Radiology Case Reports 18 (2023) 2401–2406

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Sclerosing meningioma with a large peritumoral cyst: Case report ☆,☆☆

Narushi Sugii, MD, PhD[a,*], *Takao Tsurubuchi, MD, PhD*[a], *Noriaki Sakamoto, MD, PhD*[b], *Makoto Shibuya, MD, PhD*[c], *Eiichi Ishikawa, MD, PhD*[a]

[a] *Department of Neurosurgery, Faculty of Medicine, University of Tsukuba, 1-1-1 Tennodai, Tsukuba, Ibaraki, 305-8575 Japan*

[b] *Department of Diagnostic Pathology, Faculty of Medicine, University of Tsukuba, Tsukuba, Ibaraki, Japan*

[c] *Central Clinical Laboratory, Hachioji Medical Center, Tokyo Medical University, Tokyo, Japan*

ARTICLE INFO

Article history:
Received 12 March 2023
Revised 29 March 2023
Accepted 5 April 2023
Available online 3 May 2023

Keywords:
Meningioma variant
Very low intensity
Low intensity on T2
Hypovascular tumor
Fibrous tumor
Cystic meningioma

ABSTRACT

Meningioma morphology is diverse. Although unlisted in the WHO classification, sclerosing meningioma is a rare variation featuring an extremely low signal intensity on MRI T2-weighted imaging. About 50 cases of sclerosing meningiomas, including spinal tumors, have been reported; however, cases with an accompanying large peritumoral cyst remain unreported. Here, we first report a rare case of sclerosing meningioma with a large peritumoral cyst and review relevant literature.

Abbreviations: CNS, central nervous system; CT, computed tomography; MRI, magnetic resonance imaging; TRAK, time-resolved angiography using keyhole; WHO, World Health Organization; WI, weighted image.

☆ Acknowledgments: We would like to thank Dr Mika Watanabe (Department of Pathology, Tohoku University Hospital, Miyagi, Japan) and Dr Satoko Nakada (Department of Diagnostic Pathology, Kanazawa University Hospital, Ishikawa, Japan) for pathological opinions. We thank Dr Bryan J. Mathis (International Medical Center, University of Tsukuba Hospital) for language revision. The corresponding author (N.S.) also would like to extend his indebtedness to Yuka Sugii for her endless love, understanding, support, encouragement, and sacrifice throughout his work.

☆☆ Competing Interests: The authors have declared that no competing interests exist.

* Corresponding author.
E-mail address: narushi-sugii@md.tsukuba.ac.jp (N. Sugii).

https://doi.org/10.1016/j.radcr.2023.04.012

Introduction

Meningiomas demonstrate remarkable morphological variety and current WHO classification guidelines for central nervous system (CNS) tumors (updated in 2021) divide them into various subtypes [1]. Although unlisted in these classification schema, other subtypes of meningioma that show different histopathological patterns, such as oncocytic [2], mucinous [3], and sclerosing meningiomas, have been reported [4].

Sclerosing meningioma is one such rare variation, comprising 2.5% of total cases [5]. Although not fully delineated because of rarity, about 50 cases, including spinal tumors, have been reported [4–11]. Ordinary meningioma subtypes, established and listed in the WHO classification, sometimes accompany peritumoral cysts [12]; however, no case of sclerosing meningioma accompanying a sizeable peritumoral cyst has yet been published.

Here, we first report a case of a benign fibrous tumor with a large peritumoral cyst that was eventually diagnosed as a sclerosing meningioma.

