MR Imaging of Intracranial Pediatric Meningiomas: Manifestations in 16 Patients

Hong Eo, Ji Hye Kim

Purpose : To describe the clinical, MR imaging, and pathologic findings of pediatric meningiomas.

Materials and Methods : The authors retrospectively reviewed the medical records and MR images of 16 pediatric patients with pathologically proven meningioma. Mean patient age at diagnosis was 14 years (range, 3–18). MR images were reviewed for details of lesion sizes, locations, signal intensity (SI), marginal characteristics, internal architectures, enhancements, and dural and parenchymal changes. The findings of other imaging modalities and of pathological examinations were also analyzed.

Results : Mean tumor size was 5.24 cm (range, 1.3–18.1 cm) and locations were supratentorial in 12 and infratentorial in 4. SI of masses were variable, that is, high in 9, iso in 4, and low in 3 on T2 weighted images (T2WI), and low (n = 11), iso (n = 4), or high (n = 1) on T1WI images. All lesions were visualized as well-demarcated enhancing masses. Five of the tumors were heterogeneous with cystic or necrotic components. Dural attachment was observed in 11 patients and adjacent brain edema in 10. Tumors exhibited hyperdense (n = 6) or isodense (n = 4) on non-enhanced CT scans, and 3 of the 7 angiograms demonstrated blood supply from the internal carotid artery. Pathologic examinations revealed the following subtypes; transitional cell (n = 4), meningotheliomatous (n = 4), chordoid (n = 2), fibrous (n = 2), clear cell (n = 1), hyalinized (n = 1), rhabdoid papillary (n = 1), and atypical (n = 1).

Conclusion : Pediatric meningiomas occur usually in teenagers, have diverse pathological types, and may produce atypical imaging findings, such as, a heterogeneous internal content or findings suggestive of intraaxial tumors.

Index words : Brain neoplasm
Meningioma
Childhood

Introduction

Meningioma is a common tumor of the central nervous system in adults, and accounts for 10-20% of all primary intracranial tumors. However, meningioma is rare in children, with an incidence of between 0.85-2.3% (1–7). Furthermore, meningiomas in childhood

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tend to have clinical and pathological findings that differ from those in adults. For example, meningiomas in adulthood show a female predilection, but males are more frequently affected during childhood. The MR imaging characteristics of typical meningiomas are homogeneous well-enhancing mass exhibiting high or iso- signal intensity (SI) on T2-weighted images (T2-WI) and low or iso- SI on T1 weighted images (T1-WI). They are usually observed at the convexity or in the parasagittal region with a significant dural tail. The meningotheliomatous subtype is most common (8). In contrast to meningiomas of adulthood, fewer radiologic and histopathologic findings have been reported in childhood. Findings reported include; larger tumors, cysts, unusual sites, such as, the lateral ventricles and posterior cranial fossa, a lack of dural attachment, and a more malignant nature (9, 10).

In the present study, we reviewed 16 intracranial meningiomas in children, and describe their clinical, MR imaging, and pathologic findings and compare these with meningiomas of adulthood.

Materials and Methods

Medical records and MR images were retrospectively reviewed to identify pediatric patients with pathologically proven meningioma. Spinal meningiomas and meningiomas associated with neurofibromatosis type II were excluded. Finally, a total of 16 intracranial meningiomas were identified in patients less than 19 years old and included. Clinical findings are summarized in table 1. All patients, except one, were more than 10 years old at diagnosis and mean patient age was 14 (range, 3–18). There were six girls and ten boys. Presenting symptoms were a headache (n=8), a seizure (n=4), a tingling sensation (n=2), and extremity weakness (n=1). In one patient, meningioma was detected incidentally after traffic accident.

Preoperative MR imaging was performed in all patients using variable 1.5/3.0T units (GE/Siemens/

Table 1. Patient Characteristics

Case no.	Age (years)	Gender	Neurological Presentations
1	3	F	Motor weakness
2	18	F	Headache
3	15	M	Headache
4	10	F	Seizure
5	17	F	Headache
6	14	M	No symptom
7	16	M	Headache
8	14	M	Tingling sense
9	14	M	Seizure
10	13	M	Seizure
11	16	M	Tingling sense
12	17	F	Headache
13	12	M	Headache
14	14	M	Headache
15	15	F	Seizure
16	14	M	Headache

