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Case Report

A case report of Extra-neurologic metastasis of ventricular meningioma with literature review ☆

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ABSTRACT

Ventricular meningiomas are neoplastic cells originating from the ependymal lining of the central canal of the spinal cord and the ventricles of the brain. These tumorigenic cells predominantly manifest in the fourth ventricle, followed by the spinal cord. Most intraparenchymal ventricular meningiomas are located within the brain tissue, exhibiting a higher degree of malignancy compared to their intracerebroventricular counterparts. While intracranial dissemination and metastasis to the spinal cord can occur, extra-neurologic metastasis is an exceedingly rare phenomenon that lacks a clear elucidation regarding its underlying mechanism. The authors presented a case of supratentorial brain parenchymal type ventricular meningioma surgical treatment in a young female patient, occurring two years after the development of multiple metastases in both lungs, pleura, and mediastinum. This may be attributed to the high malignancy degree and strong invasiveness of this lesion, as well as its proximity to the dura mater and venous sinus. The craniotomy provided an opportunity for tumor cells to invade the adjacent venous sinus, leading to dissemination through the blood system. Additionally, postoperative radiation and chemotherapy were administered to inhibit tumor angiogenesis; however, these treatments also increased the likelihood of tumor cell invasion into neighboring brain tissues and distant metastasis.

Introduction

Ventricular meningiomas are neuroepithelial tumors that originate from the ventricular cells of the central canal and spinal cord or the ventricular cells of the interventricular white matter in the brain. Most of these tumors are located within the brain parenchyma, originating either from ectopic ventricular cells during embryonic development or as tumors growing into the brain parenchyma from the walls of the brain's ventricles. The clinical manifestations depend on

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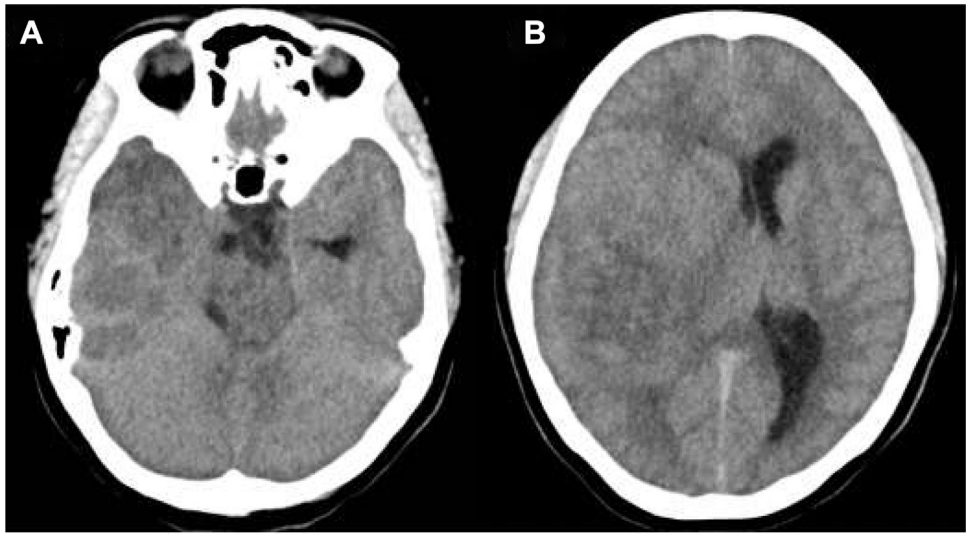

Fig. 1 – (A, B) Cranial plain CT shows a large occupying lesion with mixed density in the right temporal lobe, which is accompanied by a few high-density hemorrhages; the right ventricle is narrowed by compression, and the midline is shifted to the left.