Case presentation

A 53-year-old man with no previous medical history presented to a clinic with a headache and was diagnosed with a brain tumor in the right parietal region. He was conscious and clear, with no apparent paralysis or sensory disturbance, but with narrowing of the left lower visual field quadrant. The maximum diameter of the tumor was 5 cm, with overall high density (without calcification) on computed tomography (CT), iso intensity on magnetic resonance imaging (MRI) T1-weighted imaging (WI), uniformly very low intensity on T2WI, and high contrast on gadolinium-based contrast-enhanced MRI (Fig. 1). The tumor also showed very low intensity on diffusion WI after T2 blackout effect. The tumor was separate from the ventricles, accompanied by a large cyst anteriorly, and the signal intensity of cystic contents differed from cerebrospinal fluid. As the cyst wall was uncontrasted, the cyst was deemed a peritumoral cyst around which edematous changes were observed. The tumor was in contact with the dura mater convexity and the cerebral falx, but there were no findings suggestive of an obvious attachment nor was there a dural tail sign. Furthermore, 4D time-resolved angiography using keyhole (4D-TRAK) MRI did not show tumor staining or feeding vessels, suggesting low vascularity. Blood data was unremarkable, including tumor markers, and contrast-enhanced CT of the trunk showed no mass lesions that could be the primary tumor. Although narrowing the preoperative diagnosis was difficult, we decided on a policy of excision for pathological diagnosis and therapeutic purposes for a symptomatic tumor with a significant mass effect.

The tumor was very hard and fibrous, with almost no hemorrhaging or noticeable feeding vessels during the incision (Figs. 2A and B). The tumor was not adherent to the surrounding brain and was easily detached, suggesting an extraparenchymal tumor. Although the tumor was in contact with the falx cerebri, it detached easily and this minimal attachment allowed for total removal of the tumor and attached falx (Simpson grade I) (Figs. 2C and D). Postoperative MRIs revealed a diminished cyst, amelioration of the edema, and no residual lesions (Fig. 3).

Histologically, there was a monotonous proliferation of sparse bundles of spindle-shaped cells with thick eosinophilic collagenous interstitium, accompanied by degenerated vessel walls and areas of low cell density with some edematous degeneration (Fig. 4). The spindle-shaped cells lacked nuclear atypia and there were no mitotic figures or necrosis. Although there was a minimal meningioma component, elongated spindle cells had proliferated within the abundant collagenous stroma and whorl formations were seen in these; this area was focally weakly positive for EMA. Other immunohistochemical findings were as follows: vimentin (+), progesterone receptor (nuclear-, cytoplasmic+), CD68 (-), CD34 (-), STAT-6 (-), desmin (-), αSMA (-), Actin/HHF (-), S-100 (-), GFAP (-), Olig2 (-), NeuN (-), c-kit (-), MDM2 (-), pankeratin (-), MIC2/CD99 (-), β catenin nuclear transfer (-), p53 (-), and Ki-67 (<1%). No lesions were found in the dura (resected falx cerebri). From these data, plus the aid of several outside experts in pathology, we arrived at a diagnosis of sclerosing meningioma and treated the tumor as equivalent to WHO grade 1.

The postoperative course was good, with no additional treatment required, and the patient remains free from recurrence at 2 years of follow-up. We obtained written, informed consent concerning publication and handled clinical information anonymously in accordance with the principles of the Declaration of Helsinki and the “Act on the Protection of Personal Information” in Japan.

Discussion

We experienced a rare case of sclerosing meningioma with a peritumoral cyst that was initially difficult to diagnose. Regarding preoperative diagnosis, the imaging findings of unusually low intensity on T2WI, a large peritumoral cyst, and no dural tail sign made narrowing the diagnosis challenging. As for pathology, a lack of viable tumor components positively suggestive of meningioma and fragile immunostaining further complicated classification efforts. Nevertheless, outside experts in brain tumor pathology, plus crucial clues of whorl formations and weak EMA positivity, facilitated the final diagnosis.

