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Case report

Transformation of intracranial anaplastic astrocytoma associated with neurofibromatosis type I into gliosarcoma: Case report

Jin-Hwan Cheong[a], Choong-Hyun Kim[a,*], Jae-Min Kim[a], Young-Ha Oh[b]

[a] Department of Neurosurgery, Hanyang University College of Medicine, Seoul, Republic of Korea
[b] Department of Pathology, Hanyang University College of Medicine, Seoul, Republic of Korea

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ABSTRACT

Gliosarcoma is an uncommon malignant brain tumor composed of distinct sarcomatous and malignant glial cell elements. These tumors are defined as a variant of glioblastoma, and it can be developed by progression of the malignant glial cell tumors or primary tumors. We report a rare case with gliosarcomatous recurrence of anaplastic astrocytoma with neurofibromatosis type 1 (NF-1) followed by chemoradiation therapy. A 26-year-old male patient was presented with headache. Five years before admission, he had been diagnosed with NF-1. Magnetic resonance imaging (MRI) showed a well-demarcated, enhanced lesion in the right frontal lobe and multiple enhanced lesions in the scalp, lower cervical, thoracic, and upper lumbar regions. He underwent an osteoplastic craniotomy with total tumor resection. Histopathology of the tumor showed findings corresponding with anaplastic astrocytoma. He was followed by radiotherapy and chemotherapy postoperatively. A month later, his spinal lesion was also resected and confirmed pathologically as plexiform neurofibroma. The subsequent follow-up period of 27 months was uneventful until he developed a generalized tonic-clonic seizure. MRI showed tumor recurrence in the original site of the tumor. Re-exploration was carried out. Pathological examination displayed a biphasic pattern of the glial and sarcomatous components suggesting gliosarcoma.

1. Introduction

Gliosarcoma is uncommon primary tumors of the central nervous system (CNS) characterized by an admixture of histologically distinct glial and sarcomatous components. They were first described in 1895 by Stoebe [1]. The sarcomatous component resembles fibrosarcoma or malignant fibrous histiocytoma [2]. Although the glial component in the majority of cases of gliosarcoma is malignant astrocytoma, it comprises several kinds of glial cells tumors including oligodendroglioma or ependymoma [3]. In the 2007 World Health Organization (WHO) classification of tumors of the central nervous system (CNS), gliosarcoma is considered a subtype of glioblastoma [4]. This conclusion is supported by finding identical genetic alterations in both tumor elements [5].

On the other hand, neurofibromatosis type 1 (NF-1) is a relatively common hereditary syndrome accompanying CNS neoplasms, that are mostly pilocytic astrocytomas [6]. Gliosarcoma with NF-1 has rarely been reported. To date, 4 cases have been reported in the English written literatures [7–10]. Moreover, there are no reports with gliosarcomatous recurrence that have been originated from anaplastic astrocytoma associated with NF-1. Some cases of gliosarcoma occur spontaneously, and rarely, radiotherapy for various CNS malignancies also induces these tumors [11,12].

We present a case with gliosarcomatous recurrence of the anaplastic astrocytoma accompanying NF-1 after surgical resection and subsequent chemoradiation.

2. Case report

A 26-year-old male patient was presented with headache over a year. Five years before the current presentation, he had been diagnosed with NF-1. On neurological examination, he was alert and there were no deficits in sensory or motor functions. Ophthalmological and cranial nerve examinations also showed no significant abnormalities.

Magnetic resonance imaging (MRI) revealed a relatively well-demarcated, heterogeneous enhancement measuring 1.8 cm × 2.0 cm × 2.1 cm and an accompanying peritumoral edema in the right frontal lobe close to the central sulcus (Fig. 1A). The presumptive diagnosis was malignant glioma, lymphoma or metastatic brain tumor. Spinal MRI also demonstrated multiple lesions in the intraspinal and subcutaneous regions, and showed a huge multi-lobulated lesion in the upper lumbar region (Fig. 1B).

* Corresponding author at: Department of Neurosurgery, Hanyang University Guri Hospital, 249–1 Gyomun-dong, Guri, 471-701, Republic of Korea. Tel.: +82 31 560 2322; fax: +82 31 560 2327.
E-mail address: kch5142@hanyang.ac (C.-H. Kim).

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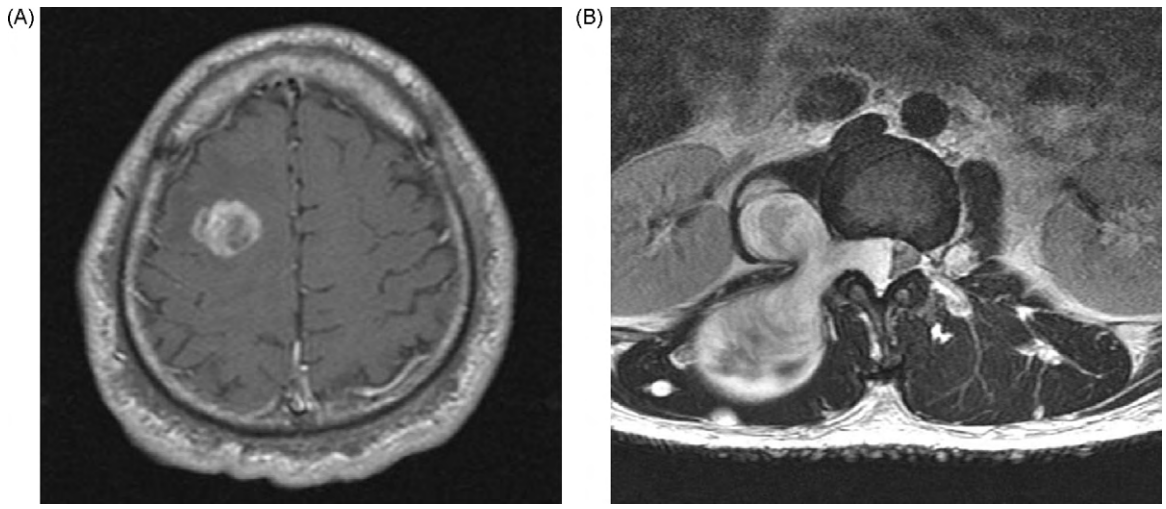