tumor location, with varying symptoms observed based on different locations. If the tumor occurs in the cortical area, it primarily manifests as headache and epilepsy; whereas if it arises in the posterior cranial fossa, it can lead to intracranial hypertension, obstructive hydrocephalus, or focal neurological symptoms. The malignancy of these tumors is high, with potential for intracranial spread or metastasis along the cerebrospinal fluid within the spinal cord; however, extra-neurological metastasis is exceedingly rare. Herein, we present a case of parenchymal (WHO grade 3) ventricular meningioma located in the temporal lobe that exhibited multiple extra-neurological metastases to the lungs, thorax, and mediastinal lymph nodes two years post-surgery. We also provide a comprehensive review of relevant literature which predominantly suggests that occurrences of extra-neurological metastases may be associated with craniotomy procedures, shunt surgeries, and prolonged patient survival. Surgical resection of cranial tumors exposes tumor cells to either lymphatic or blood circulation systems while surgical interventions such as craniotomy or shunt surgery can disrupt blood-brain barrier integrity leading to extracranial metastases; additionally, extended patient survival increases the likelihood of extracranial metastatic events. In this report and based on previous literature findings, we analyze and summarize mechanisms underlying metastasis.

Case report

The patient is a 19-year-old female presenting with a three-month history of headache and dizziness, which commenced without an identifiable etiology. The dizziness is characterized by paroxysmal episodes and lacks any sensation of rotation. Subsequently, the severity of dizziness escalated significantly, accompanied by nausea and vomiting. Notably, there were no associated symptoms such as hearing loss, tinnitus or deafness, facial numbness, dysphagia or choking. Neurological examination revealed no positive findings; however, CT imaging indicated the presence of a mixed density mass in the right temporal lobe with potential hemorrhage (Fig. 1A, B). The MRI scan revealed a mass-like long T1 and long T2 signal shadow in the right temporal lobe, with high signal intensity on T2-FLAIR imaging (Fig. 2A–C). The morphology of the lesion appeared irregular, measuring approximately 6.2 × 7.7 cm in its largest cross-section, and exhibited scattered patchy short T1 signal shadows within it (Fig. 2A), indicative of hemorrhage. The diffusion-weighted imaging (DWI) demonstrated an inhomogeneous signal pattern, characterized by both areas of high and slightly low signal intensity on apparent diffusion coefficient (ADC) mapping (Fig. 4A, B). Furthermore, the lesion involved the choroid plexus of the right ventricle (Fig. 3A–C) as well as the brain surface of the right temporal lobe. The enhanced scanning lesion exhibited prominent heterogeneous enhancement (Fig. 5A, B), accompanied by significant mass effect. Compression resulted in narrowing of the right lateral ventricle, causing a leftward shift of the midline structures by approximately 1.3 cm. Under general anesthesia, surgical resection of the tumor located in the right temporal region was performed. Intraoperatively, it was observed that the tumor extended into the lateral ventricle with substantial growth and lacked peripheral involvement. Additionally, it displayed an exceptionally rich blood supply and measured approximately 7.0 × 8.5 cm in size. Postoperative pathology revealed a poorly differentiated ventricular meningioma characterized by tumor cells forming a perivascular chrysanthemum-like pattern with evident nuclear division. Immunohistochemical results revealed the following: Ki-67 expression was observed in more than 40% of cells, GFAP showed partial positivity, S-100 exhibited focal positivity, IDH1 was negative, 0lg-2 displayed partial positivity, ATRX was positive, IGMT showed weak positivity, EMA demonstrated weak positivity, P53 expression was

