CASE REPORT

Anaplastic hemangiopericytoma manifesting as a rapidly enlarging extracranial mass lesion

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ABSTRACT

We, herein, present a patient with a recurrent anaplastic hemanigiopericytoma manifesting as a rapidly enlarging extracranial mass lesion, which was revealed by pathological and intraoperative findings. In practice, this case highlights the mandatory need for a careful long-term follow-up for patients with hemangiopericytoma, since recurrence with a greater degree of malignancy can develop following an extended disease-free interval, as such knowledge will be helpful for planning the optimal surgical procedures.

Key words: Anaplastic hemanigiopericytoma, brain tumor, gamma knife radiosurgery, meningeal neoplasm

Introduction

Hemangiopericytoma (HPC) arising from Zimmerman pericytes around capillaries and postcapillary venules is an extremely rare neoplasm, accounting for <1% of all central nervous system tumors.[1] The tumors are characteristically highly cellular and richly vascular, and are aggressive tending to recur even after macroscopic total resection. The 2007 World Health Organization classification of brain tumors was a revision of the former classification, which updated the concept of grading and added several new entities and variants; among the newly-codified entities was anaplastic HPC. The histological criteria for anaplastic HPC include at least more than five mitoses and/or necrosis, and at least two of the following items after hemorrhage, moderate to high nuclear atypia, and moderate to high cellularity.[2] Few accounts have been published of such a highly anaplastic and malignant change in a recurrent HPC. We, herein, described a patient with a recurrent anaplastic HPC after an initial complete resection and radiosurgery.

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Case Report

A 57-year-old male presented with a two-month history of right hand grip difficulty. On admission, magnetic resonance imaging revealed a well-demarked extra-axial mass lesion in the left frontal lobe [Figure 1a-c]. Partial invasion in the superior sagittal sinus (SSS) was observed on the gadolinium-enhanced T1-weighted sequences [Figure 1a-c]. After the surgical removal of the mass, a histological examination revealed HPC [Figure 1d-f]. Immunohistological findings showed that the CD34, EMA, and α-actin to be negative, while Bcl-2, S-100, vimentin, and CD99 were positive, and the MIB-1 index showed a percentage of 10-15%. Additional gamma-knife irradiation was performed for the remnant tumor in the SSS. After two years postoperatively, the remnant tumor underwent rapid regrowth [Figure 1g-i]; thus, necessitating a second surgical intervention [Figure 1j-l]. Histological examination revealed an anaplastic HPC with increasing mitotic cells [Figure 1m and n]. In addition, the recurrent tumor displayed a tumor doubling time of one week [Figure 2a-f]. A third operation was done [Figure 2g-i]. A histological examination conducted after the third procedure revealed further increasing of mitotic cells and an enlarged necrotic component [Figure 2j]. Immunohistological findings showed that CD34 was positive, and the MIB-1 index was almost 100% [Figure 2k and l]. The postoperative course was uneventful with no recurrence of the tumor evident three months following the third operation.

Discussion

HPC arising from Zimmerman pericytes around capillaries and postcapillary venules is an extremely rare neoplasm, accounting for <1% of all central nervous system tumors.[1]

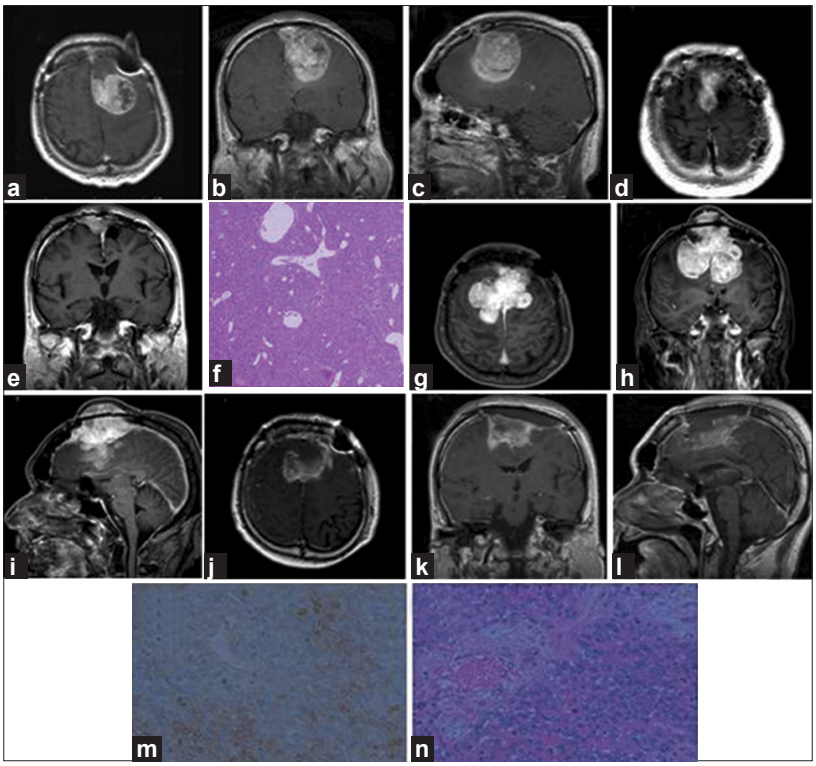

Figure 1: The initial enhanced T1-weighted MR image. (a-c) Show an extra-axial mass lesion in the left frontal lobe. (d, e) Postoperative MR images revealing the remnant tumor beside the SSS. (f) Histopathological findings showed the diagnosis of HPC. (g-l) Additional gamma-knife irradiation was performed. Two-years after the operation, the remnant tumor showed a rapidly regrowth (g-i), and the patients underwent a second surgical intervention (j-l). An immunohistochemical analysis using CD34 was positive (m) and a histological examination revealed an anaplastic HPC with increasing mitotic cells (n)