Table 2. Summary of MR Imaging and Pathological Findings

Case	MRI Findings				Pathology
	Location	Cyst	Edema	Dural Attachment	
1	Left parietooccipital convexity	Yes	Yes	Yes	Meningotheliomatous
2	Left CPA	No	Yes	No	Transitional
3	Left lateral ventricle	No	No	No	Hyalinized
4	Right parasagittal	No	No	Yes	Meningotheliomatous
5	Right temporal convexity	No	Yes	Yes	Transitional
6	Left CPA	No	No	Yes	Fibrous
7	4th ventricle	No	No	No	Clear cell
8	Right frontoparietal lobe	Yes	Yes	Yes	Chordoid
9	Right temporal convexity	No	No	Yes	Atypical
10	Frontal convexity	No	Yes	Yes	Transitional
11	Right temporal lobe	No	Yes	No	Fibrous
12	Left lateral ventricle	No	Yes	Yes	Transitional
13	Right frontal lobe	Yes	Yes	No	Rabdoid papillary
14	Left occipital lobe	Yes	Yes	Yes	Chordoid
15	Clivus	No	Yes	Yes	Meningotheliomatous
16	Left frontoparietal convexity	No	No	Yes	Meningotheliomatous

Philips). Axial T1-WI and T2-WI, sagittal T1-WI, coronal T2-WI, and two or three planes of contrast enhanced T1-WI were available for all patients.

On MR images, we reviewed lesion locations, sizes, SI, marginal characteristics, internal architectures, and enhancement patterns. The presences of dural attachment and adjacent brain parenchymal changes, such as, edema, were also reviewed. In addition, CT scans (n=10) and angiograms (n=8) were available, and were reviewed for CT attenuation, presence of calcification, tumor staining, and blood supply. Preoperative diagnosis, operation records, and pathological examination findings were also reviewed.

Results

MR imaging findings

MR imaging and pathologic findings are summarized in table 2.

Supratentorial lesions accounted for 12 (75%) of the 16 cases. Individual tumors were located at the cerebral convexity (n=5), the lateral ventricle (n=2), and the parasagittal region (n=1). Four supratentorial tumors were intraparenchymally located in frontal, frontoparietal, temporal, and occipital lobes, respectively. Four infratentorial tumors were located in the cerebellopontine angle (CPA, n=2), the 4th ventricle (n=1), and clivus (n=1). Tumors longest diameters ranged from 1.3 to 18.1 cm (average 5.24cm). Nine (56%) tumors had high SI on T2-WI images and 4 and 3 tumors exhibited iso- and low SI, respectively. On T1-WI images, the 16 tumors exhibited low (n=11), iso (n=4), or high (n=1) SI. All tumors were well-demarcated and five (31%) of the tumors were heterogeneous with cystic or necrotic components while the remaining 11 were homogenous. and homogeneous or heterogeneous enhancement was noted on post-contrast images (Fig. 1). Adjacent dural enhancement, suggesting dural attachment, was observed in 11 (69%) cases. The histopathologic