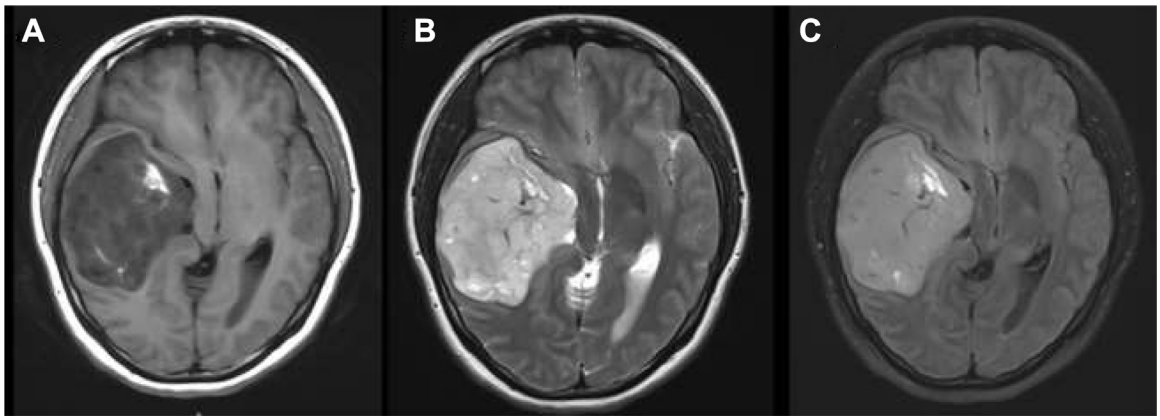

Fig. 2 – (A-C) show a right temporal lobe mass with high and low mixed signals on T1WI, T2WI, and T2 -flair sequences, and a little high signal in the mass on T1WI represents the presence of hemorrhage in the tumor; the mass is large, and the right ventricle is narrowed by the compression, and the midline structures are shifted to the left by the compression.

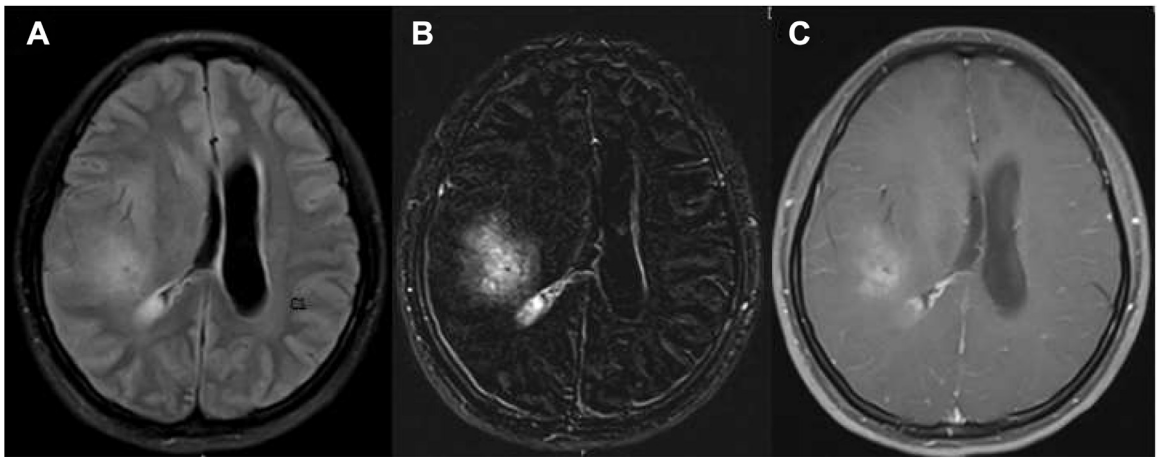

Fig. 3 – (A-C) shows that the MRI T2 -flair sequence, enhanced scanning silhouette, and enhancement maps show a patchy and markedly enhanced focus in the posterior horn of the right ventricle, suggesting that the lesion involves the right lateral ventricle.

observed in 10% or more of cells. CgA and Syn were both negative while CD56 showed positive staining (Fig. 7A). Subsequently, the patient was transferred to the radiotherapy department for radiation therapy.

Two years later, the patient began to have recurrent cough and shortness of breath, and chest CT examination showed: scattered nodular foci in both lungs, multiple nodules and mass shadows in the left thoracic cavity, and inhomogeneous enhancement of the lesions on enhancement scanning; and a large amount of pleural effusion on the left side, with the left lung tissues being compressed and aggregated in the left hilar region, and mediastinal compression shifted to the right side (Fig. 6A–D).Fiberoptic bronchoscopy showed a mass growth seen in the upper lobe opening of the left lung with luminal occlusion, and an externally compressive stenosis in the lower lobe opening of the left lung with moderate luminal narrowing. Pathology was metastatic and immunohistochemistry considered metastatic poorly differentiated ventricular meningioma, consistent with intracranial tumor metastasis (Fig. 8A).