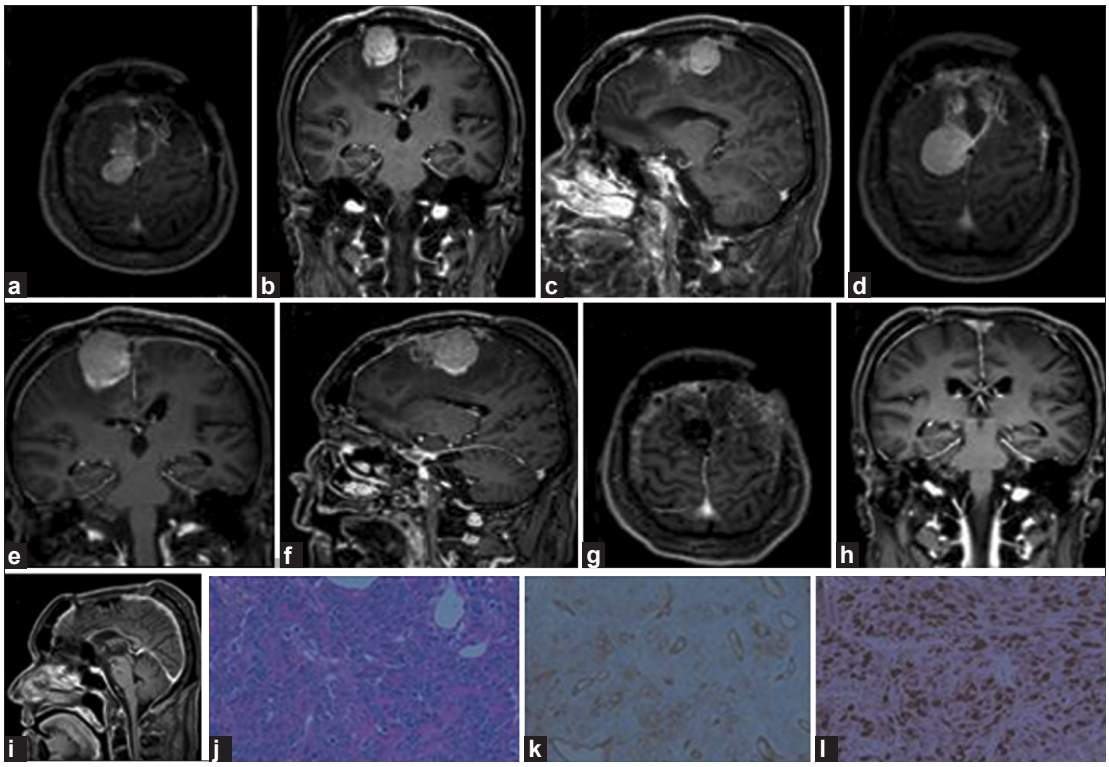

Figure 2: Synospsis of the recurrence of the tumor. (a-f) The tumor recurred, with a tumor doubling time of 1-week. (g-i) The tumor recurrence necessitated a third operation. (j) A histological examination revealed further increased mitotic cells and an enlarged the necrotic component. (k) An immunohistological examination revealed CD34 positivity. (l) The MIB-1 index was almost 100%

The tumors are characteristically highly cellular and richly vascular, and are aggressive tending to recur even after macroscopic total resection. A microscopic diagnosis is based on the recognition of an architecture characterized by a pericytomatous pattern. However, the same pattern occurs in a variety of neoplasms, such as fibrous histiocytoma, solitary fibrous tumor, and fibrous meningioma. Therefore, distinguishing HPC from other tumors can be difficult, especially when the characteristic features of other neoplasms are inconspicuous. However, because of differences in the prognosis and patient management, a proper diagnosis is critical. For example, HPC has a much higher propensity to recur and metastasize than fibrous meningioma. In addition, an immunohistochemical analysis is helpful for differentiating among meningeal tumors, since there is a considerable overlap in their immunoprofiles.

The 2007 World Health Organization classification of brain tumors was a revision of the former classification, which updated the concept of grading and added several new entities and variants; among the newly-codified entities was anaplastic HPC. The histological criteria for anaplastic HPC include at least more than five mitoses and/or necrosis, and at least two of the following items after hemorrhage, moderate to high nuclear atypia, and moderate to high cellularity.[2] A malignant clinical course is associated with a large tumor, an increased mitotic rate, high cellularity, immature and pleomorphic tumor cells, and foci of hemorrhage and necrosis.[3] The best treatment to manage the HPCs has not yet established. However, several authors suggested that a significant prolongation in recurrence-free interval as well as long-term survival was observed in patients who underwent a complete macroscopic resection and adjuvant radiation therapy.[4,5] Thus, HPCs are difficult to cure surgically; postoperative radiotherapy has been proposed, even after a gross total tumor resection.[6,7]

In the present case, the residual tumor cells, after the initial excision, grew, and accumulated genetic and phenotypic changes to become more highly malignant at the time of clinical recurrence. Few accounts have been published of such a highly anaplastic and malignant change in a recurrent HPC. Therefore, the present case serves as an important warning of this risk. In summary, we, herein, described a patient with a recurrent anaplastic HPC after an initial complete resection and radiosurgery. This case highlights the mandatory need for a careful long-term follow-up for patients with HPC, since recurrence with a greater degree of malignancy can develop following an extended disease-free interval.

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