Early dural metastasis from a case of glioblastoma with primitive neuroectodermal differentiation: A case report and literature review

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ABSTRACT

Glioblastoma with a primitive neuroectodermal (PNET) variant is a rare primary parenchymal tumor. Only a few cases of extraparenchymal metastasis are reported in world literature. Although the overall survival duration of glioblastoma multiforme (GBM) with primitive neuroectodermal tumor (PNET) variety may be prolonged in comparison to classical glioblastoma, the metastatic trend is completely different, and the prognosis is worse. We report an early dural metastasis of pure PNET component appearing in a case of primary glioblastoma with PNET variant. The lesson learned from this case is to look for early craniospinal metastasis in GBM patient with PNET component, even after completion of adjuvant radiochemotherapy.

1. Introduction

Glioblastoma with PNET is the new emerging variety of malignant tumor as reported in the literature. The prognosis is as dismal as glioblastoma. Standard radiotherapy and chemotherapy are the mode of management after gross total resection of the tumor. However, the overall survival depends on craniospinal metastasis or recurrence of the tumor. The incidence of metastasis is less than 2% [10] for glioblastoma, and for glioblastoma with PNET variety, only a few cases of metastasis were reported in he literature. We report a primary case of glioblastoma with PNET component with multiple early dural metastases of pure PNET component.

2. Case report

A 59-year-old female presented with left facial weakness for one month, followed by left side weakness of upper and lower limbs. There was no history of seizure, headache, or visual blurring. Her neurological examination was remarkable for a left-sided facial paresis and hemiparesis (power: 3/5). Computed tomography (CT) of the head showed right side frontotemporal mixed density mass with mass effect and midline shift of 5 mm. The magnetic resonance imaging (MRI) revealed a heterogeneous tumor (53 mm × 44 mm) affecting the right temporal lobe with solid and cystic components associated with moderate vasogenic edema (Fig. 1). She underwent right side frontoparietal craniotomy and gross total decompression of the tumor. The histopathology report was glioblastoma multiforme with PNET variant (Fig. 2). IDH 1, MGMT and 1p/19q studies were negative for this tumor. The immunohistochemistry report showed strong positive GFAP stain in the glial part, weak synaptophysin positivity in the neuroblastic part, and strong CD-56 positivity in both the glial and neuroblastic parts. Immediately after surgery, her left hand function returned to normal. She received radiation therapy for six weeks along with chemotherapy (temozolamide) for the same duration. She was apparently improving in motor power.

However, after 2 months, she presented with headache and vomiting. Repeat imaging showed multiple dural-based mass lesions enhanced with contrast (Fig. 3). The recurrence was outside of the irradiation field. She underwent repeat surgery and decompression of the mass lesion. Her biopsy reported a metastatic primitive neuroectodermal tumor with no malignant glial component. The biopsy slide showed fragments of a neoplasm composed of large anaplastic polygonal cells with pleomorphic, hyperchromatic nuclei, irregular nuclear borders, finely granular chromatin, prominent nucleoli, nuclear molding, and scant granular, eosinophilic cytoplasm. The tumor cells showed cell wrapping. Numerous mitoses were observed. The tumor invaded dense connective tissue and brain parenchyma. Immunohistochemistry stain confirmed the neuroblastic tumor with CD56, synaptophysin positivity and negative for Chromogranin, GFAP, AE1/3, CAM5.2, CK7, CK20, EMA, TTF1, S-100, HMB45, MART-1, and CD31 (Fig. 4). In the postoperative period, she recovered well and was discharged home. But one month after the second surgery, she began to deteriorate and was moved to hospice care.

3. Discussion

Glioblastoma multiforme has three main variants: classic GBM, gliosarcoma and giant cell GBM. The other emerging variants are GBM-PNET, GBM with oligodendroglioma, and fibrillary/epithelial GBM. Perry et al. [9] extensively discussed the clinicopathological characteristics and molecular basis of GBM-PNET. We have discussed the different patterns of metastasis as well as the recurrence rate of classic GBM and its entire variant. The primary cause of treatment failure for GBM was local recurrence. However, Stark et al. [10] reported a frequency of cerebrospinal fluid (CSF) metastasis of 1.1% in adult glioblastoma. The reason for more CSF dissemination is the resistance of the glial cell towards hematological spread as well as no lymphatic connection in the central nervous system. Furthermore, Aghakhani et al. [1] reported 6% supratentorial metastasis and 60% infratentorial metastasis related with GBM in an autopsy study. Subpial spread is the other route of dissemination into subdural space or dural metastasis. Lettau et al. [8] reported a case of dural metastasis of glioblastoma seven months after primary surgery. The median time duration between diagnosis of glioblastoma and subarachnoid dissemination was eight months to fourteen months [5].

Infiltration of dura or surrounding bone occurs frequently with gliosarcoma. However, extracranial metastasis is common with gliosarcoma, with an incidence of almost 11% [3].