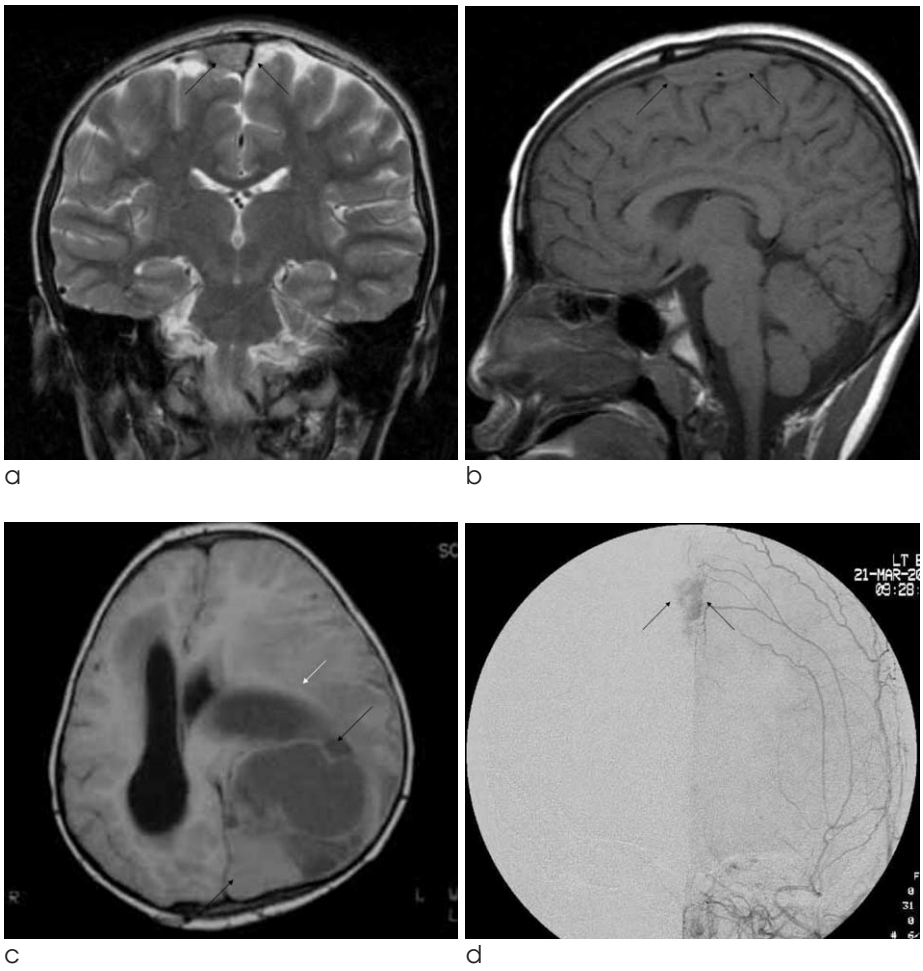

Fig. 1. A 10-year-old girl that presented with seizure.
T2WI image (a) showing a slightly hyperintense extraaxial lesion (black arrow). T1WI images (b, c) demonstrating iso-signal intensity with strong enhancement (d) and a dural tail sign (white arrow). Angiogram (d) showing tumor staining and supply from the external carotid artery. Pathology revealed a meningotheli-omatous meningioma.

subtypes of tumors containing a cystic component included, 2 chordoid, 1 rhabdoid papillary, and 1 meningotheliomatous meningiomas (Fig. 2). Parenchymal edema was demonstrated by 10 (63%) cases.

Other neuroimaging findings

On CT, variable calcifications (fine, cloud-like, dense, or a homogeneously nodular appearance) were present in 5 (50%) of the 10 tumors examined by CT. On non-contrast CT scans, tumors appeared hyperdense (n=6) or isodense (n=4) versus normal parenchyma.

Of the 8 cases with available angiographs, hypervascular tumor staining was noted in 7 (the remaining case showed no tumor staining), and in these cases, 4 tumors were supplied from the external carotid artery (ECA) (Fig. 1), and 3 from the internal carotid artery (ICA) (Fig. 3).

Preoperative diagnosis

Because of the atypical imaging findings, seven (44%) tumors were diagnosed as non-meningeal tumors preoperatively, i.e., choroid plexus papilloma (n=2), glioblastoma multiforme (n=2) (Fig. 4), ganglioglioma (Fig. 5, n=1), primitive neuroectodermal tumor (PNET, n=1), and brain stem glioma (n=1).

Histological findings

Histological subtypes were classified according to the World Health Organization (WHO) recommendations for the classification of brain tumors (11), and are summarized in Table 2. Meningotheliomatous (n=4) and transitional cell type meningioma (n=4) were most common in the present study. Histological features of the meningotheliomatous type were standard, that is, islands of poorly defined cells with homogeneous eosinophilic cytoplasm. Of the 4 meningotheliomatous tumors, 3 were located in the supratentorial area, and 1

