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SNI: Unique Case Observations

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Case Report

Contralateral metastatic cerebellopontine angle glioblastoma: A rare manifestation

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ABSTRACT

Background: Glioblastoma is the most common glioma presenting within adults with an incidence of 10 per 100,000 people globally. These are mostly supratentorial tumors with rare cases of extra-axial spread. Even rarer is the presentation of glioblastoma within the cerebellopontine angle (CPA). Here, we present a case of a previously resected and irradiated glioblastoma metastasizing from the right temporal lobe region to the contralateral CPA.

Case Description: A 24-year-old female who previously underwent surgery and concurrent chemoradiotherapy for a right temporal glioblastoma in August 2020, presented to us 6 months later with headaches, vomiting, and dizziness for the past 6 days. She had left-sided dysmetria on examination. MRI of the brain showed an extra-axial, heterogeneously enhancing lesion within the left CPA. The patient subsequently underwent a left retrosigmoid craniotomy and maximum safe resection of the lesion. Histopathology reported the lesion as a glioblastoma.

Conclusion: Glioblastoma within the CPA is rarely reported within the literature. To date, our case is the first instance of an extra-axial contralateral metastasis of glioblastoma.

Keywords: Cerebellopontine angle, Extra axial, Glioblastoma, Infratentorial, Metastasis

INTRODUCTION

Glioblastomas are WHO grade IV neoplasms that account for 15-20% of all intracranial tumors and approximately 50 percent of all adult gliomas.^[10] Incidence of glioblastoma is estimated to be around 10 per 100,000 people globally and is most commonly found in the supratentorial region.^[3] The highest incidence is found in the frontal and temporal lobes. However, the presentation of glioblastoma as an extra-axial mass is rarely encountered with a few cases described in the cerebellopontine angle (CPA).[7] An extra-axial mass within the CPA is most likely a vestibular schwannoma, meningioma or an epidermoid, with only a handful of other differentials.[6]

Previous reviews have found only ten cases of primary glioblastoma extending into the CPA that were reported. [8] Here, we describe the case of a patient who presented 6 months after gross total resection and adjuvant therapy for a right temporal glioblastoma, with new-onset symptoms secondary to a mass in the left CPA. This was later confirmed to be a metastatic glioblastoma.

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CASE PRESENTATION

A 24-year-old right-handed female initially presented to our center in August 2020 with seizures for the past 1 week. Her MRI brain scan showed a right temporal heterogeneous lesion with a peripheral rim of contrast enhancement. Due to her symptoms and the suspicion this lesion was neoplastic, she underwent a right temporal lobectomy and gross total resection of the lesion [Figure 1]. The histopathology report indicated the lesion was a glioblastoma (WHO Grade IV) and p53 positive [Figure 2]. The patient subsequently received 30 fractions of radiation therapy with concomitant temozolomide (TMZ). She had surveillance MRI scans which showed no disease progression at the primary site [Figure 3a-c].

The patient presented to our emergency department in March 2021 with complaints of vomiting, headaches, and dizziness for the past 6 days. Her vomiting was nonprojectile, two episodes per day, and associated with frontal headache. She also had complaints of imbalance and could not walk without support. She did not have any other associated history. Current medications included two antiepileptic medications and temozolomide, 300 mg for 5 days

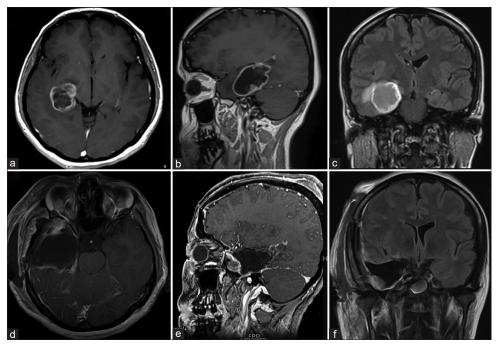


Figure 1: (a and b) Preoperative axial and sagittal T1 post contrast showing ring enhancing necrotic lesion in the right posterior temporal lobe. (c) Coronal FLAIR post-contrast showing central necrotic material. (d and e) T1 post-contrast axial and sagittal images showing gross total resection. (f) Coronal FLAIR post contrast image

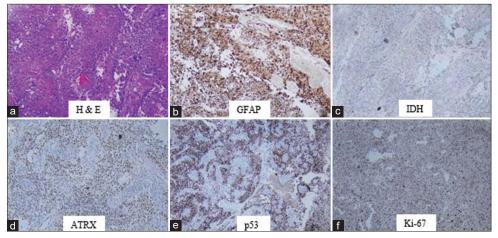


Figure 2: Histopathology from first surgery showing, (a) highly anaplastic glial cells with nuclear atypia and pleomorphism, microvascular proliferation and necrosis (b) GFAP positive, (c) IDH negative (d) ATRX retention (e) p53 positivity and (f) high Ki-67.

