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Case Report

Atypical meningeal hemangiopericytoma presenting with punched-out calvarial erosion ☆,☆☆

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ABSTRACT

Meningeal hemangiopericytoma (HPC) is an infrequent but distinct entity affecting the craniospinal axis. A previously healthy 48-year-old man sustained a gradually progressing motor weakness in the left lower extremity. CT showed a hyperdense mass in the right frontal lobe. On MRI, it was 29 × 30 × 36 mm in dimension, appeared isointense on T1 and hyperintense on T2, and was intensely enhanced with erosive changes in the inner table adjacent to the tumor. The patient underwent tumor resection. Reflection of the bone flap revealed a punched-out erosion in the inner table with a defect of the dura over the upper part of the tumor. Microscopic findings were consistent with grade III HPC with dural invasion. A punched-out calvarial erosion and dural defect caused by an extra-axial tumor may be a high-grade HPC that requires extensive surgical resection.

Introduction

Meningeal hemangiopericytoma (HPC) is an infrequent but distinct entity affecting the craniospinal axis. HPC occasionally presents masquerading a benign meningioma, but patients with HPC have high rate of recurrence and distant metastasis. Total tumor resection at initial surgery is associated with a lower risk of relapse but not prolonged survival. The efficacy of radiation therapy for HPC remains elusive. Meningeal solitary fibrous tumors and HPCs have been combined into a single classification in 2016 [1,2]. In general, HPCs are multilobulated extra-axial tumors with narrow-based dural attachments. Bone erosion by HPCs are documented to be 44%-59%, while hyperostosis or intratumoral calcification are commonly not present [3,4]. Primary calvarial erosion is an infrequent manifestation in association with brain tumor, and if present, can be caused by oligodendroglioma [5–7], astro-

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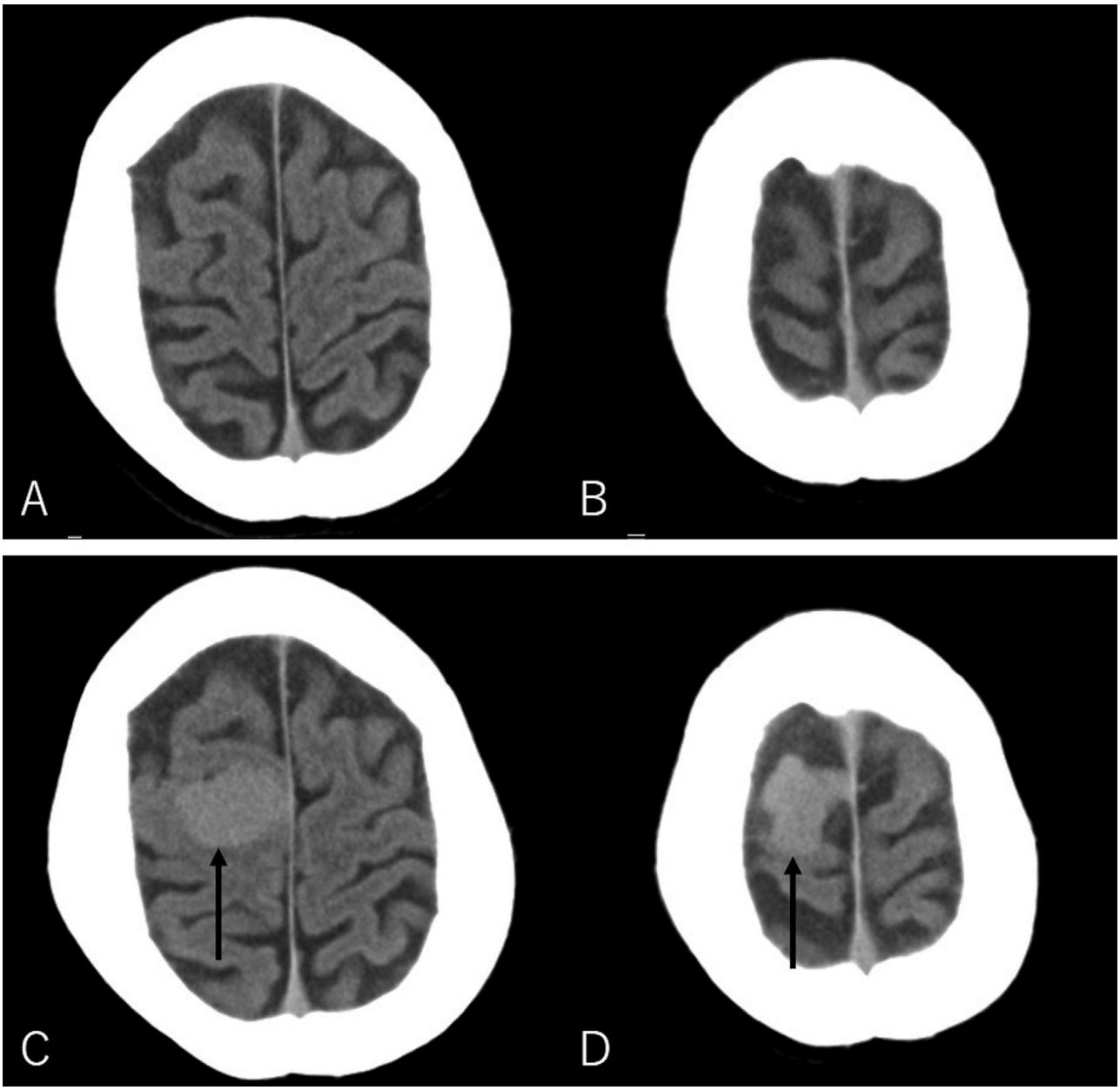