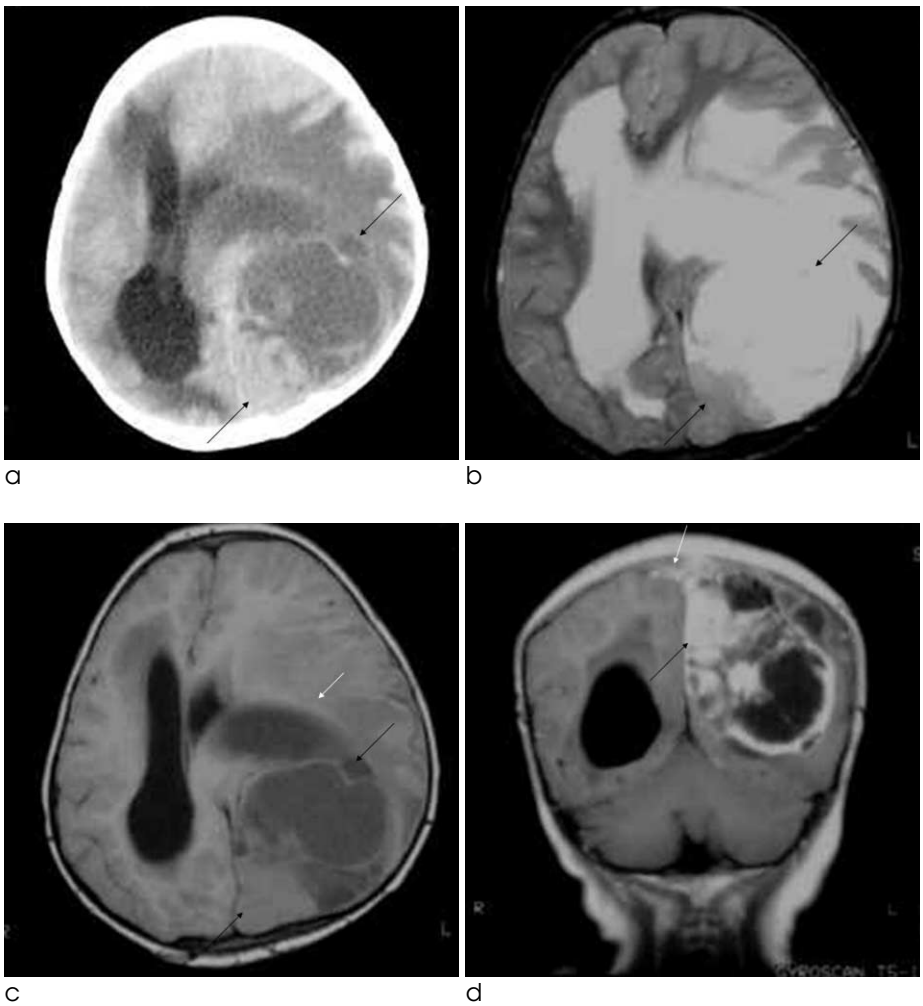

Fig. 2. A 3-year-old boy with right. side weakness.
Non-contrast CT scan (a) showing a solid cystic mass lesion (black arrow) in the Lt. parietal lobe. T2-WI and T1-WI images showing high signal intensity (b) and moderate signal intensity (c) of the solid portion, respectively. The T1-WI image (d) also shows a cystic portion (white arrow). T1-WI image (d) after contrast administration showing intense enhancement (black arrow) of the solid portion and a dural tail sign (white arrow). Pathology revealed a meningotheliomatous meningioma.

around the clivus. The transitional cell type, containing both spindle cells and round-to-oval cells separated by dense intercellular reticulum network, occurred in the frontal convexity (n = 1), temporal region (n = 1), at the CPA (n = 1), within the ventricle (n = 1). Histopathological examination of the other tumors revealed chordoid (n = 2), fibrous (n = 2), clear cell (n = 1), hyalinized (n = 1), rhabdoid papillary (n = 1), and atypical (n = 1) subtypes.

Discussion

Meningiomas of childhood and adolescence are uncommon, and mean age at diagnosis is greater than for other brain tumors of childhood (6, 12). In the present study, the majority of patients were teenagers (mean age 14 years), and boys were more affected than girls, which concurs with previous studies (9, 12), but which contrasts with that found in adults.

Clinical presentations varied and depended on tumor site. The most frequent symptom in our series was a headache (n = 8), followed by seizure (n = 4), a tingling sensation (n = 2), and extremity weakness (n = 1). Headaches are related to an elevated intracranial pressure, and their prevalence in our cohort is consistent with other observations (6, 13). Moreover, although some studies have reported that epilepsy is present in 20-40% at diagnosis (4, 14-16), more recent studies have reported much lower prevalences (12, 13, 17).

According to adult and pediatric studies, convexity and parasagittal locations are most common and are observed in more that 50% of patients (12, 18). However, our findings differ, in that these locations only accounted for 38% of the 16 tumors examined in the present study. On the other hand, other tumors had diverse locations, i.e., intraparenchymal (n = 4),

