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Case Report

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J Korean Neurosurg Soc 42 : 53-55, 2007

Cerebellar Ganglioglioma in an Old Patient

Gangliogliomas could be found anywhere throughout the central nervous system and mainly affect children and young adults during the first three decades of life. Cerebellar gangliogliomas may be rarely found, especially in old ages. Here, we present a case of ganglioglioma of the cerebellum in an old patient. The cystic cerebellar mass was associated with calcifications, intratumoral hemorrhage without ng edema. When a cystic cerebellar mass is associated with calcifications and intratumoral hemorrhage, ganglioglioma should be included in differential diagnosis. Gangliogliomas usually have good prognoses. Radiation therapy should be deferred even in subtotally removed cases.

KEY WORDS : Ganglioglioma · Cerebellum · Cystic cerebellar mass · Calcification · Intratumoral hemorrhage.

INTRODUCTION

Gangliogliomas are unusual tumors of the central nervous system comprised of variable proportions of ganglion cells and glial cells[7]. Most tumors have been reported to occur supratentorially, most commonly in temporal lobes presenting with long-standing intractable seizures[3]. Gangliogliomas occur rarely in the cerebellum (7-10% of intracranial gangliogliomas)[11]. In Korean literature, only one case has been reported previously[14].

Cerebellar ganglioglioma most commonly manifest during childhood and adolescence by symptoms due to increased intracranial pressure[13,15]. Most tumors have cystic component with mural nodules. These clinical and radiological features are very similar to those of cystic cerebellar astrocytoma[2].

Here, we report a cerebellar gangioglioma in an old patient and discuss the characteristic clinical and radiological features.

CASE REPORT

A 68-year old man presented with a 2-month history of headache and dizziness. The patient had a history of gastric carcinoma (T3N0M0, stage III) 8 years ago and underwent total gastrectomy and esophago-jejunostomy. Subsequently, the patient received 12 cycles of chemotherapy with 5-flurouracil (5-FU) and carboplatin. Complete remission was confirmed by no recurrence in endoscopic and computed tomographic studies. Neurologic examination

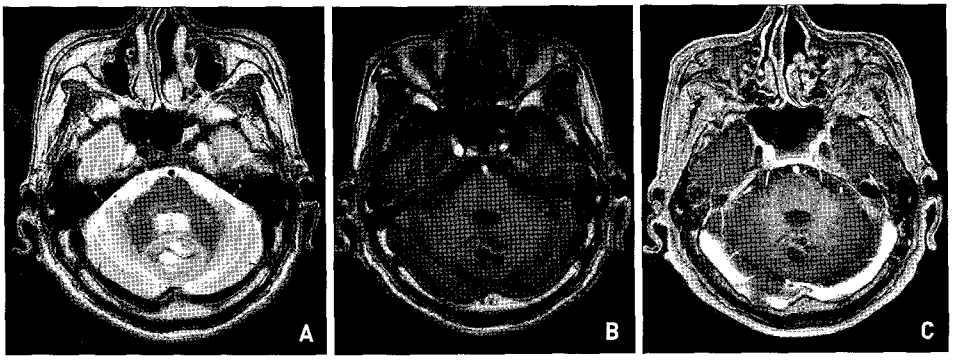

Fig. 1. The tumor being relatively well defined and heterogeneous signal intensity on T2-weighted image (A), high signal intensity on T1-weighted image (B), and only minimal marginal enhancement on postcontrast image (C).

• Received : March 28, 2007
• Accepted : June 1, 2007
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on admission revealed no neurologic deficits. Magnetic resonance imaging showed a cystic mass in the cerebellar vermis with intratumoral hemorrhage. The tumor was relatively well defined and heterogeneous and showed heterogeneous signal intensity on T2-weighted images (Fig. 1A), high signal intensity on T1-weighted images (Fig. 1B), and only minimal marginal enhancement on postcontrast images (Fig. 1C). There was no surrounding edema and ventricular compression. The most notable differential diagnostic consideration was primary brain tumor, such as hemangioblastoma or cystic astrocytoma. Metastatic brain tumor was less likely.

The patient underwent a suboccipital craniotomy and subtotal removal of the tumor was achieved. At surgery, a grayish and poorly vascular tumor with multiple non-communicating cysts was found in the cerebellar vermis. However, multiple calcified masses were hard to be removed. Postoperative CT scan showed residual calcified mass (Fig. 2). Histologically, the tumor consisted of astrocytes and a few ganglion cells with hyperchromatic nuclei (Fig. 3). Numerous microcalcifications were noted. Based on these findings, cerebellar ganglioglioma was diagnosed. Postoperative course was uneventful. The patient was discharged without radiation therapy.

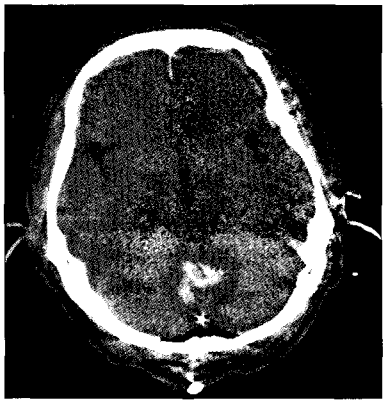

Fig. 2. Postoperative computed tomography scan showing residual calcified mass.