Fig. 1 – Noncontrast axial CT taken in June 2014 (A, B) and June 2020 (C, D) showing emergence of a hyperdense mass in the right frontal lobe (C and D, arrow).

cytoma [8–10], astroblastoma [11,12], xanthoastrocytoma [13], and glioblastoma [14]. However, to our knowledge, no study has documented brain HPC-associated calvarial erosions in detail. Here, we document the neuroimaging, intraoperative, and pathological findings of an atypical case of HPC presenting with a punched-out calvarial erosion and dural defect.

Case presentation

A 48-year-old previously healthy man sustained a progressive motor weakness in the left lower extremity for 3 months. At presentation, the patient did not show any neurological deficits other than the weakness, evaluated as grade 4 on the manual muscle test. Cranial CT showed a hyperdense mass in the right frontal lobe, which had not been noted on that performed in 2014 (6 years back) when the patient presented with sensory disturbances in the lower extremities (Fig. 1). MRI indicated a well-demarcated, extra-axial tumor. It was 29 × 30 × 36 mm in dimension, appeared isointense on T1 and hyperintense on T2, and was intensely enhanced after the infusion of a contrast agent. Furthermore, the tumor protruded superior with erosive changes in the inner table, adjacent to the tumor. A dural tail sign was not observed (Fig. 2). On catheter angiography, the tumor was mainly fed by the right

