

Case Report

Rapid Clinical Course of Cerebral Metastatic Angiosarcoma from the Heart

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We report here one case of rapid and aggressive course of cerebral metastatic angiosarcoma from the heart. A 36-year-old man presented with 10-days history of headache. Magnetic resonance imaging demonstrated subacute hemorrhage with a small region of enhancement in right parietal region and the pathological diagnosis was angiosarcoma. Transthoracic echocardiography demonstrated 3.2×3 cm sized mass on right atrial wall. Newly developed lesion was reoperated, three and four weeks later respectively, and whole brain radiotherapy of total 30 Gy was done. With the interval of two months, gamma knife surgery was done for new lesions two times, which were well controlled. Newly developed lesions rapidly happened even in the adjuvant treatment. He died 9 months after the diagnosis because of the aggravation of primary cancer. The cerebral metastatic angiosarcoma from the heart showed the rapid aggressive behavior and the closed follow-up could be needed for the adjuvant treatment.

Key Words : Angiosarcoma · Cerebral · Heart · Metastasis.

INTRODUCTION

Angiosarcoma is a rare malignant vacular tumor, which may originate in face and scalp, liver, skin and other soft tissues¹⁸. Metastatic angiosarcomas of central nervous systems are rare, a few cases have been reported and their prognosis were poor^{1,8,9,12,14,16}. We report here one case of cerebral metastatic angiosarcoma from the heart. This patient showed the poor prognosis with rapid clinical course even in combined treatment to the metastatic lesions. We would like to describe the radiologic findings of metastatic cerebral angiosarcoma and the rapid clinical course.

CASE REPORT

A 36-year-old man presented with 10-days history of headache. On admission, his neurological examination was within normal ranges. A computed tomography (CT) of the head showed the intracerebral hemorrhage of subacute stage on right parietal area (Fig. 1A). Magnetic resonance imaging (MRI) demonstrated a heterogeneous mixed signal intensity lesion on T1-weighted images with a small region of enhancement in

right parietal region. The lesion showed heterogenous mixed signal intensity on T2-weighted images (Fig. 1B-D). Provisional diagnosis was a hemorrhage from cavernous angioma. The mass was totally removed. The pathological diagnosis was angiosarcoma, which was highly cellular with cellular atypia and frequent mitosis (Fig. 2A). On immunohistochemistry, the tumor was positive for CD34, factor VIII, actin, CD31 and Ki-67 labeling index was high at 80% (Fig. 2B-E). For evaluation of primary cancer, we checked positron emission tomography-computed tomography, which showed the hot uptake of right atrial wall and right iliac bone. Transthoracic echocardiography demonstrated 3.2×3 cm sized round mass without stalk on right atrial wall and small amount of pericardial effusion. Three weeks later, he revisited with the one-week history of left hemiparesis and sensory change. Brain CT and MRI showed another intracerebral hemorrhage associated with enhancing lesion of right parietal area (Fig. 3A, B). Tumor was totally removed and symptom was improved. Four weeks later, patient complaint aggravation of previous symptom. Brain MRI showed the marginal enhanced lesion on post-resection cavity associated with perilesional edema (Fig. 3C), which was operated again.

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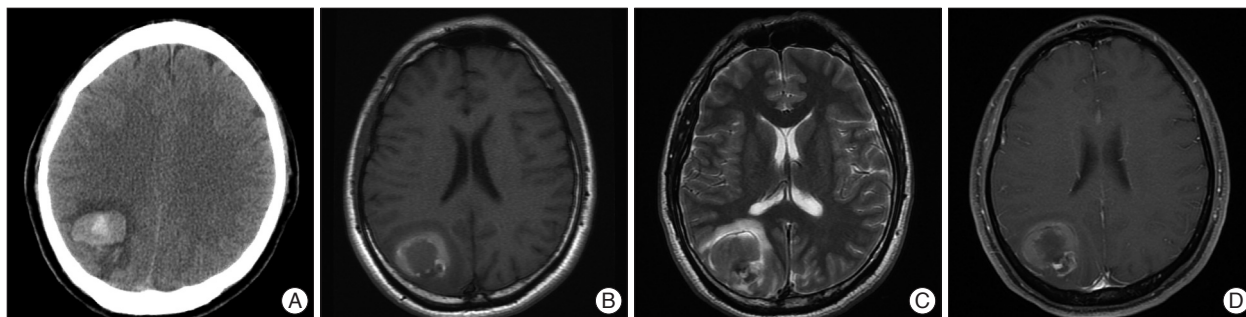


Fig. 1. Initial radiologic findings. CT showed the intracerebral hemorrhage of subacute stage on right parietal area (A). MRI demonstrates a heterogeneous mixed signal intensity lesion on T1 and T2-weighted images (B and C) with a small region of enhancement in right parietal region (D). CT : computed tomography, MRI : magnetic resonance imaging.