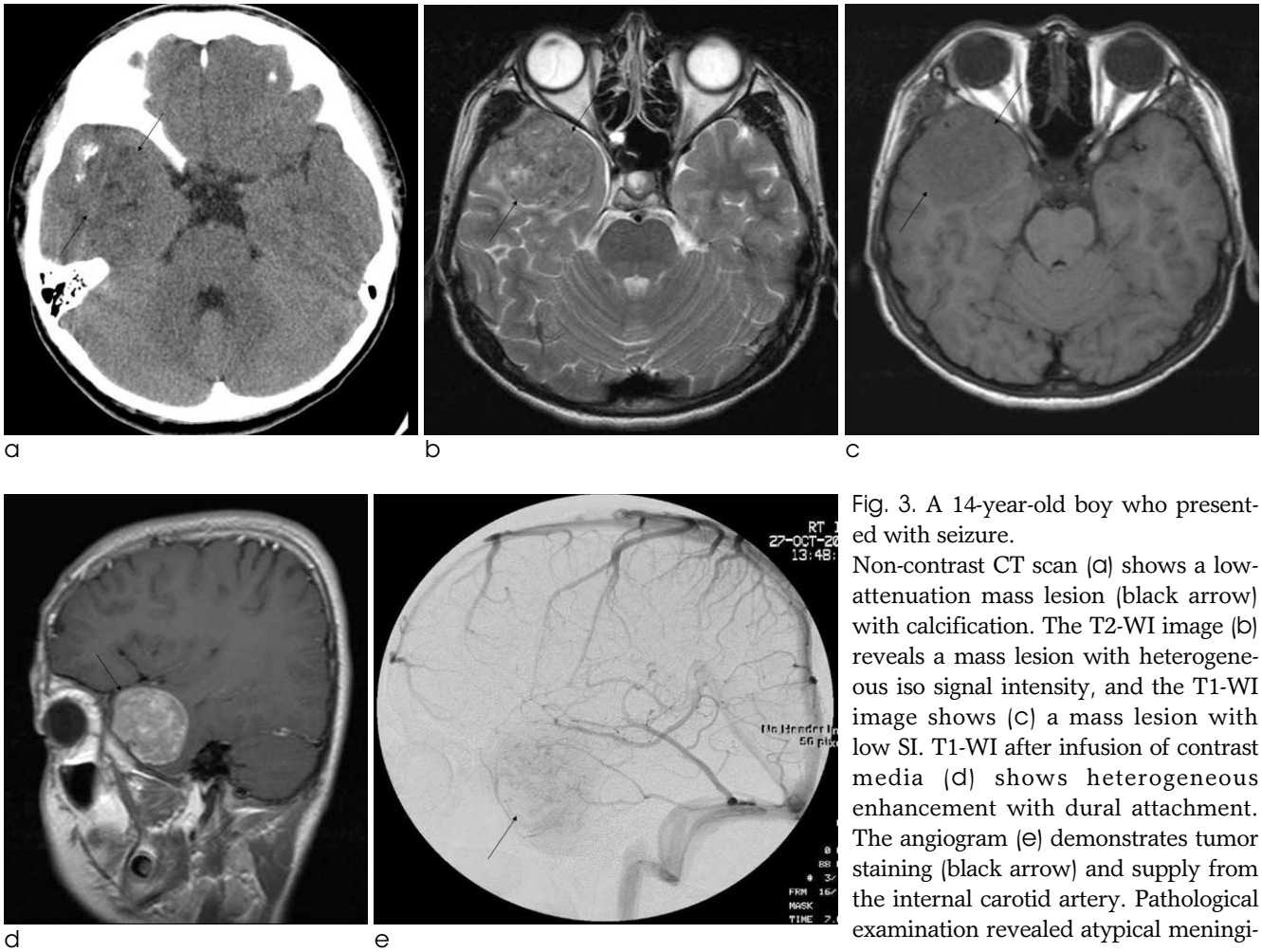

Fig. 3. A 14-year-old boy who presented with seizure.
Non-contrast CT scan (a) shows a low-attenuation mass lesion (black arrow) with calcification. The T2-WI image (b) reveals a mass lesion with heterogeneous iso signal intensity, and the T1-WI image shows (c) a mass lesion with low SI. T1-WI after infusion of contrast media (d) shows heterogeneous enhancement with dural attachment. The angiogram (e) demonstrates tumor staining (black arrow) and supply from the internal carotid artery. Pathological examination revealed atypical meningioma.

intraventricular (n = 3), CPA (n = 2), and clivus (n = 1).

A number of studies on pediatric meningioma have presented classic imaging findings (19), that is, hypo- to isointensity on T1-WI images and hyper- to isointensity versus cortical gray matter on T2-WI images, which are attributed in part to tumor cellularity. In the present study, 15 of the 16 patients displayed similar signal patterns. The remaining patient demonstrated hypointensity by T2-WI and hyperintensity by T1-WI; in this case, the tumor was pathologically proved to be a chordoid meningioma. Dural attachment is not as common as in adult meningiomas (16, 20-23), though it was seen in 69% of our patients, and importantly suggests an extraparenchymal tumor, especially for masses mimicking a parenchymal tumor on MR images. Cystic meningiomas are known to be more frequent in children than in adults (14, 22), which complicates the diagnosis, because meningiomas are usually expected to be solid tumors. In our study, 4 cases showed variably-sized cysts within or adjacent to tumors. Pathologically examinations in these cases revealed chordoid (n = 2), rhabdoid papillary (n = 1), and meningotheliomatous (n = 1) meningiomas.