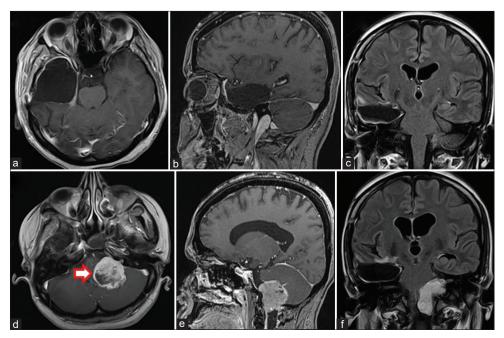


Figure 3: (a-c) Postoperative MRI Brain at 6 months post-surgery showing no evidence of disease recurrence. The patient had received CCRT prior. (d-f) MRI Brain with contrast after symptom development. (d and e) T1 post-contrast axial and sagittal images demonstrating an extra-axial CPA lesion (red arrow) with extension into the internal acoustic meatus. (f) FLAIR coronal post contrast imaging demonstrating extent to the foramen magnum.

every month, as prescribed by her primary oncologist. On examination, she was awake, alert, and oriented. Her pupils were bilaterally equal and reactive to light, and vision and speech were normal. She was found to be neurologically intact except for a mild left-sided lower motor neuron-type facial weakness. Her motor examination was normal. She had mild left-sided dysmetria and her gait could not be assessed due to severe dizziness. Her systemic examination was otherwise unremarkable. Preliminary lab investigations yielded no abnormalities.

MRI of the brain with contrast in this admission showed an extra-axial, abnormal signal intensity lesion in the left CPA [Figure 3d-f]. The lesion showed heterogeneous postcontrast enhancement and measured $3.4 \times 3.2 \times 3.2$ cm. It was causing significant compression of the medulla extending up to the cervicomedullary junction and effacement of the fourth ventricle resulting in mild proximal hydrocephalus. Contrast enhancement was seen to extend into the internal auditory canal. Of note, there was no recurrence at the primary site that had been previously operated on and given radiotherapy.

She underwent a left retrosigmoid craniotomy and resection of the lesion. Intraoperative findings showed a necrotic, soft, vascular lesion with matted lower cranial nerves passing through the bulk of the tumor. Maximum safe resection was done. Postoperative MRI with contrast showed near-total excision with a small residual. The patient was discharged from the hospital after 6 days, with mild cranial nerve deficits that were gradually improving. Final histopathology report showed a high-grade glial neoplasm with features favoring glioblastoma (WHO Grade IV) [Figure 4]. After discussion in our multi-disciplinary neuro-oncology tumor board meeting, the patient was advised to undergo conventional radiation therapy at this site as this area had not previously been irradiated and to continue with TMZ.

It was later decided by our radiation oncologists that radiation at this site near the brainstem carried significant risks and radiosurgery was advised along with intrathecal chemotherapy with a vascular endothelial growth factor inhibitor (Avastin).

DISCUSSION

Our case is a unique presentation of glioblastoma due to two reasons: glioblastoma in the CPA is a clinical rarity with few reported cases. Second, the presentation of glioblastoma metastasis is also uncommon within the literature, making the discussion of such a case necessary.

Ten cases of primary glioblastoma presenting in CPA have been previously published.^[7,8,10] Of these, six cases were intra-axial tumors originating from the brainstem or cerebellum with exophytic growth into the CPA, and three were primary extra-axial CPA glioblastoma based on cranial nerve VIII.[8] Due to internal auditory canal enhancement, the MRI findings were initially suggestive of a vestibular schwannoma, an issue that has been seen previously in the

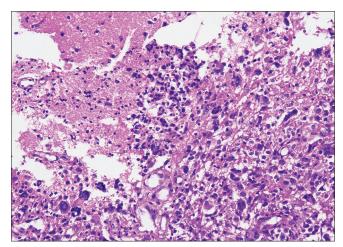


Figure 4: Photomicrograph from second surgery demonstrating highly anaplastic glial cells with nuclear atypia and microvascular proliferation and areas of necrosis.

literature regarding glioblastoma in the CPA.[9] Preoperative diagnosis is difficult in such a case, particularly as there was no recurrence at the primary site.

The other unique feature of our report is metastasis occurring to the contralateral CPA, with no local recurrence at the primary lesion site. Recurrence of glioblastoma is almost always locally with rare distant intracranial metastasis. Gliomas metastasize through the CSF channels, yet highly malignant glioblastoma metastases are rarely seen. This is likely due to the early demise of patients from the primary disease before metastatic disease has manifested. Leptomeningeal metastasis has been seen in less than 2 percent of patients with glioblastoma, however, microscopic metastases have been detected in 6 percent of supratentorial lesions.[8]

Previous studies have looked at risk factors for metastasis of CNS tumors to the CPA, which increase the risk of dissemination from the CSF causing leptomeningeal involvement, therefore further spread and disease.[11] Primary CNS tumors can spread through hematogenous pathways, direct extension of the tumor to adjacent regions, iatrogenic factors, and leptomeningeal spread due to surgically opening the ventricles or tumor abutting ventricles or basal cisterns.^[5] However, there was no disease progression at the primary site noted on any subsequent scans. Instead, the lesion is seen recurring on the contralateral side in an extra-axial space, which is unusual and previously never seen.

Intracranial metastasis of glioblastoma is rarely reported within the literature. The spread of cancer cells via the cerebrospinal fluid can potentially then create new foci of glioblastoma at a distant location, as seen in previous case reports.[1] The incidence of extracranial metastases of glioblastoma is also low (2%), with bone (38%), lymph nodes (37%), lungs (32%), and liver (18%) being the most common sites.^[4]

The management of metastatic glioblastoma includes repeat surgery, radiation (conventional and SRS), systemic chemotherapy, or combination therapies of the aforementioned. Particularly, radiosurgery has become an increasingly used salvage treatment modality for glioblastoma. This allows reduced treatment-related toxicity associated with other radiation oncology therapies and may provide benefit for cases of previously irradiated glioblastoma.^[2] However, the best management for CPA glioblastoma has not been established due to its clinical

CONCLUSION

Glioblastoma within the CPA is rare within the literature. Our case shows the possibility for extra-axial and contralateral metastasis from a previously irradiated glioblastoma, which has not been reported before.

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Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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