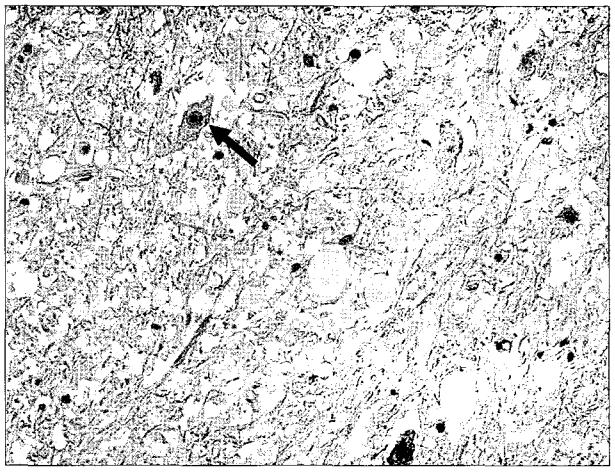

Fig. 3. Histopathological examination of the tumor demonstrating the tumor consist of astrocytes and a few ganglion cells (arrow) with hyperchromatic nuclei. Numerous microcalcifications are noted (H & E : original magnification ×200).

DISCUSSION

This case study is unique in two aspects : (1) ganglioglioma in the cerebellum and (2) ganglioglioma in old age population. Even though gangliogliomas could be found anywhere throughout the central nervous system, temporal lobe is known to be the most common location. Gangliogliomas may be rarely found in the spinal cord, brain stem, third and fourth ventricles, cerebellum, pineal region, thalamus, sella, and optic nerve[15]. These tumors mainly affect children and young adults during the first three decades of life[4,15]. In one report of gangliogliomas in 18 adult patients[5], the median age at diagnosis was 33.7 years, with a range of 21 to 55 years. Tumors were located in the temporal lobe (6 patients), temporal lobe and thalamus (1 patient), frontal lobe (5 patients), cerebellum (4 patients), and insular or thalamus (1 patient each). Unusual location of the tumor was relatively common in adults[5].

Clinically, the most common presenting symptom is seizure, with an incidence rate of 72-100% in the literature[4-6,15]. In infratentorial gangliogliomas, seizure could be presenting symptom[6]. There were some evidence of cerebellar seizure : (1) the discrete cerebellar activation seen on ictal SPECT scanning indicates that the clinical symptomatology is cerebellar in origin, (2) the cerebellum is known to have somatotopic representation of the head, face, eyes, and limbs. Electrical and mechanical stimulation of the cerebellum in cats and humans produces motor manifestations without alterations in consciousness, (3) automonic changes, such as respiratory variations, may be produced with cerebellar stimulation, and (4) electrical stimulation of the cerebellum can cause generalized seizures in animal models[9]. Rarely, myoclonus could be presenting symptom in cerebellar gangliogliomas[10].

The findings of CT scan are not specific; the mass is usually hypodense or isodense, with calcification present in 10% to 31%. In our case, preoperative CT scan was not carried out but postoperative CT scan showed definite calcifications. MR findings also are not characteristic. The tumor appears as a well-defined lesion with variable mass effect. Gangliogliomas usually have low signal on T1-weighted images but some are isointense. On T2-weighted images, solid lesions show increased signal[1]. The variable signal in cystic components depends on whether the contents are proteinaceous, hemorrhagic, or contain CSF. In our case, an intratumoral hemorrhage was observed. In the cerebellar vermis lesions, hemangioblastoma, cystic astrocytoma, and metastatic lesions should be considered in the differential diagnosis, but unlike our case, they usually enhance intensely on postcontrast MR imaging.

Although gangliogliomas are thought to be a tumor of low malignancy[15], rare cases of anaplastic gangliogliomas have been reported[8]. Some of residual tumors could be transformed to a higher grade of tumor[6]. Histologically, these tumors are comprised of neuronal and glial elements. Both cellular elements show cytologic features of neoplasia and variation in their relative proportions and regional differentiation. Malignant potential depends on the biologic behavior of gliomatous element[12].

The extent of resection appears to be the main prognostic factor[4]. The basic treatment for gangliogliomas and the best chance for a cure is a gross total resection[4-6]. Long term survival is achieved if gross total resection is possible, ranging between 7 and 17 years[4,5]. However, survival rate is less favorable when the tumor is localized midline structures (e.g. brain stem or optic nerve) and when there are anaplastic features[5]. For most gangliogliomas, radiation therapy seems to be of no benefit. Although the benefit of radiation therapy is inconclusive, it was given to patients with a tumor histology showing anaplastic features or oligodendroglial-like cells[5,12,15]. Although the tumor was subtotally removed in our case, we decided not to give radiotherapy postoperatively since there were no anaplastic features. MR surveillance should be indicated.

CONCLUSION

We experienced a case of ganglioglioma of the cerebellum in an old patient. If a cystic cerebellar mass is associated with calcifications and intratumoral hemorrhage, ganglioglioma should be included in differential diagnosis.

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