Fig. 1. (A) Postcontrast T1W axial magnetic resonance image (MRI) demonstrated a well-enhanced lesion with central hypointensity accompanying peritumoral edema in the right frontal lobe. (B) Spinal T2W axial MRI showed a lobulated lesion in the upper lumbar region.

He underwent a right frontotemporoparietal osteoplastic craniotomy and radical tumor removal was performed. The histopathology of the tumor showed relatively heterogenous in cellularity and morphologic features. It was composed of highly pleomorphic and large tumor cells with abundant cytoplasm and cytoplasmic processes. Their nuclei were pleomorphic with prominent nucleoli (Fig. 2A and B). In some areas, multinucleated tumor giant cells and spindle cells were also observed. There was no definite microvascular proliferation or tumor cell necrosis. The tumor cells including spindle cells and multinucleated giant cells revealed glial fibrillary acidic protein (GFAP) immunoreactivity and reticulin-free stroma, which were compatible with anaplastic astrocytoma (Fig. 2C and D). Ki-67 labeling index was increased up to 10%.

A month after the craniotomy, an intradural extramedullary mass in the upper lumbar region was also resected by osteoplastic laminotomy. The surgical specimen revealed abundant intervening spindle cells, wavy collagen bundles, and increased

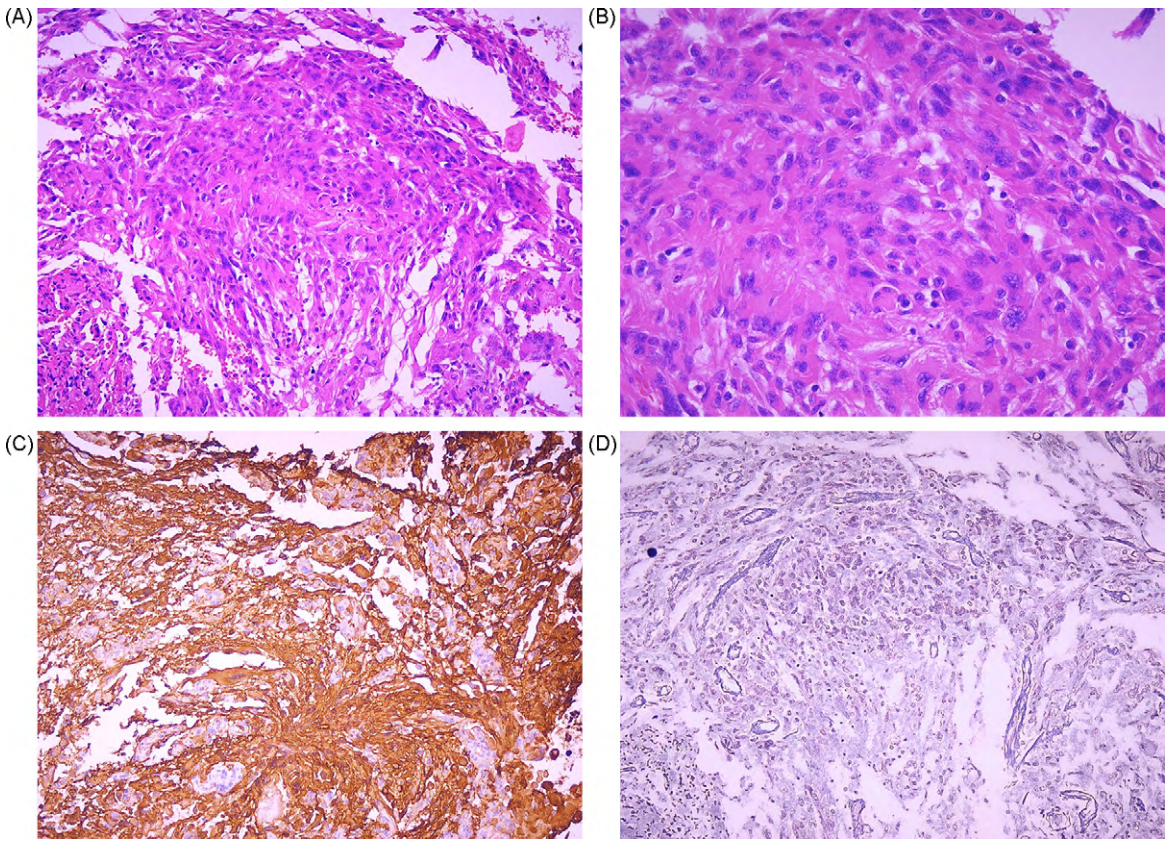

Fig. 2. Photomicrographs of the tumor. (A) Medium power view shows haphazardly arranged large pleomorphic tumor cells with some spindle cells (H&E stain, ×200). (B) High power view shows variable sized vesicular nuclei and prominent nucleoli with abundant eosinophilic cytoplasm and fibrillary processes (H&E stain, ×400). (C) Immunohistochemical stain for GFAP reveals diffuse and strong positivity (GFAP stain, ×200). (D) Most tumor cells were not immunoreactive to reticulin (Reticulin stain, ×200).