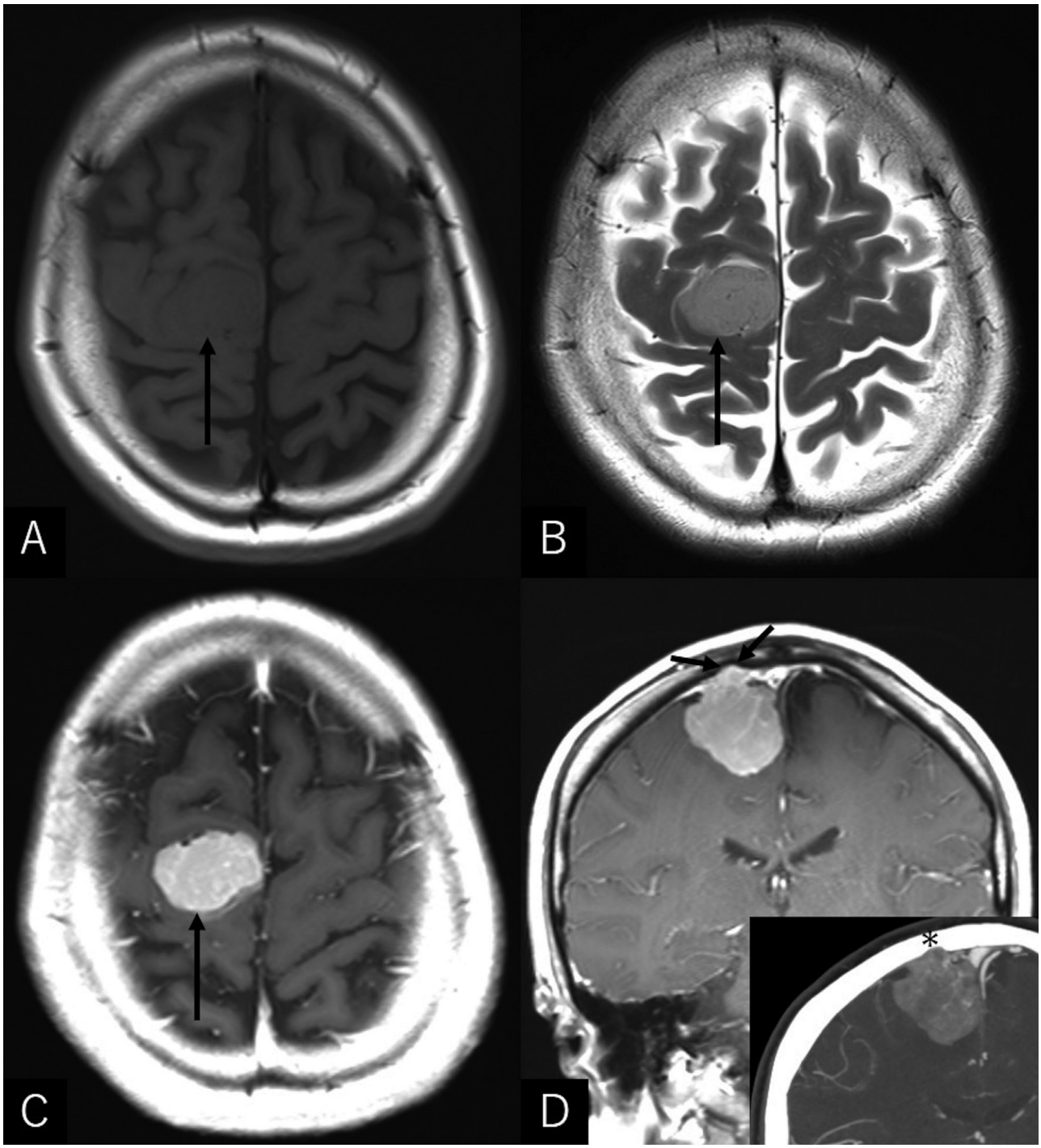

Fig. 2 – Axial T1- (A), T2- (B), and postcontrast axial (C) and coronal (D) T1- weighted MRI showing a well-demarcated, extra-axial tumor, 29 x 30 x 36 mm in dimension, appearing isointense on T1 and hyperintense on T2 (A and B, arrow). The tumor is intensely enhanced after infusion of contrast agent and protrudes superior, while lacking a dural tail sign (C and D, arrows). Inset in (D) shows erosive change in the inner table adjacent to the tumor (asterisk).

middle meningeal artery. A peripheral feeding vessel from the artery was displaced superior with the upper part of the tumor (Fig. 3). Preceding the surgical intervention, the patient underwent embolization of the feeding vessels using 33% n-butyl-2-cyanoacrylate (Fig. 4). The patient then underwent microsurgical tumor resection. Reflection of a frontoparietal bone flap revealed a punched-out erosion in the dura mater and inner table, adjacent to the upper part of the tumor. It was round, 7 mm in dimension, and showed a smooth contour. Hyperostotic changes were not observed (Fig. 5A).

