thin septum was
found between the mass and the dura mater. Microscopically, there was significant proliferation of
fibroblastic spindle-shaped cells and collagenous fibers in the subarachnoid space. Proliferated cells
had penetrated into the cortical tissue along the irregularly branched blood vessels. Immunohisto-
chemically, these penetrating perivascular cells were negative for glial fibrillary acidic protein and
S-100 protein, and positive for vimentin staining. These findings suggest that the histogenesis of the
spindle-shaped cells is most probably meningothelial.

Key words: meningioangiomatosis, von Recklinghausen’s disease, magnetic resonance imaging

Introduction
Meningioangiomatosis is a rare hamartomatous le-
sion characterized histologically by focal cortical
meningovascular proliferation and calcification, and
is generally associated with von Recklinghausen’s
neurofibromatosis. However, several cases of me-
ningioangiomatosis without neurofibromatosis have
been reported. Here we describe another patient with
meningioangiomatosis without neurofibromatosis,
and discuss this and previous cases.

Case Report
A 7-year-old girl was admitted to our clinic in
November, 1989 with loss of unconsciousness and
vomiting. She recovered some 5 hours later. There
were no stigmata or family history of
neurofibromatosis. On admission, general, physical,
and neurological examination found no abnor-
amalities. Computed tomographic (CT) scans showed
a right occipital calcified lesion, and marked en-
hancement postcontrast (Fig. 1). Magnetic reso-
nance (MR) imaging showed an inhomogeneous
low-intensity mass on the T2-weighted image, which
appeared isointense on the T1-weighted image (Fig.
2). No gadolinium-enhanced MR imaging was
available. Angiograms showed no evidence of a
mass, vascular malformation, or tumor stain. Elec-
troencephalograms showed a 4–5 Hz theta-wave
burst and sharp-wave discharges over the right pari-
eto-occipital region.

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Authors’ present addresses: K. Fujimoto, M.D., Department of Neurological Surgery, Osaka Prefectural Hospital, Osaka,
Japan; T. Yuasa, M.D., Department of Neurosurgery, Yukokai Hospital, Nara, Japan; Y. Ida, M.D., Department of Neurosurgery, National Senboku Hospital, Sakai, Osaka, Japan; M. Fujioka, M.D., Department of Emergency Medicine, Nara Medical University, Kashihara,
Nara, Japan.
The lesion was totally removed via a right occipital craniotomy. The lesion was firm and well defined (Fig. 3), touching the falx, but separated by a thin capsule. Microscopically, there was very significant leptomeningial proliferation composed of fibroblastic spindle-shaped cells interpolated with collagenous fibers in the subarachnoid space, and psammomatous calcification in the leptomeninges. Proliferated spindle-shaped cells had penetrated into the cortical tissue along irregularly branched blood vessels. Neurons were seen in the neural tissue (Fig. 4). The penetrating perivascular cells were negative to S-100 protein and glial fibrillary acidic protein (GFAP), and positive to vimentin immunohistochemical staining (Fig. 5).

Postoperatively, she developed left homonymous hemianopsia, but seizures were controlled by sodium valproate.

**Discussion**

Partington et al.\(^\text{10}\) classified meningioangiomatosis into two types: 1) symptomatic children and young adults presenting with seizures or headaches, and 2) asymptomatic individuals (mostly with neurofibromatosis) in whom meningioangiomatosis was discovered as an incidental finding. Table 1 summarizes the clinical features of the reported cases of meningioangiomatosis without neurofibromatosis.\(^\text{1,2,4-12}\) Patient ages ranged from 1 to 70 years (mean 19.2 yrs). There were 13 males and eight females. Many lesions were located in the right lobe (right 16, left 5), frequently in the temporal lobe (13 cases). The clinical presentation of seizures is the most common presenting symptom, as in our patient. One patient presented with left hemiparesis.\(^\text{11}\) Other lesions have been discovered incidentally after head trauma or at autopsy.\(^\text{2,5}\)

The CT appearance of the lesion in our case was similar to previous reports in which the lesion was hyperdense, with remarkable enhancement postcontrast. MR imaging showed the lesion as isointense on the T\(_1\)-weighted image, and hypointense on the T\(_2\)-weighted image. Previous reports indicated that MR imaging showed hypointense areas on both T\(_1\)- and T\(_2\)-weighted images.\(^\text{1,6-10}\) Angiography showed abnormal vessels in only one reported case, and normal or avascular mass in the other cases and in our present case. Therefore, the abnormal vascular component of meningioangiomatosis seems to behave as an occult cerebral vascular malformation angiographically.

The histological features observed in our patient are similar to those previously reported, including
leptomeningial fibrous proliferation, psammomatous calcification in the leptomeninges, and penetration by spindle-shaped cells into the cortical tissue along the malformed vessels. These cells stained negatively for S-100 protein, GFAP, desmin, and cytokeratin, and positively for vimentin. These findings indicate that the histogenesis of the spindle-shaped cells is most probably meningothelial. There were neurocytes in the cortical tissue among these leptomeningial cells, but whether or not these neurocytes were functioning is unknown. There were neurofibrillary tangles of the neurons within the lesion and adjacent cortex in six previous cases,\(^{2,7,11}\) indicating degenerative change.\(^3\) This suggests that these neurons are not functioning or are hypofunc-

Fig. 4 Photomicrographs of the tumor specimen. A, B: Irregularly branched thick-walled blood vessels (arrow) penetrating into the cortical tissue from the meningeal surface. left, arachnoid side; right, cortical side. HE stain, ×100. C: These blood vessels are surrounded by collagenous fibers and spindle-shaped cells. L: blood vessel lumen, S: surrounding cells. HE stain, ×400. D: Neurocytes (arrow) among intervening neural tissue. HE stain, ×400.

Fig. 5 Immunohistochemical staining of the tumor specimen, showing penetrating perivascular cells negative for GFAP stain (upper: ×100) and positive for vimentin stain (lower: ×100). left, arachnoid side; right, cortical side.

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This disorder is thought to be a benign hamartoma. Therefore, patients with medically controllable seizures and asymptomatic patients should be treated conservatively. Surgery is advised if seizures are medically intractable or the lesion is easily accessible. However, no previous case was diagnosed as meningioangiomatosis preoperatively, and definite diagnosis prior to surgery is quite difficult.

References


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*Address reprint requests to: K. Fujimoto, M.D., Department of Neurological Surgery, Osaka Prefectural Hospital, 3-1-56 Mandai-higashi, Sumiyoshi-ku, Osaka 558, Japan.*