Two years later, the patient presented with recurrent cough and dyspnea. Chest CT examination revealed scattered nodular foci in both lungs, multiple nodules and mass shadows in the left thoracic cavity, and heterogeneous enhancement of the lesions on contrast-enhanced scanning. Additionally, a significant amount of pleural effusion was observed on the left side, resulting in compression and consolidation of lung tissues in the left hilar region as well as mediastinal shift towards the right side (Fig. 6A–D). Fiberoptic bronchoscopy demonstrated luminal occlusion caused by a growing mass at the upper lobe opening of the left lung, along with moderate luminal narrowing due to external compression at the lower lobe opening. Pathological analysis confirmed metastatic poorly differentiated ventricular meningioma through immunohistochemistry staining, consistent with intracranial tumor metastasis (Fig. 8A).

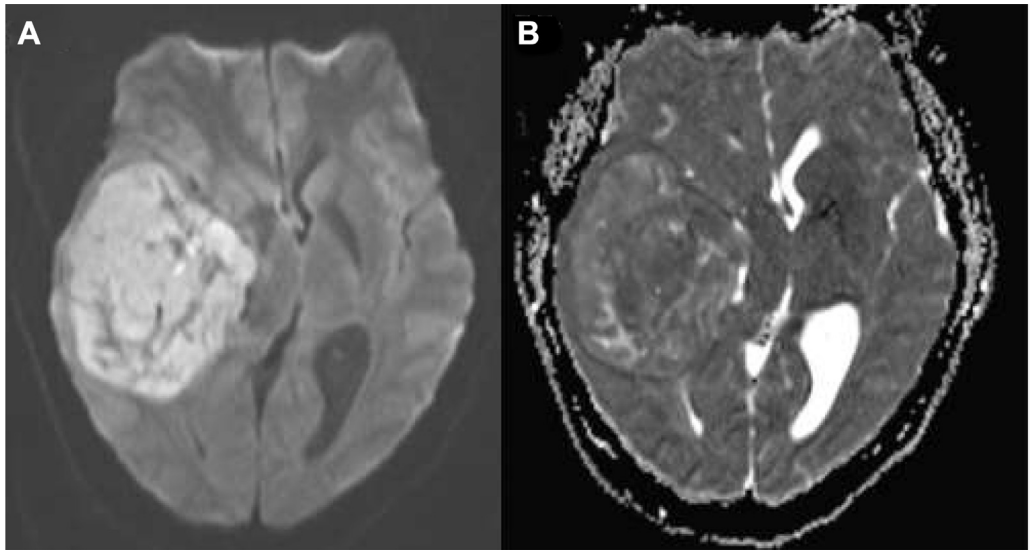

Fig. 4 – (A and B) show that the mass has most of its diffusion restricted on DWI, with a small area of unrestriction.

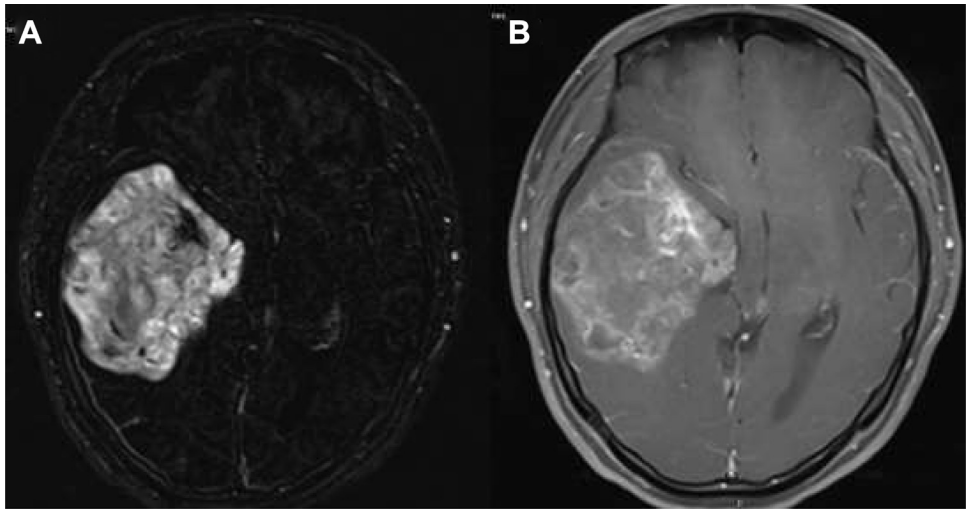

Fig. 5 – (A, B) MRI enhancement scan of the solid portion of the tumor shows inhomogeneous significant enhancement.