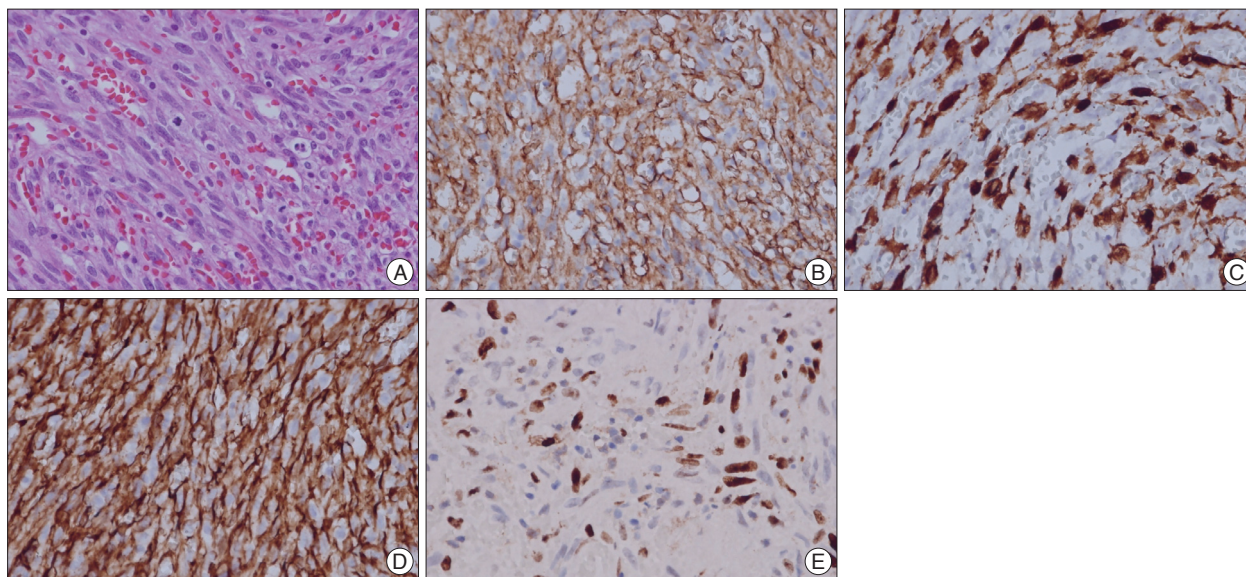


Fig. 2. Pathologic findings. A : Highly cellular with cellular atypia and frequent mitosis (H&E, original magnification $\times 400$). B : Immunopositive for CD34 (original magnification $\times 400$). C : Immunopositive for actin (original magnification $\times 400$). D : Immunopositive for CD31 (original magnification $\times 400$). E : Ki-67 labeling index at 80% (original magnification $\times 400$).

And, the whole brain radiotherapy of total 30 Gy was done. Two months later, routine follow-up brain MRI showed less than 1 cm-sized, four newly enhancing lesions (Fig. 3D) and there was no recurrence on two post-resection cavity. Gamma knife surgery (GKS) was done for these newly developed lesions (marginal dose, 20 Gy; 50% isodose curve). Two months later, he complained dizziness and nausea sensation. Brain MRI demonstrated the decrease of GKS-treated enhanced lesions and there were newly developed lesions; 2.3 cm sized hemorrhagic mass lesion with heterogeneous enhancement in right cerebellar hemisphere (Fig. 3E) and multiple small lesions in the cerebral hemisphere. GKS was done for new lesions (marginal dose, 18-20 Gy; 50% isodose curve). Six weeks later, headache was developed with improved previous symptoms. Brain MRI showed that the newly multiple lesions were detected (Fig. 3F) with the decrease of previous GKS-treated lesions. Even if he had received chemotherapy at the oncology department, cardiac mass and pericardial effusion were increased and he died 9 months after the diagnosis.

DISCUSSION

Angiosarcomas are rare malignant tumor originating from endothelial cells of arteries, veins, and lymphatic channels and are one of the most rare type of human tumors^{9,20}. These tumors constitute less than 1% of all sarcomas which usually locate in the head, face, liver, skin, and soft tissue. Primary and secondary cerebral angiosarcomas have been rarely reported and one study summarized these rare tumors¹⁶. Metastatic cerebral angiosarcomas are more frequent than primary angiosarcomas and most often occur in the right atrium. Angiosarcomas of the heart are common cardiac sarcoma. In the previous study of Donsbeck et al.⁷, angiosarcoma was the second most frequent histological type of cardiac sarcoma and occurred more frequently in men, with a common age of presentation between the third and fifth decades of their lives. Metastatic lesions often manifest prior to the diagnosis of cardiac angiosarcoma^{13,20}. Metastases occur to the lung, liver, central nervous system and bone. This case was a cerebral metastasis from an-

giosarcoma of the heart manifested by intracerebral hemorrhage.

