Unusual Location of Uncommon Tumor: one case report of intraventricular gliosarcoma

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ABSTRACT

Gliosarcoma is an unusual malignant neoplasm of the central nervous system. It shares the same clinical presentation, epidemiology and prognosis with glioblastoma multiforme. It is usually located within intraparenchymal compartment, especially in the supratentorial area adjacent to the cortical surface. Intraventricular location is extremely rare for gliosarcoma. We report a rare case of intraventricular gliosarcoma. The features of the tumor, as observed by computed tomography (CT), magnetic resonance imaging (MRI) are described, along with a review of the literature.

CASE REPORT

This 37 year-old male had suffered from headaches for two to three months, associated with dizziness, occasional fainting, poor sleep and appetite, recent memory deficit, ever vomiting, personality change and slurred speech. At our emergency room, the Glasgow coma scale (GCS) revealed E4V4M6, and impaired JOMAC mental status examination. Left facial palsy and right extremities muscle power decrease were also implied. Computed tomography (CT) of the brain without contrast medium revealed two isodense to slightly hyperdense mass lesions in the left paraventricular and suprasellar regions with hydrocephalus (Fig. 1). Magnetic resonance image (MRI) with gadolinium (Gd) showed a heterogeneously enhancing mass in the left lateral ventricle surface with brain parenchyma extension, about 4.7 × 3.8 × 4.0 cm in size. The lesion showed low signal intensity on T1-weighted images (T1WI), slight high signal intensity on T2-weighted images (T2WI), and mild high signal intensity on diffusion weighted images (DWI), and foci of dark signal on susceptibility weighted images (SWI). There was another heterogeneously enhancing nodule in the suprasellar region, about 1.8 × 1.6 cm in size, sharing the same image characteristic patterns with the ventricular lesion (Fig. 2). The differential diagnosis included high-grade glioma, germ cell tumor, ependymoma, lymphoma, metastasis or...
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others. These lesions caused optic chiasm compression and midline shift to the right side. There was abnormal contrast enhancement around the left lateral ventricle, surface of midbrain and pons on T2FLAIR post gadolinium images, which is indicative of leptomeningeal metastasis. Then the patient received craniotomy and stereotactic biopsy. The pathology showed a hypercellular tumor with hyperchromatic rounded cells intermingled with bizarre spindle cells (Fig. 3a). Geographic necrosis and endothelial proliferation were also appreciated. Immunohistochemically, some of the tumor cells were positive for GFAP (Fig. 3b), whereas the spindle cell elements were negative. The vimentin was focally positive and focal increased reticulin fibers was noted (Fig. 3c, 3d). The cytology of cerebrospinal fluid (CSF) yielded malignant cells. In conclusion, the final diagnosis was gliosarcoma (WHO grade IV). Follow-up cervical spine MRI with contrast medium showed leptomeningeal tumor spreading along the pontine surface and C1-3 spinal cord surface (Fig 4.). Afterwards, this patient only received palliative radiotherapy. No further chemotherapy was given due to the poor socioeconomic status and personal reasons. After one year-follow-up, this patient is still alive, living in the nursing home. No further treatment or imaging was pursued.

DISCUSSION

One-tenth of all central nervous system (CNS) tumors are present within or adjacent to the ventricular system. These neoplasms include ependymoma, subependymal giant cell astrocytoma, subependymoma, and, rarely, glioblastoma. Gliosarcoma is a rare CNS malignant tumor with biphasic pattern of unequivocal sarcomatous and glial cell populations. Most of the sarcomatous components appear as fibrosarcoma or malignant fibrous histiocytoma [4]. It was first described by Stroebe in 1895 and officially accepted by WHO in the modern time [5]. Based on the clinical and epidemiological and pathologic aspects, gliosarcoma is regarded as a variant of glioblastoma multiforme and accounts for about 2-8% of all CNS tumors [6]. It usually occurs in the population aged 40-60 years with male predilection (1.8:1) [7]. Like glioblastoma, it is mainly a lobar tumor, usually located in the supratentorial region, and has the propensity for temporal, frontal, parietal, and occipital lobes in the order of preference [8], often surfacing and abutting to the meninges [9] and sometimes mimicking meningioma [1]. Intraventricular location is extremely rare and up to now, only four cases have been reported [1, 3]. The first case arose from malignant transformation of a preexisting ependymoma. The second case involved the frontal lobe with extension into the lateral ventricle [3]. The third case was an exclusively lateral ventricular tumor probably arising from the intraventricular septum and blocking the CSF pathway [3]. The fourth case was a lobulated mass within the temporal horn of the right lateral ventricle [1]. The image patterns of gliosarcoma often show low signal on T1WI and high signal on T2WI and heterogeneous enhancement post Gd. Sometimes it may present tumor necrosis, intratumoral hemorrhage, and diffusion restrictive appearance on DWI. MR spectroscopy shows elevated choline, decreased NAA and may show lactate or lipid peak, the same pattern as high-grade glioma. The conventional image pattern is like the four cases of intraventricular gliosarcoma, but the DWI and SWI pictures are not mentioned in the previous cases. We did not perform MR spectroscopy on this patient due to his rapid transfer to the other campus of our hospital after the intracranial