Although meningiomas are benign, they are often accompanied by symptomatic brain edema (24, 25). Nakano et al. (25) reported that brain edema in 27 of 51 meningiomas was correlated with an irregular tumor margin, the disappearance of a tumoral rim, and T2-WI hyperintensity. In the present study, 10 of 16 meningiomas had associated brain edema.

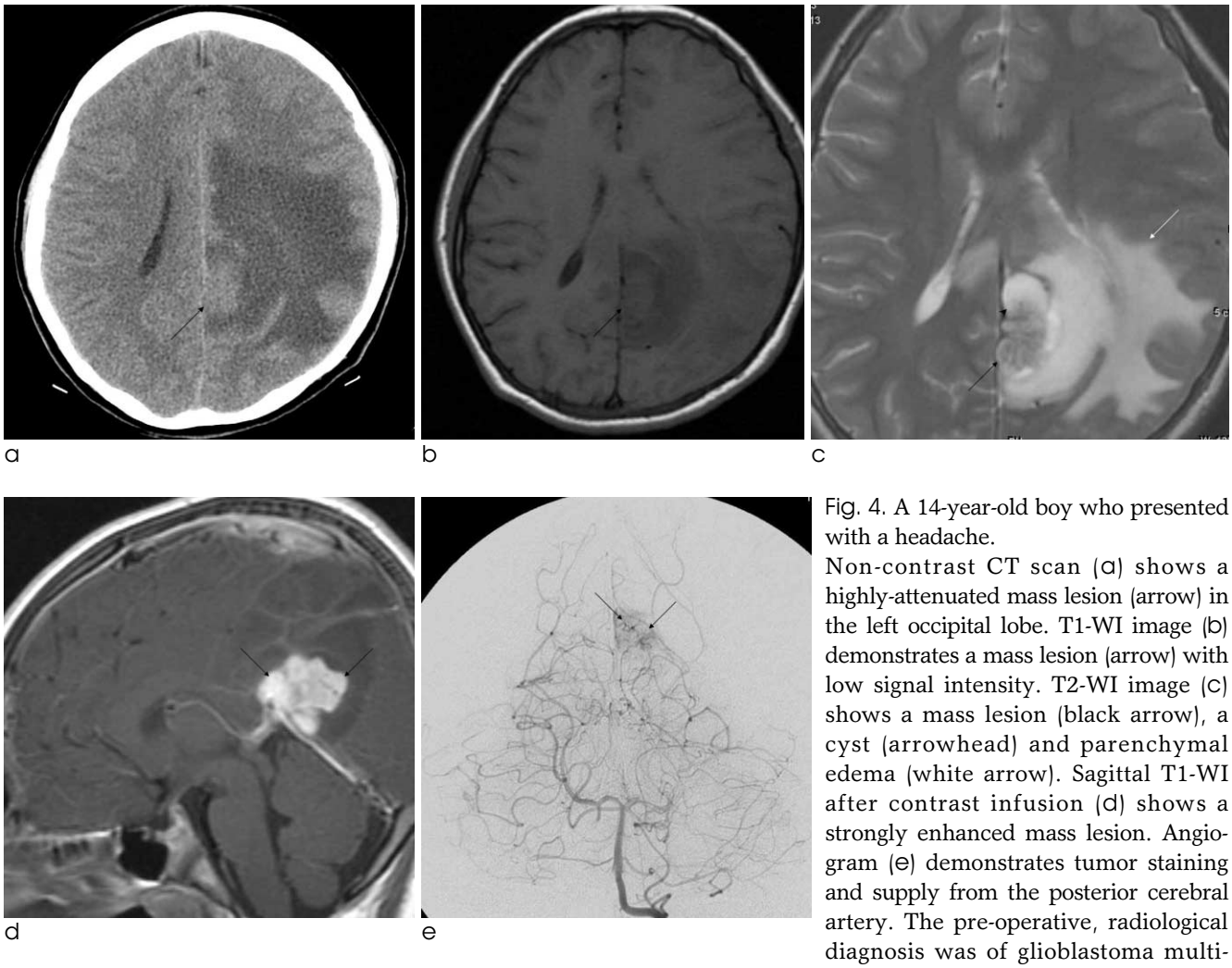

Fig. 4. A 14-year-old boy who presented with a headache.
Non-contrast CT scan (a) shows a highly-attenuated mass lesion (arrow) in the left occipital lobe. T1-WI image (b) demonstrates a mass lesion (arrow) with low signal intensity. T2-WI image (c) shows a mass lesion (black arrow), a cyst (arrowhead) and parenchymal edema (white arrow). Sagittal T1-WI after contrast infusion (d) shows a strongly enhanced mass lesion. Angiogram (e) demonstrates tumor staining and supply from the posterior cerebral artery. The pre-operative, radiological diagnosis was of glioblastoma multiforme but pathological examination subsequently revealed chordoid meningioma.