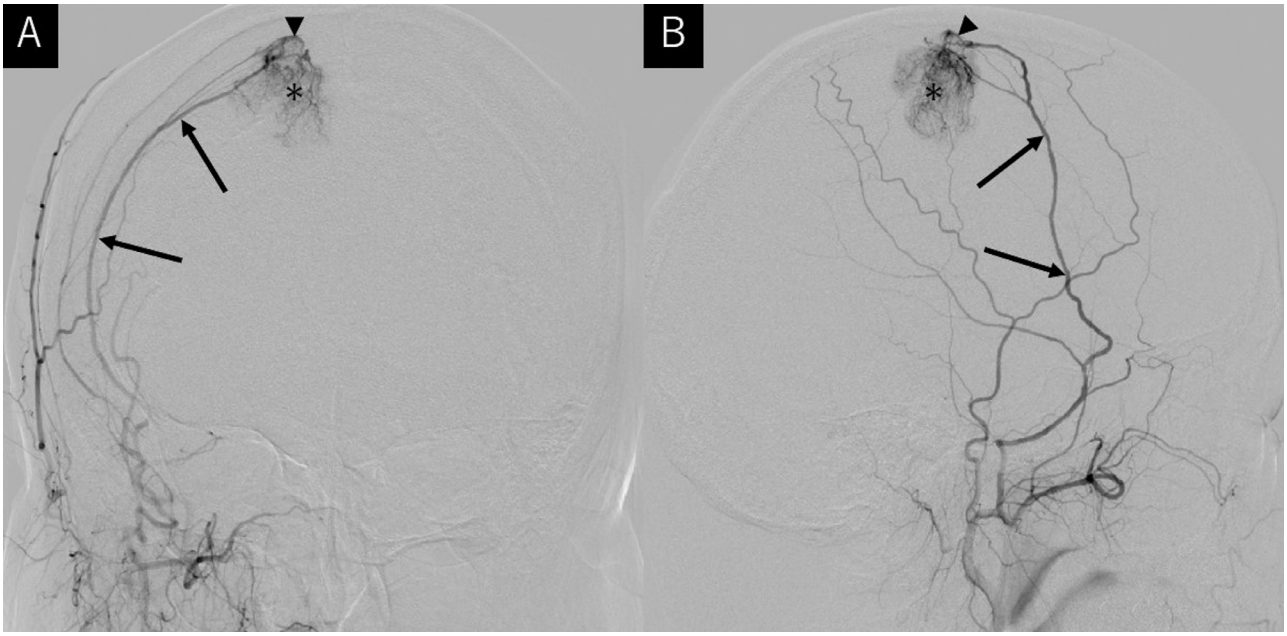

Fig. 3 – (A, B) Right external carotid angiography, anteroposterior (A) and lateral (B) views, showing the tumor stain (A and B, asterisk) supplied by the middle meningeal artery (A and B, arrows). Note that a peripheral feeding vessel from the middle meningeal artery is displaced superior with the upper part of the tumor (A and B, arrowhead).

