Recurrent Pilocytic Astrocytoma after 5 Years with Anxiety and Headache

Abstract

Pilocytic astrocytomas (PAs) account for 25% of all pediatric brain tumors and commonly occur in the first two decades of life. Recurrence and/or regrowth of residual tumors are not common and mostly occur within 4–5 years of the first surgery. Herein, we reported a 16-year-old male adolescent with neurological symptoms and signs that pathologic specimens confirmed the diagnosis of PA. The patient underwent 27 sessions of radiotherapy without surgery. After 5 years, at the age of 21, the patient readmitted with anxiety and headache and the recurrence of the same tumor with the same grade. We suggest follow-up of the patient after initial treatment for at least 5 years with considering any neurological symptoms including behavioral changes.

Keywords: *Case report, pilocytic astrocytoma, recurrence*

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Introduction

Pilocytic astrocytomas (PAs) were recognized by pathologists over 70 years ago.[1] They are forming 8% of all gliomas[2] and 25% of all pediatric brain tumors and commonly occur in the first two decades of life.[1-7] PA has classified as Grade I gliomas by the World Health Organization (WHO).[8] These tumors have excellent outcomes and can be curable after complete surgical resection.[1,4-6,8,9] Anaplastic and metastatic features are rare.[1,3,8,10] The most common places that tumor involved are the cerebellum, diencephalon, brainstem, and optic pathway. The neurological signs and symptoms of the tumor are related to tumor localization and mass effects.[1,8] Recurrence and/or regrowth of residual tumors are not common and mostly occur within 4–5 years of the first surgery.[10] Many factors have an influence on recurrence and regrowth of the residual tumors such as patient-related, treatment-related (the extent of surgery, total, or subtotal), and tumor-related factors.[5] According to many researches on PA, at the moment, the exact details of biological behavioral of PA are not clear yet.[5] In this article, we report a recurrent PA after 5 years and try to achieve a good overview of PA properties and its management.

Case Report

A 16-year-old male adolescent was presented to the neurosurgery ward on May 31, 2011, with headache for 2 weeks ago and diplopia for the last week. Vertigo and occasional nausea and vomiting were present. There was no significant medical history. Drug history was analgesic, omeprazole, and Vitamin B1. Papilledema was noted in fundoscopic examination of both sides. The patient was conscious, and physical examination was otherwise unremarkable. Vital signs were stable with blood pressure of 90/70 mmHg. Brain computed tomography (CT) scanning without contrast showed cystic mass with hypodense area measuring 5 cm × 6 cm with peripheral edema in the left parietal lobe and slight right midline shift. Magnetic resonance imaging (MRI) was recommended for better evaluation. On June 3, 2011, MRI of the brain reported a solid cystic mass lesion measuring 70 mm × 50 mm × 45 mm in the left temporo-parietal lobe, leading to compression on lateral ventricle and cerebral peduncle. Midline shift was seen to the right. Solid portion of the lesion measuring 32 mm × 22 mm showed marked enhancement. Lateral ventricle tumor with extension to midline was biopsied after craniotomy with primary diagnosis of choroid plexus papilloma. The specimen consisted of several pieces of creamy tissue with soft consistency measuring 0.8 cm × 0.6 cm × 0.5 cm. The pathologist reported PA [Figure 1]. Brain MRI with gadolinium (gad) on August 6, 2011, reported a 20 cm × 30 cm mass in medial part of the left temporal lobe. After

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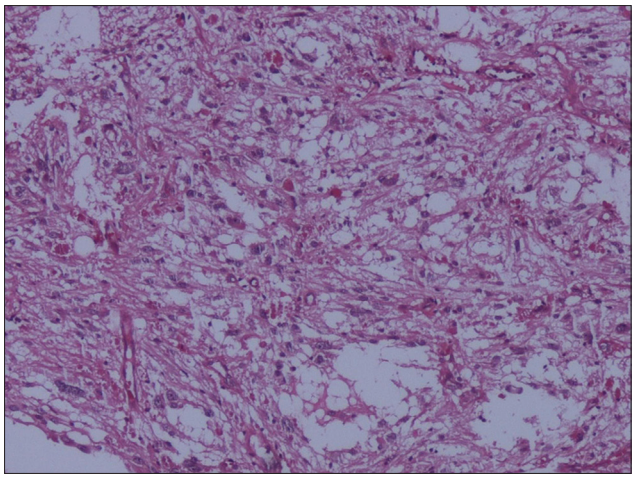

Figure 1: Pilocytic astrocytoma (H and E, ×100)

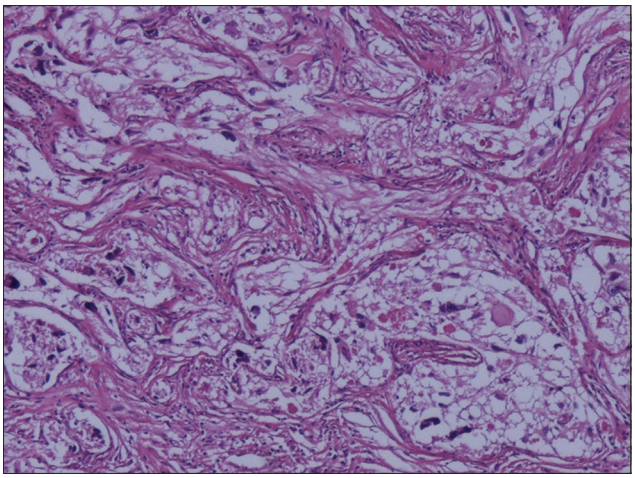

Figure 2: Recurrent pilocytic astrocytoma after 5 years of presentation (H and E, ×100)

gad injection, intense enhancement of the mass was noted. There were two cystic lesions in the atrium of left lateral ventricle (12 mm) and another in posterolateral to the left midbrain. MRI on December 16, 2011, with and without gad noted a 30 mm × 20 mm mass lesion in medial part of the left temporal lobe including hippocampus. After gad injection, intense homogeneous enhancement was noted in the lesion (tumor remnant).

