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# Meningioangiomatosis: MR imaging and pathological correlation in two cases

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University Children's Hospital, 28 Yongon-dong, Chongno-gu, Seoul 110-744, South Korea Abstract Meningioangiomatosis is a rare, benign neoplastic disorder involving the cortex and leptomeninges. The pathological findings are characterised by proliferation of meningothelial cells and leptomeningeal vessels and calcifications within the mass. We experienced two cases of pathologically confirmed meningioangiomatosis, one as a solitary cortical mass with calcification and the other as a cortical lesion manifested as extensive intracranial haemorrhage. On MRI, the first case showed an isointense cortical mass in the left frontal lobe and homogeneous enhancement on the

contrast-enhanced study. The second case showed a target-like lesion with a peripheral dark signal rim on T2-weighted images accompanied by extensive haemorrhage in the adjacent frontal lobe and lateral ventricles.

Keywords Meningioangiomatosis · MR imaging · Pathological correlation

### Introduction

Meningioangiomatosis (MA) is a rare, benign hamartomatous lesion of cerebral cortex and leptomeninges [1]. Pathologically, it is characterised by cortical meningovascular proliferation and leptomeningeal calcifications [2]. The common clinical symptoms are headache and seizure. Although the aetiology and pathogenesis are unknown, neurofibromatosis is associated in half of the patients with MA. We report the imaging and pathologic findings of two cases of MA.

# **Case reports**

#### Case 1

An 8-year-old boy had a 7-year history of recurrent, simple partial seizures. During a seizure he had clonic movement predominantly in the right upper extremity. The EEG showed rhythmic spikes in the left temporo-parietal lobe and both frontal lobes. MRI revealed

a  $3\times2\times4$ -cm, ill-defined subcortical mass in the left frontal lobe that had a broad base on the adjacent dura. The signal intensity of the tumour was isointense with the cortex on T1-weighted (T1-W) and T2 weighted (T2-W) images. On contrast-enhanced images, the tumour was better defined with mild enhancement, and the attached dura enhanced moderately (Fig. 1). Dark signal spots suggestive of calcifications were identified within the mass on all sequences. The mass was diffusely decreased in signal intensity on the gradient-echo image, suggesting associated haemorrhage within the tumour (Fig. 1d).

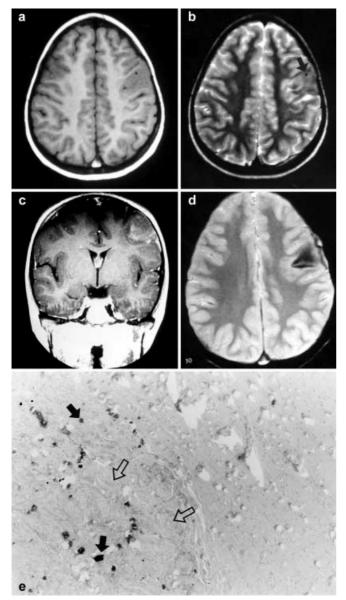
During surgery, the tumour was found in the left precentral gyrus as an ovoid cortical mass with firm consistency. It was easily demarcated from the surrounding normal tissue and there was no invasion of the overlying dura.

Microscopic examination (Fig. 1e) revealed leptomeningeal and vascular proliferations and some psammomatous calcifications within the mass, which was compatible with meningioangiomatosis. Some foci of haemorrhage were revealed in the tumour.

The patient has remained free of seizures since surgery.

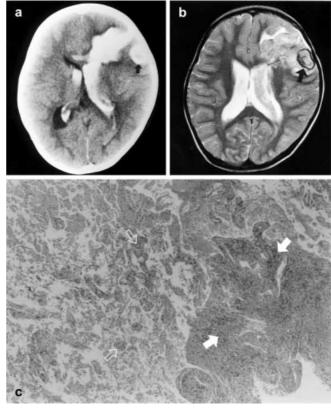
#### Case 2

A 3-year-old boy complained of headache and suffered a seizure 3 days after minor head trauma. CT (Fig. 2a) showed extensive



**Fig. 1a–e** An 8-year-old boy with partial seizures for 1 month. **a** An ill-defined isointense mass in the subcortical area of the left frontal lobe on T1-W image. The mass extends to the surface and has a broad base. **b** On T2-W image, the mass shows heterogeneously intermediate and low-signal intensity, including a darksignal intensity focus (*arrow*) suggesting calcification. **c** On contrast-enhanced image, the mass demonstrates mild homogeneous enhancement, and there is prominent enhancement of adjacent meninges over the mass. **d** On gradient-echo image, the mass shows diffuse signal decrease which suggests the presence of haemorrhage. **e** Meningothelial cells and leptomeningeal vascular proliferations (*open arrow*) are demonstrated on light microscopic examination. Numerous psammomatous calcifications (*arrows*) are seen

haemorrhage in the left frontal lobe extending into both lateral ventricles and crossing the midline along the corpus callosum. On MRI, an ovoid target-like lesion surrounded by a peripheral dark signal rim was seen in the left posterior frontal cortex accompanied