Sclerosing meningioma is a rare variation; according to the previous report, 21 out of 851 meningioma cases were histopathologically proven as sclerosing meningioma, with a calculated rate of about 2.5% of all meningiomas [5]. Its crucial pathological features are a scarcity or absence of viable tumor components in the meningothelial cells of massively sclerotic lesions and that pauci-cellular collagenous tissue with spindle cells show immunohistochemical staining patterns that differ from conventional meningothelial cells [7]. In the present case, finding a tiny fraction of viable meningioma cells from massively degenerated tumor components was challenging and immunohistochemical findings were not indicative of fibrous histiocytoma, solitary fibrous tumor/hemangiopericytoma, myogenic tumor, or conventional

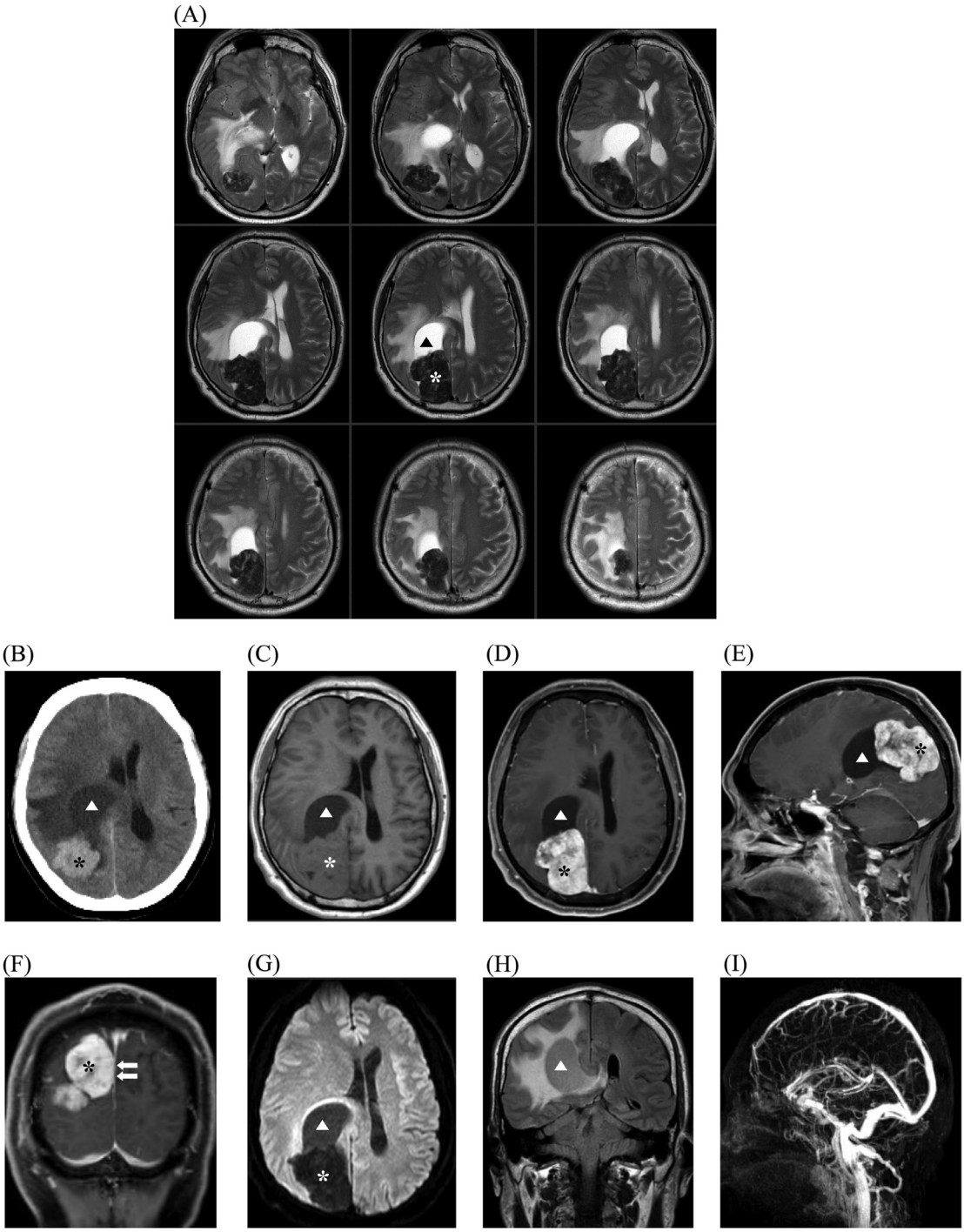

Fig. 1 – Preoperative CT and MRI. Preoperative images are shown. The tumor (*), demonstrating markedly low signal intensity on MRI T2WI and accompanied by a peritumoral cyst (arrowheads) anteriorly with marked edema (A). CT, revealing