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Case Report

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Cerebellar Anaplastic Oligodendroglioma Presenting with Hemorrhage

The authors describe an anaplastic oligodendroglioma of the cerebellum which is distinctly uncommon. The patient presented with sudden onset of decreased consciousness associated with hemorrhage in the cerebellum, which appeared to origin from a vascular malformation or a tumor on a initial computed tomography (CT) scan. Subsequent magnetic resonance (MR) imaging suggested a high grade glioma with abundant vascularity in the right cerebellum. The histological examination revealed the findings compatible with those of an anaplastic oligodendroglioma. A complicated clinical course had led him to a poor outcome.

KEY WORDS : Anaplastic oligodendroglioma · Cerebellum · Hemorrhage.

INTRODUCTION

Oligodendrogliomas represent approximately 4-7% of all primary intracranial tumors and the third most common glioma following glioblastoma and astrocytoma[2]. They are found principally in adults and primarily in the cerebral hemisphere, where the frontal lobes are greatly favored. They rarely appear in the cerebellum and have been anecdotally reported[6,10]. Although magnetic resonance (MR) imaging is the diagnostic choice, as many as 60% of oligodendrogliomas calcify and the use of computed tomography (CT) with its superior visualization of calcium is considerably valuable. Russel and Rubinstein[12] mentioned that spontaneous hemorrhage seemed to have a special propensity in oligodendrogliomas. Histologically, perinuclear halos in the cytoplasm are characteristic. Anaplastic oligodendroglioma may present de novo or evolve from a well-differentiated oligodendroglioma, of which the diagnosis was made based on histological criteria including the presence of oligdendroglial morphology, hypercellularity, nuclear pleomorphism and necrotic activity[4]. The treatment of anaplastic oligodendrogliomas consists of maximum feasible surgery followed by chemotherapy and radiation therapy.

The authors report a case of cerebellar anaplastic oligodendroglioma presenting with tumor bleeding.

CASE REPORT

History and examination

The 37-year-old man was quite well until he presented with a sudden onset of decreased consciousness. The CT scan obtained at a local clinic demonstrated a 4.9×3.4 cm hematoma in the right cerebellar hemisphere with surrounding edema that caused a non-communicating hydrocephalus. There was also a enhancing lesion abutting the hematoma which suggested a vascular malformation or a tumor (Fig. 1). After emergency ventricular drainage, he was transferred to our hospital.

On physical and neurological examination, he was in deep-drowsy state was hardly responsive to the verbal order. The cranial nerves were intact and overall motor activity was depressed. Brain MR images showed an 5.4×3.3×3.8 cm sized irregular hematoma (acute to subacute) and multiple small vascular signal void structures around the hematoma in the right cerebellar hemisphere (Fig. 2).

Operation and postoperative course

The patient underwent a suboccipital craniectomy and hematoma removal. Biopsy was performed at the several sites around the hematoma, which demonstrated normal cerebellar

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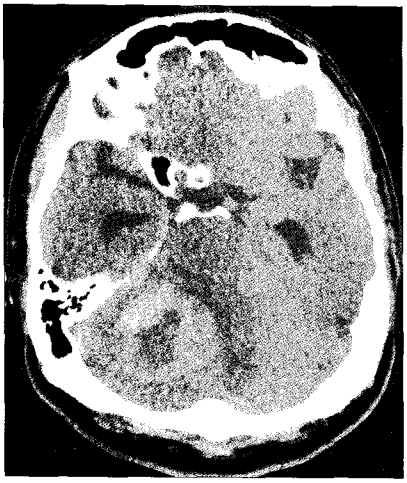

Fig. 1. Initial computed tomography scan shows 4.9×3.4 cm hematoma in the right cerebellar hemisphere with surrounding edema which caused a non-communicating hydrocephalus. An enhancing lesion abutting the hematoma suggests a vascular malformation or a tumor.