**Fig. 2a–c** A 3-year-old boy with headache and seizure for 1 day. **a** On unenhanced CT, extensive haemorrhages are noted in the left anterior frontal lobe, crossing the midline along the corpus callosum and extending into both lateral ventricles. A welldefined high-density mass (*arrow*) with central low density is noted posterior to the parenchymal haemorrhage in the left frontal lobe. **b** On T2-W image, acute extensive parenchymal and intraventricular haemorrhage is seen in the left frontal lobe and a well-defined target-like lesion (*arrow*) surrounded by low-signal rim is present in the posterior portion of the haemorrhage. **c** Light microscopic finding shows numerous vessels (*arrows*) surrounded by meningothelial cells. Foci of haemorrhage (*open arrows*) are present

with haemorrhage, which showed intermediate-to-high-signal intensity on proton and T2-W images (Fig. 2b). Haemorrhage surrounding the mass was heterogeneously intermediate and highsignal intensity on proton-density and T2-W images, crossing to the right cerebral hemisphere through the genu of the corpus callosum and extending into both lateral ventricles. On angiography, there was no evidence of vascular malformation or hypervascular mass.

Emergency left frontal craniotomy and excision of the mass were performed. The haematoma was present in the left fronto-temporal lobe and a well-demarcated cortical mass of slightly hard consistency, measuring about  $3\times 2\times 2$  cm, was found posterior to the haematoma.

On microscopic examination, proliferated meningothelial cells were seen with abundant vessels (Fig. 2c). Innumerable red blood cells due to haemorrhage were scattered around the mass but there was no evidence of a vascular malformation.

The patient has been free of seizures since surgery.

## Discussion

MA is a rare, benign, hamartomatous intracranial lesion, characterised by a cortical mass and leptomeningeal hypervascularisation. It usually affects children and young adults. The first case was described by Bassoe and Nuzum in 1915 as an incidental autopsy finding in a 15-year-old boy and it was first named by Worster-Drought et al. in 1937 [1,4]. The most common symptoms are headaches and seizures, but it is sometimes asymptomatic in older patients, or in patients with neurofibromatosis or other congenital disorders [1, 2,3]. MA is associated with neurofibromatosis in nearly half of the patients, there being a stronger association with type II than with type I [4]. Our patients had no stigmata or family history of neurofibromatosis.

Although the pathogenesis is unknown, three possible theories have been suggested. First, the lesions could be hamartomas; second, they could be secondary to direct invasion of brain tissue by a leptomeningeal meningioma; third, they could represent a vascular malformation [3].

The pathological criteria may be summarised as leptomeningeal proliferation of nodules, or whorls or bands of meningothelial (i.e. arachnoid cap) cells exhibiting marked degenerative reactions such as calcification, fibrocartilage or bone formation in association with sharply demarcated intracortical plaques of proliferating small vessels and perivascular cuffs of spindleshaped fibroblast-like cells [5]. Calcifications are variable, from psammomatous to dense osteoid type [3]. In the first case, the tumour contained psammomatous calcifications on microscopic examination.

The imaging findings of MA are variable [1, 2, 3, 4, 5,6]. It is usually seen as a solitary cortical lesion, although multifocal lesions have been reported [4]. It may manifest as an intra-axial or extra-axial lesion [2]. It is usually located in the frontal or temporal lobe [2], but may unusually locate in the brainstem [4]. The reported CT findings of MA are hypodense, round masses containing various types of calcification in the cortical or leptomeningeal area with little or no contrast enhancement [1, 2, 3,4]. In several cases, MR findings have been reported; abnormalities are confined to the cortex and consist of isointensity or hypointensity on T1-W images and a heterogeneous cortical mass surrounded by an area of increased intensity on T2-W images, probably due to oedema or gliosis. Contrast-enhanced images are not described in most cases but when reported it varies from no enhancement to strong enhancement [1, 2, 3,4].

Our second case was unusual in that extensive haemorrhage was present around the tumour and within the lateral ventricle; cerebral angiography revealed no other cause of haemorrhage such as vascular malformation or tumour vascularity. Therefore, we assume that the cause of the haemorrhage may have been due to hypervascularity of the tumour complicated by head trauma.

MA is a slowly growing tumour that is always benign without malignant potential [2]. Therefore, accurate diagnosis is important because the prognosis after excision is excellent. However, differential diagnosis from other cortical tumours such as oligodendroglioma, ganglioglioma, dysembryoplastic neuroepithelial tumour (DNET), metastasis, meningioma and granulomatous meningitis is not easy because of non-specific radiological characteristics of the lesion [4,6]. In a case with haemorrhage-associated MA, like the second case, vascular malformation should be considered as differential diagnosis.

In conclusion, the imaging findings of MA are characterised by a cortical or subcortical mass with variable calcification and enhancement, which is sometimes difficult to differentiate from other cortical lesions. Although it is unusual, intracranial haemorrhage can be a major abnormality of MA as seen on the second case.

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