a high-density lesion without calcification (B) and iso intensity on MRI T1WI (C) with good enhancement by gadolinium-based contrast-medium in axial (D), sagittal (E), and coronal sections (F). The tumor, touching the falx cerebri, but a dural tail sign is absent (arrows). The lesion, showing low intensity on DWI (G), and cystic contents with signal intensity differing from cerebrospinal fluid on FLAIR coronal section (H). The SSS is patent; no tumor staining or feeding vessels are visible on 4D-TRAK MRI (I).

Abbreviations: CT: computed tomography, DWI: diffusion weighted image: FLAIR: fluid-attenuated inversion recovery; MRI: magnetic resonance imaging; SSS: superior sagittal sinus; TRAK: time-resolved angiography using keyhole; WI: weighted image.

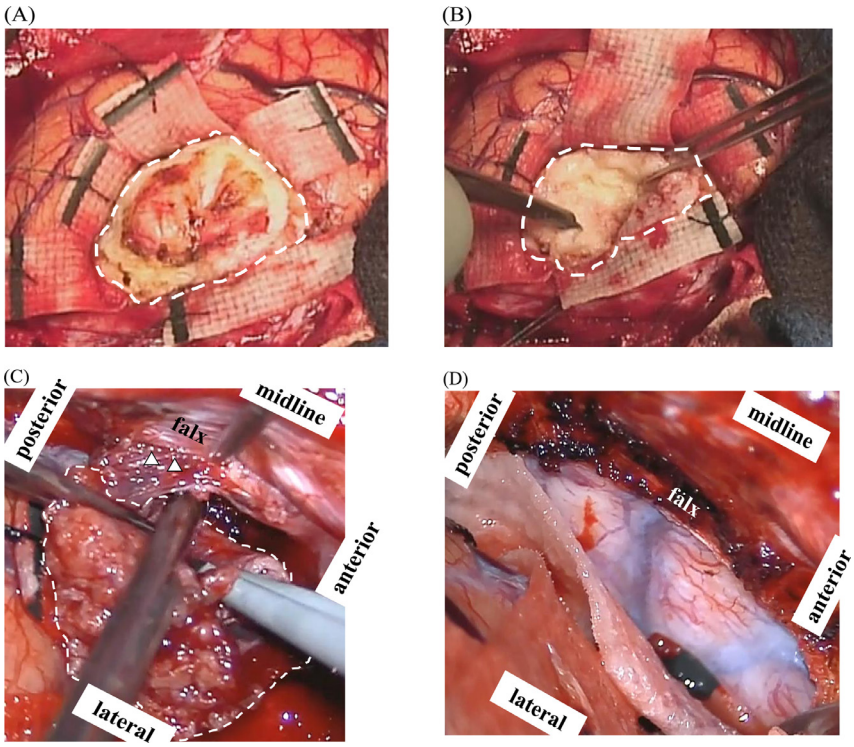

Fig 2 – Intraoperative findings. The tumor (areas enclosed by the dotted line), extremely solid and with clear boundaries (A). The tumor, with almost no blood flow (B) and a minimal attachment to the falx cerebri (C, arrowheads), was removed, including the attached falx (D).

