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Case Report

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Management of intracranial angiosarcoma metastasis to the right lateral ventricle choroid plexus by radiotherapy: A case report

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Abstract

Angiosarcomas are a type of rare malignant endothelial tumor. While its most common metastasis site is the scalp, cerebral metastases are rare. A 46-year-old woman came with a headache. A cranial magnetic resonance imaging (MRI) showed a hemorrhagic lesion within the right lateral choroid plexus and T1 contrast-enhancing lesions in the falx cerebri and right frontal dura. We used mini-craniotomy and neuronavigation to remove a purple-colored extradural lesion in the right frontal region. Histopathological analysis later confirmed the diagnosis of angiosarcoma. Cardiac MRI demonstrated the presence of a left atrial mass, which led to the decision to operate on the patient by the cardiovascular surgery team. After the surgery, the patient underwent radiotherapy and chemotherapy, as there was no cranial metastasis necessitating surgical resection. The cranial metastases responded to the treatment on follow-up, and there was no evidence of recurrence on the three-year follow-up. Although they are uncommon, pathology that indicates the presence of angiosarcoma requires cardiac screening. These patients may not require aggressive surgery as they respond well to adjuvant therapies.

Keywords: angiosarcoma, metastasis, radiotherapy, case report

1. Introduction

Cerebral metastases of angiosarcomas are a rare but very severe clinical situation, which confirms the malignant nature of these vascular tumors. Angiosarcomas, which develop from endothelial cells, are capable of forming brain metastases, resulting in substantial neurological sequelae and complicated treatment strategies (1). Brain metastases from angiosarcomas are a harbinger of a dismal clinical course, reflecting the aggressive behavior of the tumor and the challenges of managing metastatic disease in the central nervous system (2). Despite the advances in diagnostic and therapeutic approaches, treating metastatic cerebral angiosarcoma poses great challenges, requires a multidisciplinary approach, and emphasizes the need for further studies on targeted therapies and prognostic markers to improve the patient's outcome. We present a case that underscores the importance of the multidisciplinary approach to cerebral metastases of cardiac angiosarcomas.

2. Case Presentation

A 46-year-old female was referred to us from the emergency department. She has had episodes of headaches for the last three months that have become more severe recently. Computed tomography (CT) imaging revealed a hemorrhagic mass within the right ventricle (Fig. 1). Subsequent MRI imaging revealed multiple lesions in the falx cerebri, right frontal dura, and choroid plexus of the right lateral ventricle that were T1-contrast enhancing (Fig. 1).

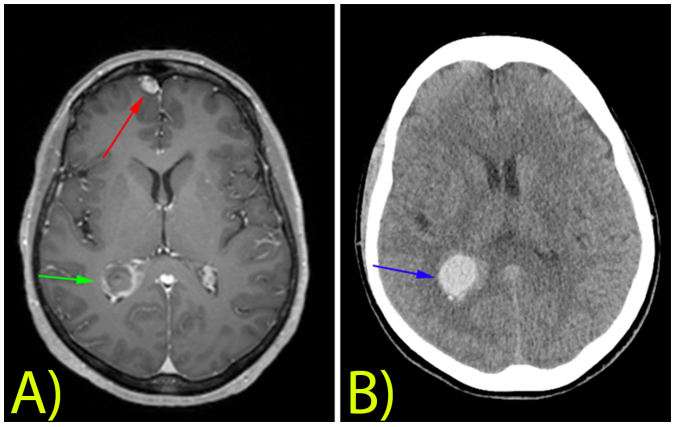

Fig. 1. T1 Contrast enhancing sequence(A), right atrial peripherally contrast-enhancing lesion is shown with the green arrow, while right frontal metastasis is shown with red arrow. On CT imaging (B), note the hemorrhagic lesion in the right ventricle, shown with a blue arrow

The patient was awake with no neurological symptoms. Apart from the headache, there were no notable features. She had no medical history, no tobacco use, and no history of contraceptive use.

Since there was no mass effect and no neurological symptoms, a biopsy from the right frontal dura was planned. The biopsy was obtained using neuronavigation and a mini-craniotomy. The lesion was extradurally located, purple-ish in color, and soft in nature. Postoperatively, the patient was

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discharged without complications.

On morphological analysis, the tumor exhibited atypical epithelioid cell nodules with eosinophilic cytoplasm on a highly inflammatory background. Immunohistochemical analysis revealed the tumor cell expression of endothelial markers CD31, CD34, and also ERG. The morphological findings and the immunophenotypic profile supported a diagnosis of grade 2 angiosarcoma.