Figure 1

Figure 1. a. b. CT of the brain without contrast medium revealed two isodense to slightly hyperdense mass lesions over left paraventricular and suprasellar regions with hydrocephalus.
Figure 2. a. axial T1WI; b. axial T2WI; c. DWI (b value=1000); d. ADC; e. SWI; f. and g. post Gd axial T1WI; h. post Gd axial T2FLAIR (Fluid attenuated inversion recovery). One mass lesion over left lateral ventricle and paraventricular region showed low signal on T1WI (a) and mild high signal on T2WI (b). The DWI (c) with corresponding ADC (d) revealed mild diffusion restrictive pattern. There are dark signal foci on SWI (e) within the mass lesion. There was heterogeneous enhancement of these lesions over left paraventricular region and suprasellar region on post Gd T1WI (f, g). The post Gd T2FLAIR (h) image showed abnormal enhancement over surface of pons, suggesting leptomeningeal carcinomatosis.
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Susceptibility-weighted imaging (SWI) is a high-spatial-resolution 3D gradient-echo MR imaging technique with phase post-processing that accentuates the paramagnetic properties of blood products such as deoxy-hemoglobin, intracellular methemoglobin, and hemosiderin. It is particularly useful for detecting intravascular venous deoxygenated blood as well as extravascular blood products. It is also quite sensitive to the presence of other substances such as iron, some forms of calcification, and air. In our case, there is SWI dark signal within the tumor, indicating that previous tumor bleeding with hemosiderin deposition is favored than other etiology. It may imply high malignant potential of this mass. Intracranial germ cell tumor is often referred to as germinoma or seminoma. It is usually located within the midline third ventricle/suprasellar region or pineal region, so-called bifocal germinoma. It is often in the male population aged 20 years old or younger. The tumor marker serum hCG is usually elevated, and it could be helpful to our diagnosis. Metastasis could be any form of image appearance. However, this patient did not have known malignancy history. So, metastasis is listed in the last differential diagnosis. Ependymoma is usually detected within the infratentorial area, with the age younger than 10 years old. It frequently expands through the foramen of Luschka into the cerebellopontine angle or the foramen of Magendie. It could be also located in the supratentorial area.  

Figure 3. a. H &E stain; b. GFAP; c. reticulum stain; d. vimentin stain. The pathology showed a hypercellular tumor with hyperchromatic rounded glial cells intermingled with bizarre spindle cells (a). Immunohistochemically, glial cell elements of the tumor cells were positive for GFAP (b), while the spindle cell elements were positive in the stain of vimentin and reticulin fibers (c, d).
area, with the juxtaventricular region most often seen. CSF dissemination seeding is often observed. Primary CNS lymphoma usually presents well enhancing lesion within basal ganglia and periventricular white matter. It could show hyperdensity on precontrast CT and diffusion restrictive appearance on MRI, a character also detected in our case. So the differential diagnosis of ependymoma and CNS lymphoma could not be excluded completely. Pathological diagnosis, however, is crucial for definite diagnosis of gliosarcoma. The histopathological examination indicates that the sarcomatous component is rich in reticulin and vimentin and glial component is positive for glial fibrillary acidic protein (GFAP) and S-100 protein [10].

There are many hypotheses of the histogenesis of these biphasic tumor patterns. With the development of molecular biochemistry, now it is believed to be derived from the monoclonal origin of the glial and sarcomatous components [3]. Gliosarcoma shares the same poor prognosis with glioblastoma. The treatment option includes aggressive surgical resection, external beam radiotherapy and chemotherapeutic agents such as temozolomide or nitrosourea.

Although the well-circumscribed and surfaced nature facilitates radical resection, the treatment response to multimodality therapy is still poor as GBM [1]. The median survival rate has been reported to be 6-14 months [11]. Moreover, gliosarcoma is thought to metastasize more often than GBM and frequently undergoes extracranial metastasis via hematogeneous dissemination, mainly to the liver and the lung [9]. The sarcomatous component seems to have a higher propensity for such dissemination, often being the only component (without glial cells) in the metastatic lesions.

REFERENCE