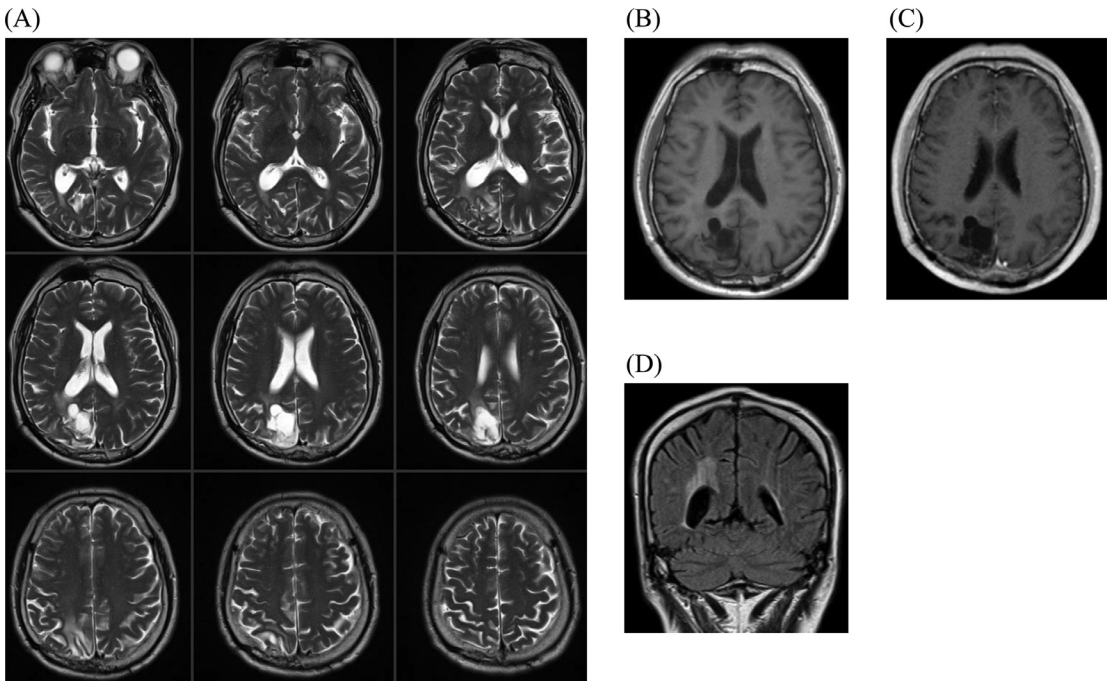

Fig. 3 – Postoperative MRI. Resolved tumor, completely excised, and cyst, which disappeared with minimal edematous changes, on T2WI (A) pre-(B) and post-(C) enhanced T1WI, and FLAIR coronal section (D).
Abbreviations: FLAIR: fluid-attenuated inversion recovery; MRI: magnetic resonance imaging; WI: weighted image.