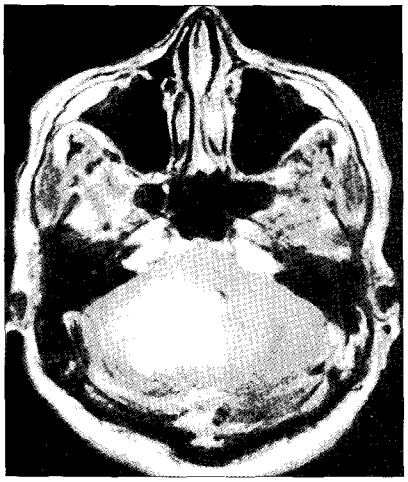

Fig. 2. Enhanced brain magnetic resonance image shows 5.4×3.3×3.8 cm hematoma (acute to subacute) in the right cerebellar hemisphere.

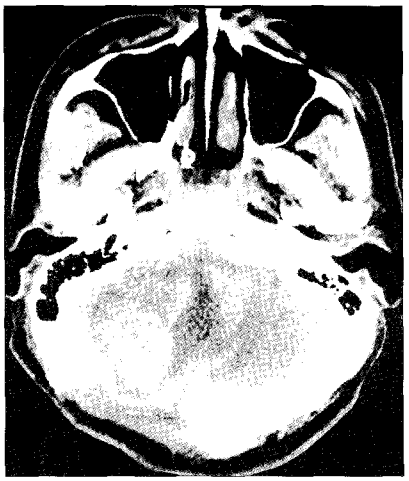

Fig. 3. Rebleeding and an irregulary enhancing heterogeneous mass around the hematoma.

tissues. Postoperatively, he gained consciousness to be nearly alert. He was relatively well until intense headache had developed. The CT scan showed marked ventricular dilatation secondary to compression of the fourth ventricle by cerebellar edema.

After first operation, a ventriculo-peritoneal shunt was carried out in 45 days, which made his headache relieve. Four days later, he suddenly became drowsy. The CT scan revealed rebleeding and a irregularly enhancing heterogeneous mass around the hematoma (Fig. 3).

He subsequently underwent gross total resection of the mass after hematoma removal. However, he did not make a good postoperative recovery. Histological examination of the mass revealed characteristic findings of an anaplastic oligodendroglioma. He had maintained a poor stationary clinical condition. Further treatments, including a posterior fossa decompression, chemotherapy and radiation therapy, were denied by his family. He died afterward.

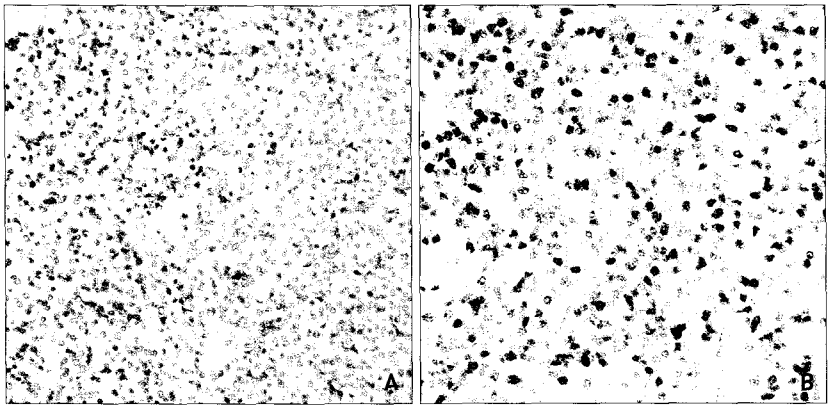

Fig. 4. A : Groups of cells, with relatively irregular round nuclei and empty-looking cytoplasm, bounded by distinct cell membrane, are demarcated by fine fibrovascular stroma. Nuclei are mildly to markedly pleomorphic, and mitoses are occasionally noted (H&E, ×400). B : Immunohistochemistry for Ki-67 shows positive nuclear staining more than 50% of tumor cells (ABC, ×400).

Histological examination

The permanent section obtained after rebleeding showed a tumor composed of relatively small, densely packed cells containing little cytoplasm. Nuclei were mildly to markedly pleomorphic, and mitoses were occasionally noted (Fig. 4A).

The tumor was found to be positive for glial fibrillary acidic protein, but negative for synaptophysin. A Ki-67 index was above 50% (Fig. 4B).