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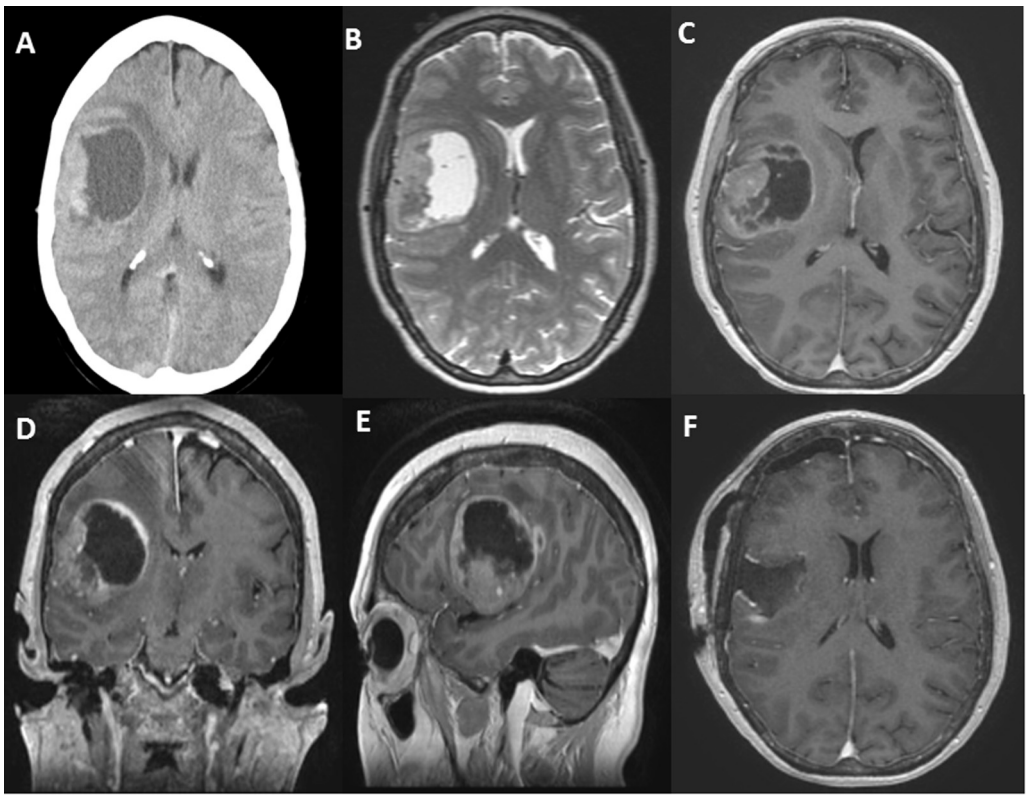

Fig. 1. (A) Head CT showed right side posterior frontal mass with solid and cystic component, (B) MRI (T2W) showed right posterior frontal heterogeneous mass with solid part at periphery, (C) MRI Contrast (Axial) showed enhancement of solid part, (D) MRI Contrast (Coronal) showed tumor involving the inferior frontal gyrus, (E) MRI (Sagittal) showed solitary lesion, (F) MRI Post-op (axial) gross total resection of tumor.

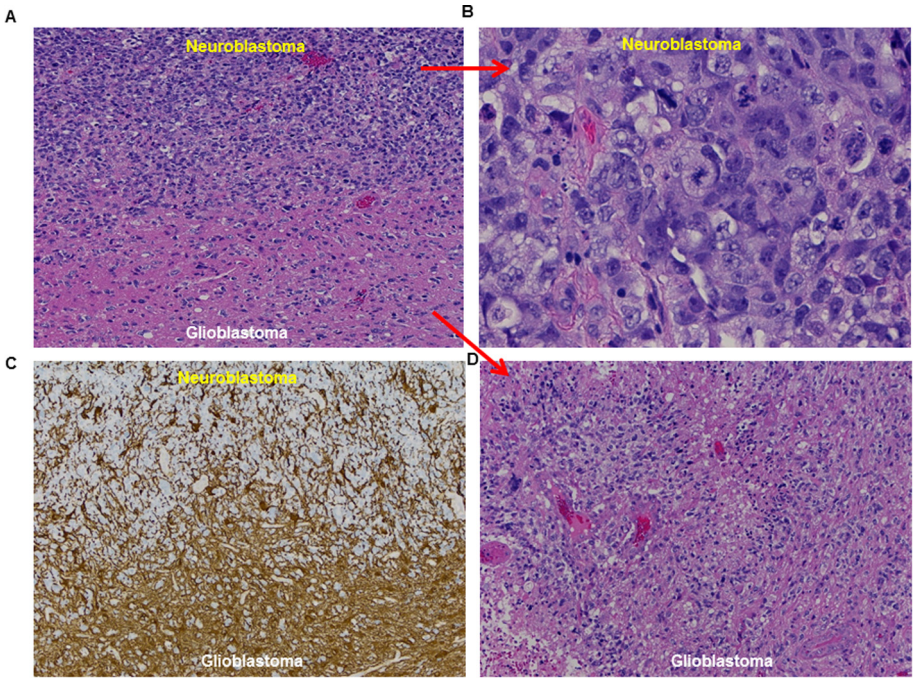

Fig. 2. A (H and E stained) and C (Glial fibrillary acidic protein, GFAP stained) show the concurrence of PNET and glioblastoma. B and D (Hand E stained) represent the magnification of the neuroblastic and glial portion of the primary tumor.

Beaumont et al. [3] reported a case of gliosarcoma with multiple extracranial metastases 19 months after diagnosis. However, Witwer et al. [14] reported a case of cervical cord metastasis one month after primary diagnosis. Wharton et al. [13] diagnosed extraneural metastasis from the gliosarcoma with areas of primitive neuroepithelial differentiation arising from temporal lobe in a patient who presented five months after primary diagnosis. Ammerman et al. [2] reported a case of giant cell GBM with spinal