Discussion

Although gliomas are the most prevalent tumors of the central nervous system, extracranial metastases are infrequent. Liwnicz et al [1] reported that glioblastoma (41.4%) was the most common type of glioma to metastasize, followed by medulloblastoma (26.7%), ventricular meningioma (16.4%), astrocytoma (10.3%), and oligodendroglioma (5.25%). Metastases can occur in any organ or structure in the body, with lymph nodes, bones, lungs, and abdominal organs being the typical sites of metastasis. The most frequent sites for metastasis are bones and multiple locations throughout the body, followed by the lungs. Previous cases suggest that metastasis likely occurs through cerebrospinal fluid dissemination, as well as blood and lymphatic system spread. The mechanism of metastasis remains unclear, and some studies suggest that it may be associated with tumor cell exposure to the lymphatic system or blood circulation during surgery, disruption of the blood-brain barrier (e.g., craniotomy, shunt surgery), and prolonged patient survival. Specifically, these factors include: ① Intracranial barriers play a crucial role in impeding metastasis outside the nervous system; however, surgical procedures such as dura mater opening or disruption of the blood-brain barrier provide an opportunity for tumor cells to interact with extracranial lymphatics and blood vessels. Consequently, exposed tumor cells can directly invade damaged tissues leading to vascular invasion, meningeal infiltration, and cranial lymphatic system involvement. Several reports indicate that nearly 96% of patients who develop extracranial metastases have undergone craniotomy [2]. Therefore, craniotomy is often considered a significant contributing factor to extracranial metastasis development. Additionally, ventriculo-peritoneal shunt represents another important cause of CNS tumors'