DISCUSSION

Oligodendrogliomas account for approximately 4-7% of all primary intracranial gliomas[12], predominantly occurring in the supratentorial location. They are distinctly uncommon in the posterior fossa. In a large study of 323 patients including oligodendrogliomas and anaplastic oligodendrogliomas, Ludwig, et al.[9] found that the primary location was frontal in 55%, and cerebellar in 3%. Ernest, et al.[5] reported 14 cases (7%) out of 200 cases of oligodendrogliomas ; 5 in the cerebellar vermis, 4 in the fourth ventricle, 3 in the hemisphere, and 2 in the cerebello-pontine angle. Packer, et al.[11] found 4 cases (1.6%) arising in the posterior fossa out of 249 children with primary intracranial neoplasms. In Korea, Kim, et al.[13] reported one case of anaplastric oligodendroglioma arising from pons.

The symptoms for oligodendroglioma do not reliably distinguish them from other tumors. The most common presenting symptom is seizure. Classically, patients with oligodendroglioma often experience seizure for several years prior to their diagnosis that are usually present after an apoplectic event such as hemorrhage. Recently, this situation has become less common with the increased sensitivity of CT and MR imaging, which have led to earlier diagnosis.

This patient had experienced no seizures until he presented with an intracerebellar hemorrhage, which may be attributable to location in the posterior fossa.

Brain tumors constitute the cause of spontaneous intracranial hemorrhage only in rare instances. Tumor bleeding had been reported to have a range of from 0.9% to 11.0%[7]. Of the glioma, the hemorrhage rate ranged 2.0% to 25.7%[7]. Russel and Rubinstein[12] mentioned that spontaneous hemorrhage seemed to have a special propensity in oligodendrogliomas.

Hinton, et al.[8] performed an etiological study in 84 cases diagnosed as spontaneous intracerebral or intracerebellar hemorrhage, of which 54 cases included blood clot plus brain. Seven cases (13%) denoted a brain tumor in which one was an oligodendroglioma. He thus emphasized a biopsy of adjacent brain tissue on evacuation of spontaneous intracerebral hemorrhage. Well-differentiated oligodendroglioma are composed of uniform cells with round nuclei and fine chromatic pattern with small nuclei. Perinuclear halos in the cytoplasm are characteristic, which are a result of autolysis due to delay in fixation. The halos are therefore absent in frozen sections. Accoridng to a report[1] from the Central Brain Tumor Registry of the United States, 77% of oligodendroglioma are low grade and 23% are anaplastic. An anaplastic oligodendroglioma may present de novo or evolve from a well-differentiated oligodendroglioma. Anaplastic oligodendrogliomas are more cellular with significant nuclear pleomorphism, elevated mitotic rate, vascular proliferation and often necrosis. Our case represented typical characteristics of anaplastic oligodendrogliomas being exclusively oligodendroglial. A Ki-67 labeling index may be useful in assessing tumor grade. Deckert, et al.[3] reported a good correlation between tumor grade and Ki-67 index. Anaplastic oligodendroglioma shows a significantly higher labeling index as compared to well differentiated oligodendroglioma. This case had a Ki-67 index of above 50% and followed suit. Because of the low incidence of oligodendrogliomas in the posterior fossa, there has been little reference to outcomes of the treatment. The treatment of anaplastic oligodendrogliomas consists of maximal feasible surgery, followed by chemotherapy and radiation therapy. It has been well-known that anaplastic oligodendrogliomas are uniquely chemosensitive tumors at initial diagnosis or recurrence[4]. In this patient, further treatments were recommended, but were denied from the family mermbers.

CONCLUSION

The authors report an anaplastic oligodendroglioma arising in the right cerebellum. The patient presented with suddenly decreased consciousness associated with cerebellar hemorrhage. An initial CT scan suggested hemorrhage by a vascular malformation or a tumor. The findings of subsequent MR imaging preferred a high grade glioma with abundant vascularity to a vascular malformation.

A biopsy showed a typical anaplastic oligodendroglioma, exclusively oligodendroglial. The outcome was grave following several events.

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