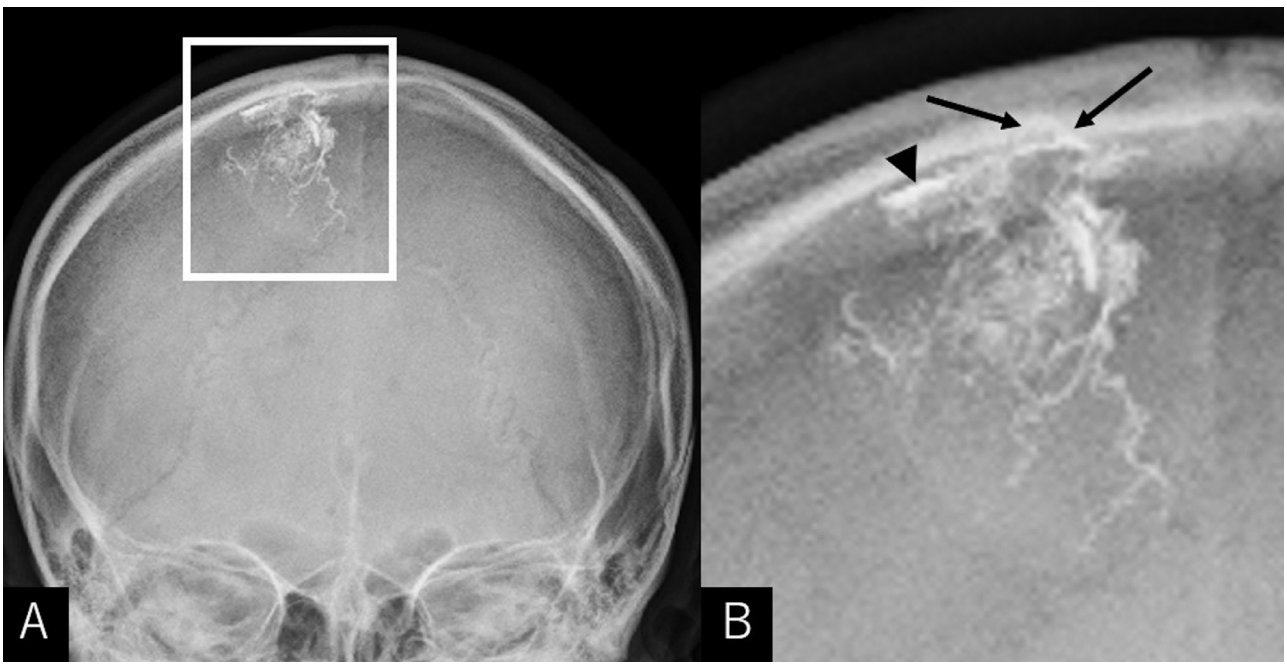

Fig. 4 – (A, B) Cranial radiography after embolization, anteroposterior view (A), and magnified view of the boxed area (B) showing an embolized segment of the middle meningeal artery (B, arrowhead) and tumor vessels. Arrows in (B) indicate the upper part of the tumor protruding into the skull.

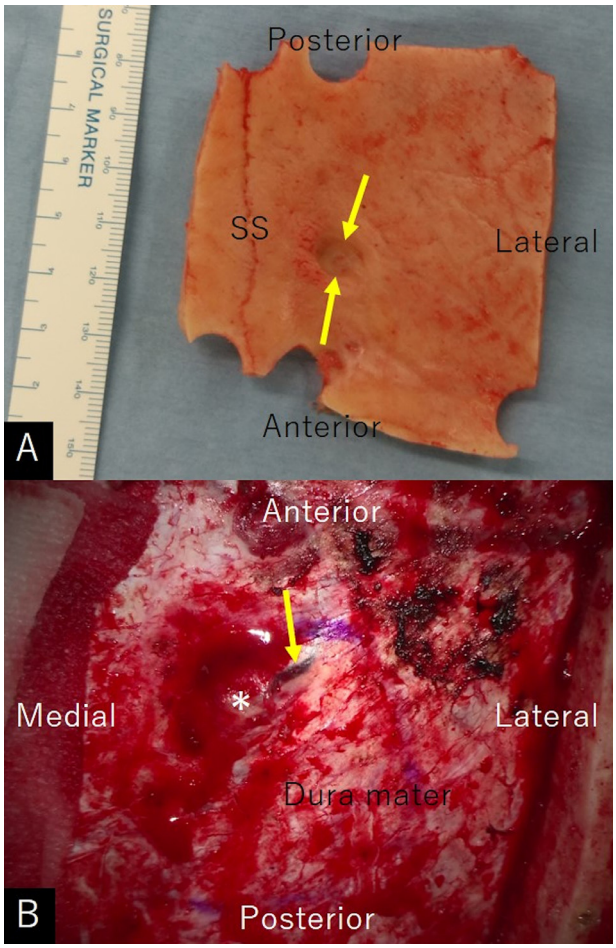

Fig. 5 – (A) Intraoperative photos showing a punched-out bone erosion with round and smooth contour, 7 mm in dimension, in the inner table of the bone flap (arrows). Hyperostotic change is not noted. (B) An embolized middle meningeal artery segment (arrow) connecting to the exposed tumor (asterisk) passes through the margin of the dural defect over the upper part of the tumor. SS: sagittal suture.

An embolized middle meningeal artery segment connecting to the exposed tumor was identified as passing through the margin of the dural defect (Fig. 5B). The tumor was elastic and hard in consistency and vascular. With the internal debulking maneuver using a cavitron ultrasonic surgical aspirator (SONOPET), total resection was eventually achieved. Part of the dura surrounding the defect was extensively resected. On microscopical examination, the tumor comprised spindle-shaped cells with branching vascular structures and cell atypia (Fig. 6A). Immunohistochemical staining was positive for CD34 and STAT6 (Figs. 6B and C), but negative for glial fibrillary acidic protein and epithelial membrane antigen. The MIB-1 index was up to 60%. These were consistent with a World Health Organization grade III HPC. Furthermore, tumor invasion into the dura was identified (Fig. 6D). Postoperatively, the patient's motor weakness improved. Systemic ^{18}F-fluorodeoxyglucose positron emission tomography/CT scans did not reveal any abnormal accumulation. Immediate adjuvant radiotherapy was not administered.