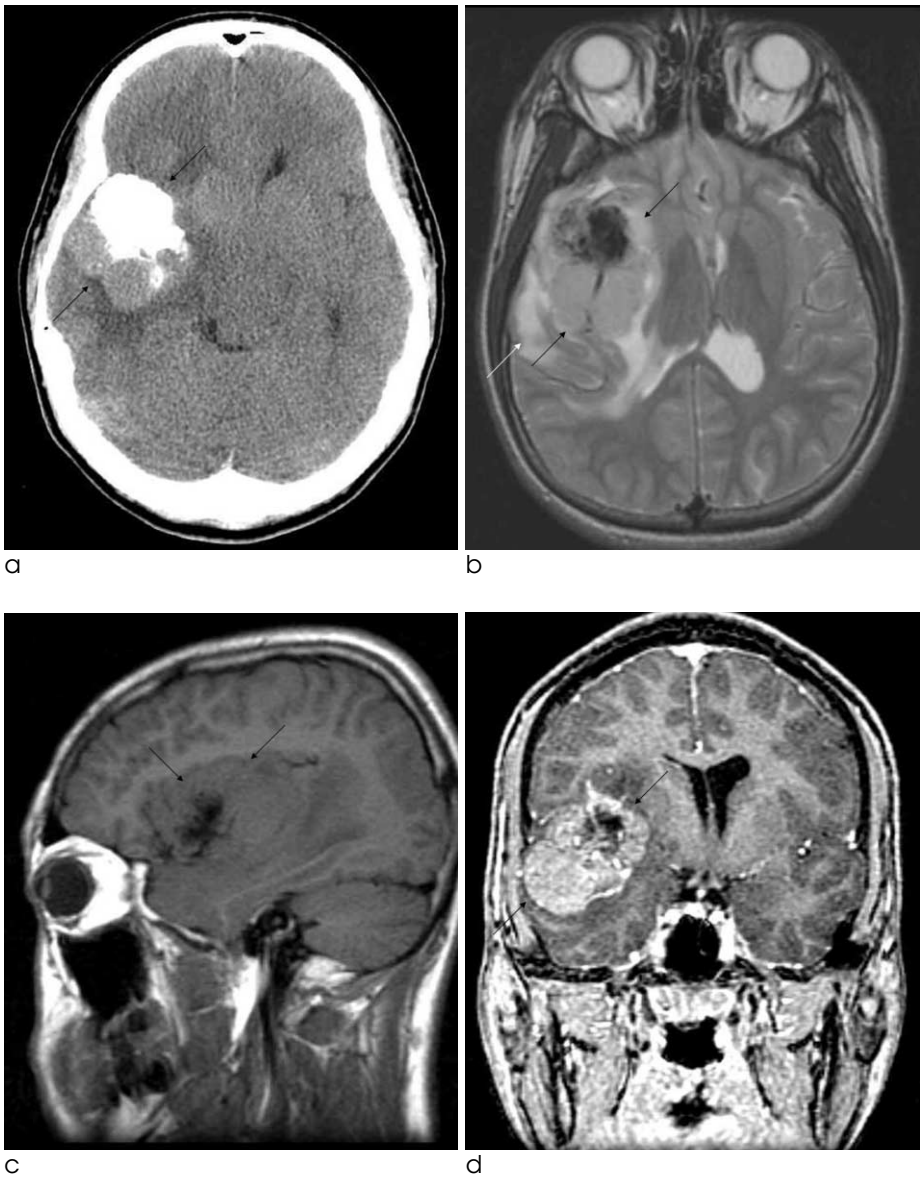

Fig. 5. A 16-year-old boy who presented with a tingling sensation. Non-contrast CT scan (a) shows a highly attenuated mass lesion (arrow) with dense calcification in the right temporal lobe. T2-WI image (b) demonstrates a high signal intensity mass lesion (black arrow) with low signal intensity region (suggestive of calcification), and parenchymal edema (white arrow). T1-WI image (c) shows a low signal intensity mass lesion (arrow) with a low signal intensity region, suggestive of calcification. T1-WI contrast enhanced image (d) shows strong enhancement. The pre-operative radiologic diagnosis was of ganglioglioma, but pathologic examination revealed fibrous meningioma.