The patient underwent 27 sessions of radiotherapy on the next year and prescribed anticonvulsant (unaware of the name of the drug). After that, he refused follow-up. He readmitted in the neurosurgery ward on September 8, 2016, with the chief complaint of headache, anxiety, and nervousness for 2 weeks ago. MRI with and without contrast revealed a cystic mass lesion with enhancing wall and mural nodule of 69 mm × 51 mm in the left temporo-parieto-occipital lobe. The lesion caused pressure effect on the left lateral ventricle, midline shift, and the right with subfalcine herniation. Lumbar puncture on October 5, 2016, showed scanty cellular smear with no malignant cell. Surgery was done on October 6, 2016. The specimen consisted of several fragments of creamy brown tissue measuring 2.5 cm × 2.5 cm × 1.2 cm. The pathologist reported PA (WHO Grade I/IV) the same as the primary tumor [Figure 2].

Discussion

The present case was a 16-year-old adolescent admitted to the neurosurgery ward with neurological symptoms and signs. Brain CT scanning revealed hypodense and cystic mass and pathologic specimens confirmed the diagnosis of PA. The patient underwent 27 sessions of radiotherapy without surgery. After 5 years, the patient readmitted to the neurosurgery ward with the same pathological features. For discussing this case, the first of all, we should overview features and management of PA to understand better about this tumor. PAs have survival rate up to 95% in 10 years despite their different behavior.[1,4-6,8,9] Its clinical sign and symptoms are insidious because of slow growth, and it depends on the localization of tumor and their mass effects. These include ataxia and other gait disturbances, signs of increased intracranial pressure (nausea, vomiting, and headache), visual disturbance of optic pathway involvement, cranial nerve defects, and behavioral changes such as anxiety and psychosis due to the effect on cerebral lobes. On neuroimaging studies, PAs' features on CT scan are well-defined round/oval lesions that are hypodense or isodense and enhance with contrast. On MRI, they appear with hypointense or isointense on T1 and hyperintense masses on T2 weighted with enhancement. They also show cystic or cystic solid compartments on neuroradiology modalities. In histopathology specimens, PAs have low-to-moderate cellularity with fibrillated areas (Rosenthal fibers) and also have mucoid background materials with microcyst compartments. Microvascular and endothelial proliferation and hyalinized vessels are often seen, but these do not show malignancy. Mitoses, calcification, and necrotic foci are rare.[1,8] Metastasis and seeding are not common.[3,8,10] PAs are almost sporadic tumors; however, in some cases, particularly in optic pathway, gliomas have the association with familial tumor syndromes such as neurofibromatosis type 1.[1,7,9] Most studies showed that the extension of tumor resection with surgery is the main factor that influenced on patient outcomes such as true recurrence, regrowth, or regression of residual tumors.[4,5,10] In follow-up patients with PAs, we have some terms that need attention: (1) true recurrence that defined as the discovery of new tumors on neuroimaging after total resection, (2) regrowth, and (3) regression that these terms are according to residual tumors after partial surgery or nonsurgical approaches.[5] Total resection has higher survival rate and lower recurrence rates than partially removed PAs.[8] Although total resection is choice of treatment, in tumors localized in the brainstem, optic structures, and other deep brain or midline places, because of adverse neurological deficit after surgery, total

resection is not possible and patients should undergo partial surgery or other approaches. In these cases, appropriate approaches are "close observation" or "wait and see" policy with neuroimaging follow-up for at least 5 years to detect regrowth or regression residual tumors as soon as possible,[3,5,6,10] radiation therapy (controversial), radiosurgery, or targeted therapy. The use of radiation therapy after surgery in patients with PAs is controversial. Several studies showed no benefits and also higher rate of recurrence, and it also makes anaplastic features and vascular malformation.[5,10,11] Clinical symptoms and histopathology signs are the same in recurrent and nonrecurrent tumors. Although total resection is needed to avoid recurrence of tumors, this approach is debatable. Some studies reported no recurrence or in other words no growth progression, even seen regression in incomplete resections. It is demonstrated that the extent of surgery and other treatment modalities are not the sole factors in the prognosis of PAs, and other factors such as genetic profiles and pathological features should be considered.[10] The present case was not a good candidate for surgery, and he underwent radiotherapy.

Conclusions

According to our knowledge from the past studies, surgery is the first-line therapy in PAs, and radiotherapy is controversial with the high rate of recurrence and transformation to anaplastic PAs. This case confirmed this opinion that radiotherapy may not be a good treatment for PAs, and we should try to improve our studies about other factors such as genetic profiles and histological behavior of tumors to discover new better therapeutic approaches such as target and cell therapy. We suggest follow-up of the patient after initial treatment for at least 5 years with considering any neurological symptoms including behavioral changes.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. The patient understands that her name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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