Discussion

The 2007 World Health Organization classification of tumors of the central nervous system distinguished HRCs from solitary fibrous tumor. However, it has recently been demonstrated that both tumors share inversion at 12q13, fusing the NAB2 and STAT6 genes. This leads to a nuclear expression of STAT6 that can be detected by immunohistochemistry [1,2]. Despite extensive investigation and aggressive treatment, the outcome of high-grade HPCs is still dismal [1,2]. Thus, early surgical intervention followed by determination of histological grade and stage of the disease are critical for HPCs. In the present case, the calvarial erosion was punched-out and accompanied by a dural defect. In addition, a peripheral branch of the middle meningeal artery supplied the tumor through the margin of the dural defect, which was confirmed on catheter angiography and intraoperative observations. Therefore, we assumed that the site may be the origin of the HPC. Furthermore, such bony and dural alterations, in addition to tumor invasion into the dura mater, were thought to reflect the aggressive nature of the tumor.

Calvarial erosion caused by brain tumors has been assumed to be associated with their peripheral location and slow-growing nature [6]. Based on CT findings, the tumor in the current case was assumed to arise and reach 36 mm in maximal dimension in less than 6 years. Given that symptomatic HPCs are commonly 40 mm or more, the period may indicate the natural history of HPCs, from genesis to appearance of symptoms [4].

In the present case, the dural tail sign and hyperostotic changes characteristic of meningiomas were not found. An extra-axial tumor atypical to meningioma may suggest an HPC with more aggressive biological behavior. Even after total tumor resection, careful follow-up is necessary for a long time, especially in the first 5 years after diagnosis, due to high rates of local recurrence and distant metastasis [2].