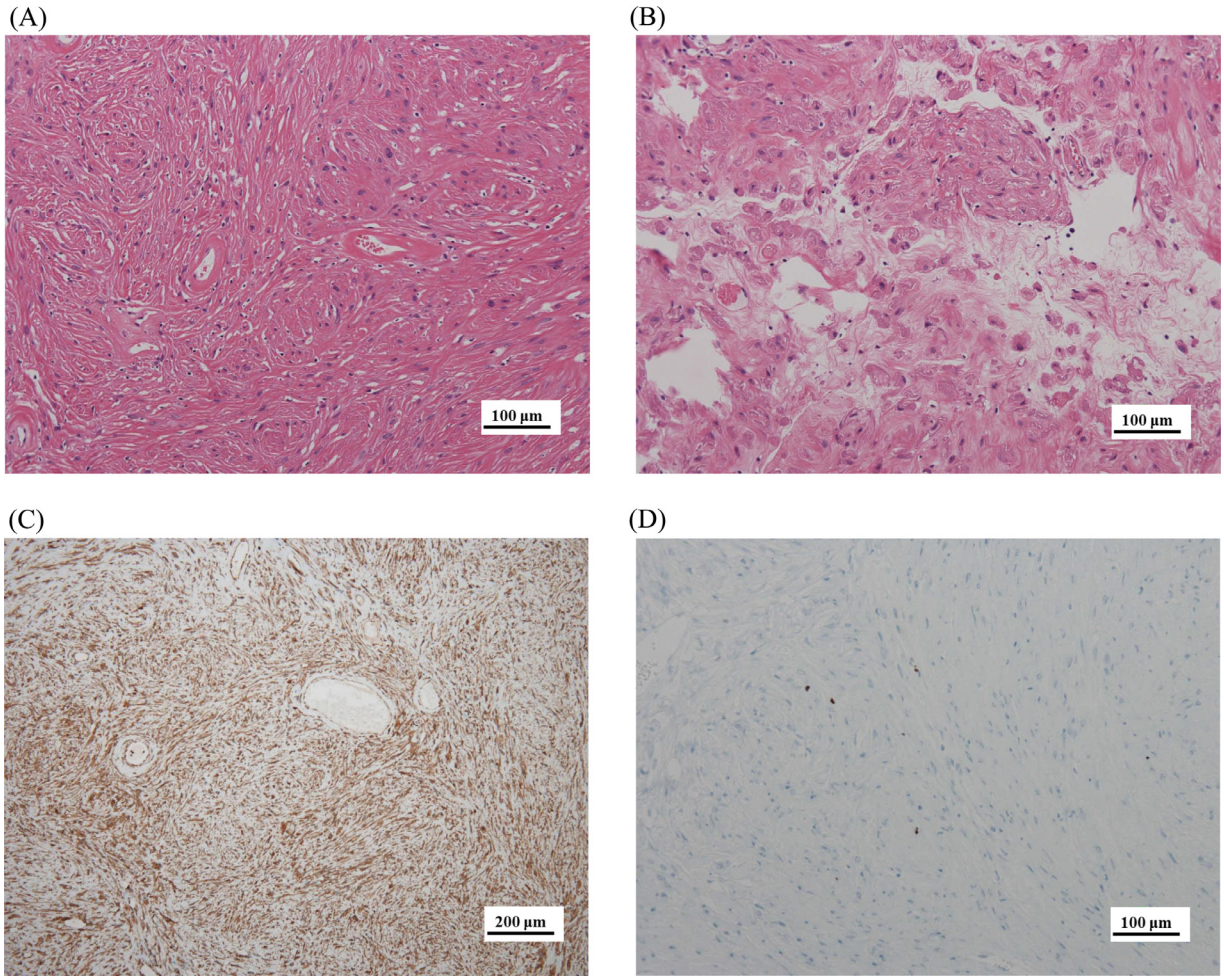

Fig. 4 – Pathological findings. H&E staining, revealing a monotonous proliferation of spindle-shaped cells with thick eosinophilic collagenous interstitium (A) and low cell density areas with edematous degeneration (B). The tumor, positive for vimentin (C) with an MIB-1 labeling index (Ki-67) of less than 1% (D).
Abbreviation: H&E: hematoxylin and eosin.

meningiomas. However, we finally found a minimal meningioma component that stained strongly positive for vimentin, focally weakly positive for EMA, and negative for GFAP, CD34, progesterone receptor, and S-100; those findings were compatible with reported sclerosing meningioma. Thus, even if most of the tumor component is degenerated or non-viable, there may still be characteristic findings suggestive of meningioma and, thus, persevering in diagnosis is essential.