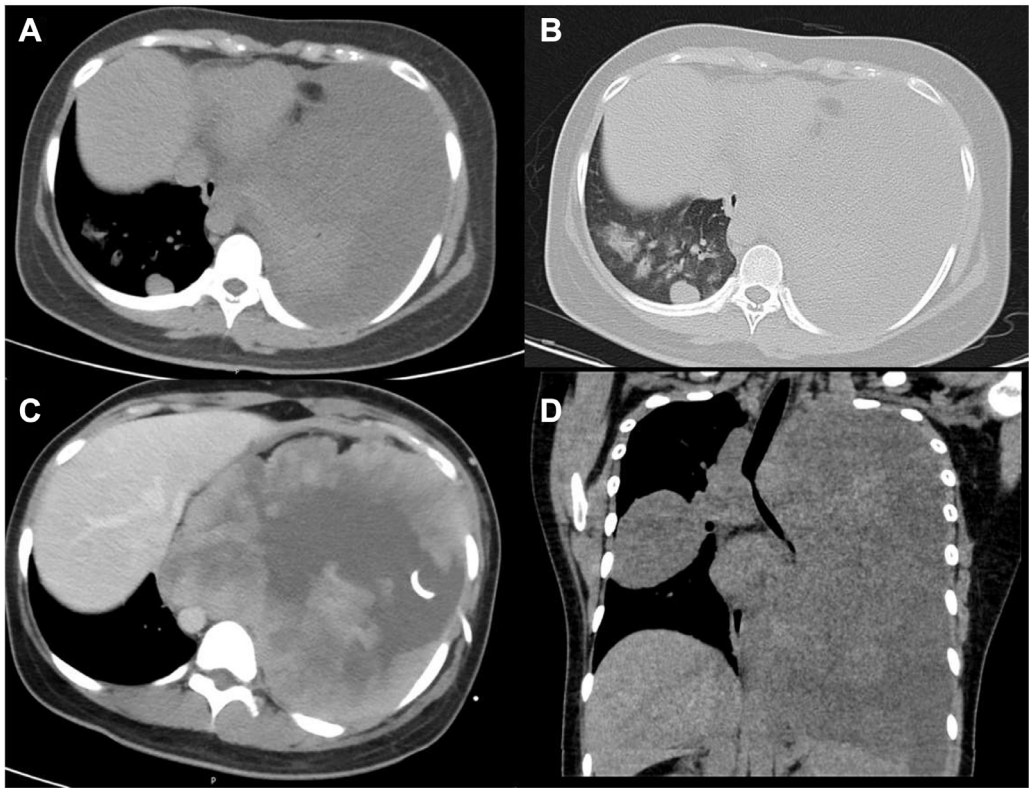

Fig. 6 – (A-D) Chest CT in April 2023 showed multiple nodules and masses in both lungs and pleura, predominantly in the left lung, with marked inhomogeneous enhancement of the lesions on enhancement scan (C); the mediastinum was shifted to the right by compression, and there was enlargement of the mediastinal lymph nodes, as well as pleural effusion on the left side.

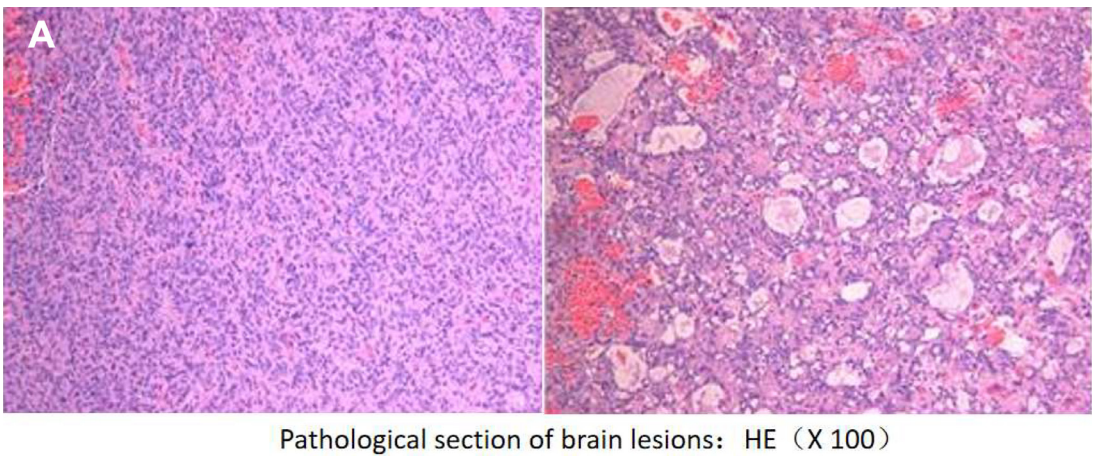

Fig. 7 – (A) shows an intracranial lesion: tumor cells form a daisy-shaped mass around blood vessels with active nuclear division.

extracranial metastasis [3]. ② The central nervous system possesses robust protective mechanisms against metastasis occurrence; for instance, dense dura wrapping around cerebral venous sinuses prevents tumor cell infiltration [4]. However, leukopenia-induced immunosuppression and decreased dural density resulting from long-term radiotherapy can contribute to extracranial metastasis. ③ Advances in diagnostic techniques and treatment have significantly extended the survival of cancer patients. Previously, patients might succumb to intracranial hypertension or other complications before experiencing extracranial metastasis [5]; however now with prolonged survival comes an increased likelihood of developing such distant spread. Moreover, evidence suggests that longer patient survival enhances the probability of glioma tumor