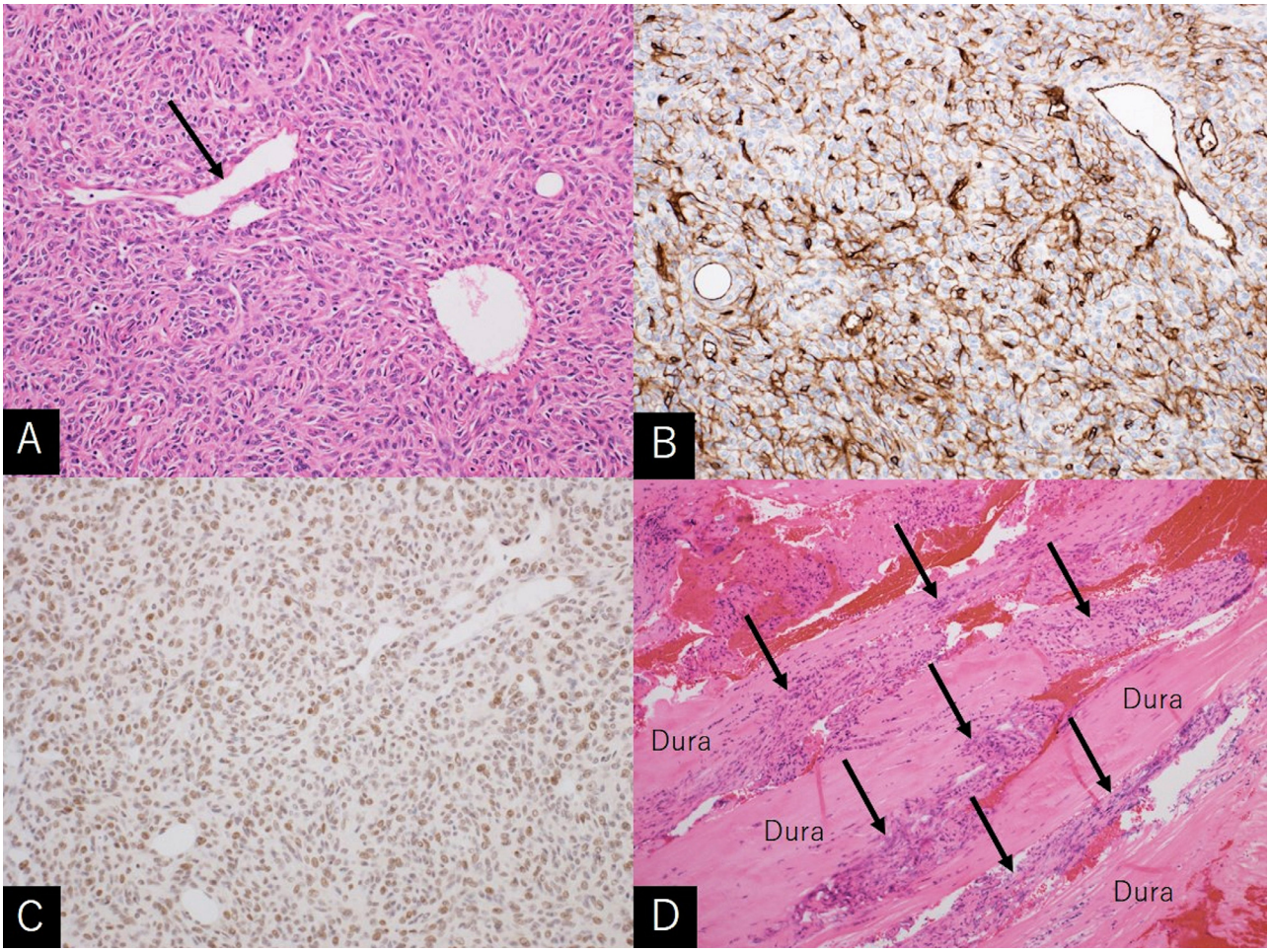

Fig. 6 – Microscopic appearance of the tumor comprising spindle-shaped cells accompanying branching vascular structures (arrow) and cell atypia (A). Immunohistochemical stains showing positive staining for CD34 (B) and STAT6 (C). Tumor invasion into the dura mater (arrows). (A) hematoxylin and eosin stain, × 200; B: CD34; C: STAT6, × 200; (D) hematoxylin and eosin stain, × 100.

Conclusion

A punched-out calvarial erosion and dural defect caused by an extra-axial tumor may be a high-grade HPC that requires extensive surgical resection and a long-time follow-up.

REFERENCES

[1] Champeaux C, Khan AA, Wilson E, Thorne L, Dunn L. Meningeal hemangiopericytoma and solitary fibrous tumour: a retrospective bi centre study for outcome and prognostic factor assessment. J Neurooncol 2017;134:387–95.
[2] Giordan E, Marton E, Wennberg AM, Guerriero A, Canova G. A review of solitary fibrous tumor/hemangiopericytoma tumor and a comparison of risk factors for recurrence, metastases, and death among patients with spinal and intracranial tumors. Neurosurg Rev 2020 Online ahead of print. doi:10.1007/s10143-020-01335-x.
[3] Chen Q, Chen XZ, Wang JM, Li SW, Jiang T, Dai JP. Intracranial meningeal hemangiopericytomas in children and adolescents: CT and MR imaging findings. AJNR Am J Neuroradiol 2012;33:195–9.
[4] Chiechi MV, Smirniotopoulos JG, Mena H. Intracranial hemangiopericytomas: MR and CT features. AJNR Am J Neuroradiol 1996;17:1365–71.
[5] Bajaj Multicystic oligodendroglioma with calvarial destruction. Asian J Neurosurg 2014;9:93–5.
[6] Lee YY, Van Tassel P. Intracranial oligodendrogliomas: imaging findings in 35 untreated cases. AJR Am J Roentgenol 1989;152:361–9.
[7] Muzumdar DP, Goel A, Pakhmode CK. Oligodendroglioma causing calvarial erosion. Neurol India 2003;51:129–30.
[8] Handzhiev DD, Kalevski SK, Handzhieva SV, Dzhenkov DL, Salieva-Badi S. Low grade astrocytoma causing dural and calvarial destruction. J Clin Neurosci 2017;40:84–9.
[9] Khan MA, Hashmi S. Low-grade astrocytoma causing calvarial scalloping. Pediatr Neurosurg 2007;43:155–7.
[10] Osborn RE, Ley CE. Astrocytoma with calvarial erosion. AJNR Am J Neuroradiol 1986;7:178.

[11] Cunningham DA, Lowe LH, Shao L, Acosta NR. Neuroradiologic characteristics of astroblastoma and systematic review of the literature: 2 new cases and 125 cases reported in 59 publications. Pediatr Radiol 2016;46:1301–8.
[12] Yunten N, Ersahin Y, Demirtas E, Yalman O, Sener RN. Cerebral astroblastoma resembling an extra-axial neoplasm. J Neuroradiol 1996;23:38–40.
[13] Maleki M, Robitaille Y, Bertrand G. Atypical xanthoastrocytoma presenting as a meningioma. Surg Neurol 1983;20:235–8.
[14] Satyarthee GD, Mahapatra AK. Giant pediatric glioblastoma multiforme causing primary calvarial erosion and sutural diastasis presenting with enlarged head. J Pediatr Neurosci 2015;10:290–3.