Typical sclerosing meningiomas show extremely low signal intensities on MRI T2WI and have intratumoral calcifications [5]. Although calcification was absent, an MRI revealed unusually low intensity on T2WI in the present case. Generally, meningiomas with hypointensity on T2WI tend to be hard [13] and, since sclerosing meningioma has a more prominent fraction of collagen bundles and psammomatous calcification than other subtypes [5], very low signal intensity is expected. Indeed, intraoperative findings showed that the tumor was very firm and mostly fibrous, with pathology revealing a high degree of collagen fiber hyperplasia. In addition, a lack of clear dural attachment and dural tail signs, plus the absence of apparent feeding arteries on 4D-TRAK, were atypical for conventional meningiomas. In this regard, the imaging findings were consistent with sclerosing meningioma. We excised the cerebral falx because of a putative attachment but no tumor was found pathologically, implying that the tumor may have originated from elsewhere. There are no previous reports concerning the dural attachment or the origin of sclerosing meningioma cells except for a report of a sclerosing meningioma in the deep sylvian fissure [6]. They speculated that the meningioma arose from the arachnoid membrane or the Virchow-Robin space, but the exact origin of the tumor was unclear. While recent molecular profiles of meningiomas have revealed that differing molecular backgrounds lead to different clinical courses [14,15], no molecular information on sclerosing meningiomas, including our case, are reported. Although rare, further accumulation of cases with molecular evaluation is warranted to explore tumor origins.

In comparison to previous reports, this case had the unique presence of a sizeable peritumoral cyst. Although the precise mechanism remains unclear, brain edema proceeds peritumoral cyst formation [16], taking approximately 36 months from edema onset to form cysts [17]. Sclerosing menin-

giomas exhibit more severe peritumoral edema than conventional meningiomas [5], and their slow-growing nature allows enough time to produce cysts. Based on these hypotheses, it would be unsurprising to see cyst formation in sclerosing meningiomas, yet there are no reports of sclerosing meningioma cases with prominent peritumoral cysts. Overall, approximately 10% of CNS tumors, including benign, malignant, and metastatic, have accompanying cysts [16], and the incidence of meningiomas with cyst(s) (cystic meningiomas) is reported to be 3.5%-9.6% among all meningiomas [12,18–20]. These numbers include intra-tumoral degenerative cyst formation and the proportion of peritumoral cysts similar to the present case accounted for 9.3%-19.5% of cystic meningiomas [12,18,19]. Thus, meningiomas with apparent peritumoral cysts are rather rare, with a calculated rate of about 1% of all meningiomas, a rarity that could simply explain the lack of cyst-associated sclerosing meningioma reports [4–11]. Regardless, the presence of a peritumoral cyst is no reason to exclude sclerosing meningioma from differential diagnosis.

We experienced a rare benign fibrous tumor with a large peritumoral cyst, which eventually led to the diagnosis of sclerosing meningioma. Sclerosing meningiomas are an important differential diagnosis when the lesion shows very low signal intensity on T2WI and is accompanied by a peritumoral cyst.

Patient consent

We obtained written, informed consent concerning publication and handled clinical information anonymously in accordance with the principles of the Declaration of Helsinki and the "Act on the Protection of Personal Information" in Japan.

REFERENCES

[1] Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D, et al. The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. Neuro Oncol 2021;23:1231–51. doi:10.1093/neuonc/noab106.

[2] Gallina P, Buccoliero AM, Mariotti F, Mennonna P, Di Lorenzo N. Oncocytic meningiomas: cases with benign histopathological features and a favorable clinical course. J Neurosurg 2006;105:736–8. doi:10.3171/jns.2006.105.5.736.

[3] Berho M, Suster S. Mucinous meningioma. Report of an unusual variant of meningioma that may mimic metastatic mucin-producing carcinoma. Am J Surg Pathol 1994;18:100–6.

[4] Haberler C, Jarius C, Lang S, Rössler K, Gruber A, Hainfellner JA, et al. Fibrous meningeal tumors with extensive non-calcifying collagenous whorls and glial fibrillary acidic protein expression: the whorling-sclerosing variant of meningioma. Neuropathol Appl Neurobiol 2002;28:42–7. doi:10.1046/j.0305-1846.2001.00364.x.

[5] Kang H, Kim JW, Se YB, Dho YS, Choi SH, Park SH. Sclerosing Meningioma : radiological and clinical characteristics of 21 cases. J Korean Neurosurg Soc 2016;59:584–9. doi:10.3340/jkns.2016.59.6.584.