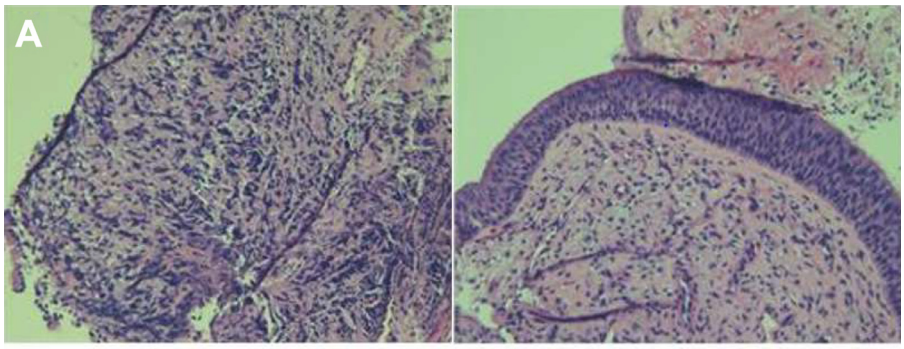

Fig. 8 – (A) shows local epithelial squamous metaplasia, local tissue extrusion deformation, nuclear hyperchromic cells found in the interstitium.

cell detachment into the lymphatic system [6]. Younger age at onset, incomplete tumor resection, high-grade or mucopapillary ventricular meningiomas, and higher tumor proliferation index are risk factors associated with the development of disseminated and distant metastases in patients with ventricular meningiomas. 2. In this case, the patient was a young woman who presented with a supratentorial parenchymal type (WHO grade 3) ventricular meningioma located in the right temporal lobe. The tumor had a high proliferation index Ki-67 of 40%+. The occurrence of pulmonary, thoracic, and mediastinal lymph node metastasis two years after surgery may be attributed to the malignant nature of this lesion, its strong invasiveness, and its location near the temporal lobe dura mater and venous sinus. Additionally, invasion of tumor cells into adjacent venous sinuses can lead to dissemination through the blood system. Furthermore, postoperative radiotherapy was administered to inhibit angiogenesis but inadvertently increased the likelihood of tumor cell invasion into adjacent brain tissues and subsequent distant metastasis.

In clinical practice, it is imperative to establish precise diagnostic criteria for suspected cases of metastatic CNS glioma. In 1955, Weiss [7] proposed the subsequent diagnostic criteria: (1) confirmation of a characteristic primary CNS tumor; (ii) clinical history indicating initial symptoms caused by a CNS tumor; (iii) thorough exclusion of the possibility of a primary tumor at any other site through meticulous autopsy; and (iv) consistency between CNS tumor morphology and distant metastases, with allowance for varying degrees of interstitial degeneration. Although advancements in imaging technology, molecular biology, pathologic testing, and other techniques have reduced the necessity for autopsies, the aforementioned remaining diagnostic criteria retain their clinical significance.

Conclusion

The incidence of parenchymal ventricular meningiomas in the supratentorial brain is relatively low, while their malignancy is notably high. Although intracranial tumor spread and implantation metastases in the cerebrospinal fluid can occur, extra-central nervous system metastases are exceptionally rare. Limited reports exist on such cases. In this study, we present a case of supratentorial parenchymal (WHO grade 3) ventricular meningioma with multiple metastases in the lungs, thorax, and mediastinal lymph nodes that developed post-surgery. After thoroughly reviewing and analyzing the pertinent literature, we postulate that the observed phenomenon may be attributed to several factors: the tumor's substantial size, its proximity to both the dura mater and venous sinuses, and the craniotomy procedure potentially facilitating tumor cell invasion into adjacent venous sinuses, thereby leading to hematogenous dissemination. Additionally, administration of postoperative radiotherapy likely suppressed tumor angiogenesis while concurrently increasing the likelihood of infiltration into neighboring brain tissues, mimicking distant metastasis. Although extracranial metastases from central nervous system (CNS) tumors are exceedingly rare occurrences, they should be considered within the realm of differential diagnosis. In cases where such infrequent suspected metastatic lesions arise, a comprehensive assessment encompassing an understanding of primary lesion characteristics must be undertaken in conjunction with multidisciplinary knowledge encompassing clinical history, histology evaluation, immunophenotyping analysis, and molecular genetic alterations profiling. This approach is crucial for accurate diagnosis while minimizing misdiagnosis or underdiagnosis probabilities.

Patient consent

The study involving human subject was reviewed and approved by the Medical Research Ethics Committee of Hainan General Hospital in accordance with the Helsinki Declaration. In this retrospective study, this patient's written informed consent was obtained.

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