We obtained a cardiac MRI scan, revealing a left atrial mass, which was removed by the cardiovascular surgery team, and the patient was discharged without complications.

The patient was discussed on our multidisciplinary tumor board. Since there was no mass effect of the right lateral ventricular metastasis, close follow-up and radiotherapy followed by chemotherapy using paclitaxel were planned. The patient received ten fractions of radiotherapy followed by 18 months of paclitaxel therapy and is disease-free after three years of follow-up (Fig. 2).

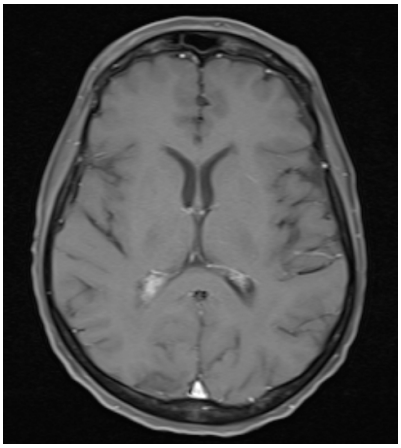

Fig. 2. T1 Contrast-enhancing sequence showing no recurrent lesions

3. Discussion

Intracerebral metastases originating from angiosarcomas are rare but emphasize the malignant nature of these vascular tumors and their ability to cause devastating neurologic sequelae (3). Approaches to the management of intracerebral angiosarcoma metastases usually include surgical resection, radiotherapy, and chemotherapy (4). Nevertheless, the effectiveness of such treatments is not high enough, which demonstrates that angiosarcomas are highly aggressive and do not respond to regular therapies. The prognosis of patients with cerebral metastases is still unfavorable (3), which emphasizes the urgent need for new therapeutic strategies and targeted therapies.

Molecular biology and immunotherapy have recently made great strides (5,6) and could provide new opportunities for treatment, thereby making the research into the pathogenesis of angiosarcoma even more critical. In addition, the importance of a multidisciplinary team must be considered as interdisciplinary care, including neurosurgeons, medical oncologists, radiologists, and pathologists, which is critical for the best patient outcomes.

To sum up, the treatment of intracranial angiosarcoma metastasis should be individualized according to the patient's clinical picture and the nature of the metastatic lesions. Our case and the recent literature case highlight the effectiveness of the multidisciplinary approach, including radiotherapy and chemotherapy. These cases add to the increasing evidence for the requirement of personalized treatment regimens and underscore the need for further research to formulate more conclusive treatment recommendations for this uncommon clinical entity.

Conflict of interest

The authors declare that they have no competing interests.

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None to declare.

Authors' contributions

Concept: H.B., T.Y., Design: H.B., T.Y., Data Collection or Processing: B.B.A., Analysis or Interpretation: B.B.A., M.S.B., Literature Search: B.B.A., M.S.B., Writing: B.B.A., M.S.B.

Ethical Statement

The need for ethics approval was waived as this is a case report.

References

1. Young RJ, Brown NJ, Reed MW, Hughes D, Woll PJ. Angiosarcoma. Lancet Oncol. 2010 Oct;11(10):983–91.

2. Jung S-H, Jung T-Y, Joo S-P, Kim H-S. Rapid clinical course of cerebral metastatic angiosarcoma from the heart. J Korean Neurosurg Soc. 2012 Jan 31;51(1):47–50.

3. Drosos E, Kalyvas A, Komaitis S, Skandalakis GP, Kalamatianos T, Liouta E, et al. Angiosarcoma-related cerebral metastases: a systematic review of the literature. Neurosurg Rev. 2020 Aug;43(4):1019–38.

4. Schlemmer M, Reichardt P, Verweij J, Hartmann JT, Judson I, Thyss A, et al. Paclitaxel in patients with advanced angiosarcomas of soft tissue: a retrospective study of the EORTC soft tissue and bone sarcoma group. Eur J Cancer. 2008 Nov;44(16):2433–6.

5. Ravi V, Subramaniam A, Zheng J, Amini B, Trinh VA, Joseph J, et al. Clinical activity of checkpoint inhibitors in angiosarcoma: A retrospective cohort study. Cancer. 2022 Sep 15;128(18):3383–91.

6. Florou V, Rosenberg AE, Wieder E, Komanduri KV, Kolonias D, Uduman M, et al. Angiosarcoma patients treated with immune checkpoint inhibitors: a case series of seven patients from a single institution. J Immunother Cancer. 2019 Aug 8;7(1):213.