Radiologically, primary and secondary cerebral angiosarcoma have a frequent tendency of hemorrhage with marked perilesional edema on CT, and MRI may show well-circumscribed areas of hemorrhage with surrounding edema and increased signal intensity with partial contrast enhancement^{3,11}. This is the reason that we misdiagnosed angiosarcoma as a cavernous angioma at the first time and we should concern another intracranial tumor in the consideration of perilesional edema.

The biological behavior of angiosarcomas is not well known because these tumors are rare. After surgical resection of angiosarcoma, chemotherapy and radiation therapy could be useful for the prevention of brain metastasis theoretically. However, once angiosarcomas have metastasized to the brain, chemotherapy is not effective, because useful drugs for treatment of sarcomas do not

penetrate the central nervous system effectively⁸). The outcomes of patients with angiosarcomas in general are poor because these tumors are mostly malignant and aggressive. According to the reported data, the prognoses of cardiac angiosarcoma are poor, and once these have metastasized, the prognoses become worse with a mean survival of 2 to 24 months from the time of detection²). In primary cerebral angiosarcoma, the clinical course is characterized by the rapid onset of symptoms and poor prognosis^{6,17}). This patient also showed the rapid clinical courses. The interval to the newly developed or recurred lesions after operation was three to four weeks. After whole brain radiotherapy (WBRT), the interval to the newly developed lesions was two months. For newly developed lesions, gamma knife surgery was a good treatment modality for local control. Traditionally, sarcoma, renal cell carcinoma and melanoma have been considered as radioresistant tumors. In the past, brain metastasis from sarcoma were often treated primarily with fractionated WBRT for improving local control and decreasing distant brain failure, but a median survival of 3 months was the usual outcome⁹). For WBRT, the total dose is 25 to 39 Gy and daily fractions of 3 to 6 Gy are administered. With the invent of stereotaxic radiosurgery, there has been renewed interest in more aggressive management of potentially radioresistant brain metastases^{4,5,15}). Local control rate was 88% for radioresistant tumors, which result was similar to the results reported in other radiosurgery series of brain metastases^{10,19}). Out of radioresistant tumors, stereotaxic radiosurgery was less effective in treating melanoma or sarcoma brain metastases than renal

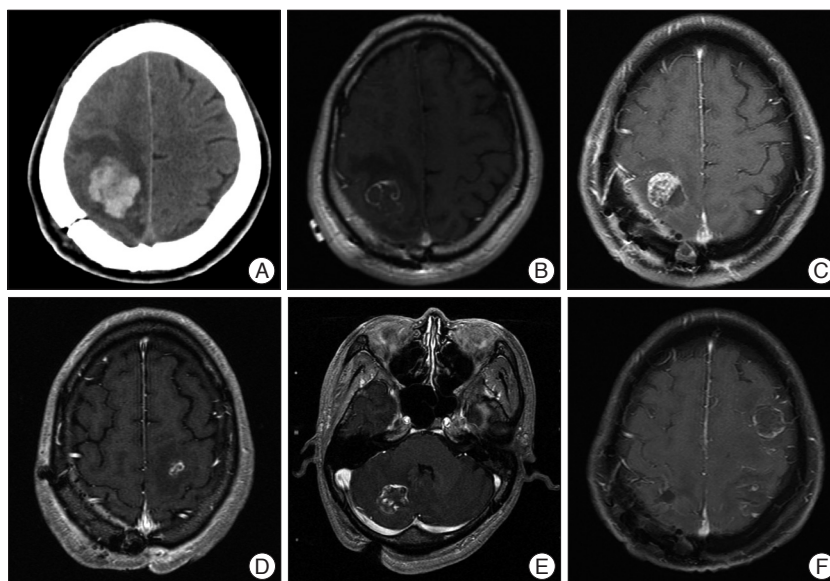


Fig. 3. Follow-up radiologic findings. A and B : Three weeks later, CT and MRI show another intracerebral hemorrhage associated with enhancing lesion of right parietal area. C : Four weeks later, MRI shows the marginal enhanced lesion on post-resection cavity associated with perilesional edema. D : After WBRT two months later, MRI shows less than 1 cm-sized, four newly enhancing lesions. E : Two months later, there are newly developed lesions; 2.3 cm sized hemorrhagic mass lesion with heterogeneous enhancement in right cerebellar hemisphere. F : Six weeks later, MRI shows that the new multiple lesions are detected. CT : computed tomography, MRI : magnetic resonance imaging, WBRT : whole brain radiotherapy.

cell carcinoma brain metastases⁵). In this case, the progression-free survival was 2 months after WBRT and GKS was a good adjuvant treatment modality for newly developed lesions. Even in combined treatment, newly developed lesions rapidly happened with the interval of 6 weeks to 2 months and the cause of death was the aggravation of primary cancer.

CONCLUSION

Metastatic cerebral angiosarcoma showed the rapid clinical courses and closed follow-up could be needed for the adjuvant treatment.

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