[6] Fukushima S, Narita Y, Yonezawa M, Ohno M, Arita H, Miyakita Y, et al. Short communication: sclerosing meningioma in the deep sylvian fissure. Brain Tumor Pathol 2014;31:289–92. doi:10.1007/s10014-013-0167-8.

[7] Kim NR, Im SH, Chung CK, Suh YL, Choe G, Chi JG. Sclerosing meningioma: immunohistochemical analysis of five cases. Neuropathol Appl Neurobiol 2004;30:126–35. doi:10.1046/j.0305-1846.2003.00517.x.

[8] Im SH, Chung CK, Cho BK, Kim MK, Chi JG. Sclerosing meningioma: clinicopathological study of four cases. J Neurooncol 2004;68:169–75. doi:10.1023/b:neon.0000027759.54516.7c.

[9] Elmaci İ, Altinoz MA, Sav A, Bolükbaşı FH, Önöz M, Başkan Ö, et al. Whorling-sclerosing meningioma. A review on the histological features of a rare tumor including an illustrative case. Clin Neurol Neurosurg 2017;162:85–90. doi:10.1016/j.clineuro.2017.09.009.

[10] Cömert D, Tosuner Z, Hatiboglu MA. A rare case of sclerosing meningioma with immunohistochemical features. Br J Neurosurg 2019:1–2. doi:10.1080/02688697.2018.1556780.

[11] Alabbas Z, Tamer Y, Jneidi M, Issa R. Whorling-sclerosing meningioma invading skull bone and subcutaneous tissue with an incidental toxoplasmosis: a case report. Neuropathology 2022 [e-pub ahead of print] First published: 15 November 2022. doi:10.1111/neup.12882.

[12] Boukobza M, Cebula H, Pop R, Kouakou F, Sadoun A, Coca HA, et al. Cystic meningioma: radiological, histological, and surgical particularities in 43 patients. Acta Neurochir (Wien) 2016;158:1955–64. doi:10.1007/s00701-016-2898-x.

[13] Yao A, Pain M, Balchandani P, Shrivastava RK. Can MRI predict meningioma consistency?: A correlation with tumor pathology and systematic review. Neurosurg Rev 2018;41:745–53. doi:10.1007/s10143-016-0801-0.

[14] Nassiri F, Liu J, Patil V, Mamatjan Y, Wang JZ, Hugh-White R, et al. A clinically applicable integrative molecular classification of meningiomas. Nature 2021;597:119–25. doi:10.1038/s41586-021-03850-3.

[15] Maas SLN, Stichel D, Hielscher T, Sievers P, Berghoff AS, Schrimpf D, et al. Integrated molecular-morphologic meningioma classification: a multicenter retrospective analysis, retrospectively and prospectively validated. J Clin Oncol 2021;39:3839–52. doi:10.1200/jco.21.00784.

[16] Baggenstos MA, Butman JA, Oldfield EH, Lonser RR. Role of edema in peritumoral cyst formation. Neurosurg Focus 2007;22:E9. doi:10.3171/foc.2007.22.5.10.

[17] Lonser RR, Vortmeyer AO, Butman JA, Glasker S, Finn MA, Ammerman JM, et al. Edema is a precursor to central nervous system peritumoral cyst formation. Ann Neurol 2005;58:392–9. doi:10.1002/ana.20584.

[18] Go KO, Lee K, Heo W, Lee YS, Park YS, Kim SK, et al. Cystic Meningiomas: correlation between radiologic and histopathologic features. Brain Tumor Res Treat 2018;6:13–21. doi:10.14791/btrt.2018.6.e3.

[19] Jung TY, Jung S, Shin SR, Moon KS, Kim IY, Park SJ, et al. Clinical and histopathological analysis of cystic meningiomas. J Clin Neurosci 2005;12:651–5. doi:10.1016/j.jocn.2004.09.020.

[20] Liu M, Liu Y, Li X, Zhu S, Wu C. Cystic meninigioma. J Clin Neurosci 2007;14:856–9. doi:10.1016/j.jocn.2006.06.003.