Intraparenchymal location and surrounding edema (Fig. 2) are, however, may lead to misdiagnosis of brain parenchymal tumor rather than meningioma.

CT is excellent for demonstrating intratumoral calcifications in meningiomas and mass hyperattenuation is characteristic on unenhanced CT. In the present study, calcification and hyperattenuation on unenhanced CT scans were observed in 50 and 60%, respectively. Although cerebral angiography is not necessary in all cases of pediatric meningioma, it can provide additional information about the tumor base and its vascularity, which can be useful during surgical planning. Typical angiographic findings are a prominent tumor blush and delayed "washout". Meningiomas are usually supplied by meningeal branches of the ECA (26) although, 3 of 8 meningiomas in our study received a blood supply from the ICA (Fig.3, 4), which led to the mistaken diagnosis of an intraaxial tumor in some cases. Furthermore, one tumor showed no blush by cerebral angiography.

Because of these atypical imaging findings and the low incidence of meningioma in children, several cases were regarded as intraparenchymal tumors in this study preoperatively; for example, a right temporal lobe mass without dural attachment was diagnosed as ganglioglioma preoperatively, but a postoperative pathologic investigation subsequently revealed it to be a fibrous meningioma, and pathologically-proven

chordoid meningioma was preoperatively diagnosed as glioblastoma multiforme, because the cystic lesions had a parenchymal location with accompanying extensive brain edema. Furthermore, intraventricular meningiomas were diagnosed preoperatively as ependymoma and choroid plexus papilloma due to an unusual location.

In the present study, histopathological examinations revealed grade I meningioma in 11 cases (69%), grade II in 4 (25%), and grade III in 1 (6%), according to the WHO classification (11). Therefore, 5 of the 16 (31%) were atypical malignant meningiomas, which supports previous reports concerning higher incidences of atypical histopathological features in pediatric meningiomas (10, 27, 28)

We conclude that pediatric meningiomas usually occurs in teenage patients, represent diverse pathological types, and frequently exhibit atypical imaging findings mimicking intra-axial tumors.

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두개강내 소아 수막종 16예의 자기공명영상 소견

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어 홍 · 김지혜

목적: 이 연구의 목적은 소아 수막종의 임상적, 영상의학적, 그리고 병리학적 특징을 기술함에 있다.
대상 및 방법: 병리학적으로 진단된 수막종을 가진 16예의 소아환자를 대상으로 임상기록과 자기공명영상을 후향적으로 분석하였다. 평균연령은 14세 (3-18세)였다. 자기공명영상은 병변의 크기, 신호강도, 경계부의 특징, 내부 구조, 조영증강, 그리고 경막과 뇌실질의 변화를 검토하였고 다른 영상 방법 및 병리소견을 함께 분석하였다.
결과: 종양의 평균 크기는 5.24 cm (1.3-18.1 cm)였으며 12예는 천막상부에 4예는 천막하부에 위치하였다. T2 강조 영상에서 높은 신호강도를 보이는 종양이 9예 있었고 4예에서 동신호강도를, 3예에서 낮은 신호강도를 보였다. T1 강조 영상에서는 낮은 신호강도 11예, 동신호강도 4예, 그리고 높은 신호강도 1예가 있었다. 모든 종양의 경계가 좋았고 조영증강을 보였다. 5예에서 종양은 균질한 양상을 보였으며 나머지에는 낭종이나 괴사에 의해 비균질한 양상을 보였다. 경막 부착 소견이 11예에서, 뇌실질 부종이 10예에서 동반되었다. 컴퓨터 단층촬영 소견은 6예에서 밝은 음영을 보였으며 5예에서 석회화를 동반하였다. 혈관 조영술 상 3예는 내경동맥에서, 4예는 외경동맥으로부터 혈액 공급이 이루어졌다. 병리검사결과 종양의 아형은 이행성 (4예), 수막세포성 (4예), 척삭성 (2예), 섬유성 (2예), 투명세포성 (1예), 유리질화성 (1예), 횡문근양 유두모양 (1예), 그리고 비정형(1예) 수막종으로 진단되었다.
결론: 소아의 수막종은 주로 10대에 발생하며 다양한 병리학적 아형을 보일뿐만 아니라 비정형적인 영상 소견으로 인해 축내 종양으로 오인될 수 있다.

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