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Case report

Malignant transformation of diffuse infiltrating glial neoplasm after prolonged stable period initially discovered with hypothalamic hamartoma

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ABSTRACT

We present a case of malignant transformation of diffuse infiltrating glial neoplasm after a prolonged stable period on magnetic resonance imaging (MRI) and spectroscopy (MRS) initially discovered with a hypothalamic hamartoma. Although MRI and MRS suggest the possibility of malignant transformation in future, they cannot precisely predict the timing of rapid growth.

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1. Introduction

Diffuse astrocytoma is characterized by a high degree of cellular differentiation and slow growth corresponds to WHO grade II which may be located in any region of central nervous system including the region of junction of the three cerebral lobes. Gliomatosis cerebri (GC) is a rare, diffusely infiltrating glial neoplasm with little mass effect that is usually associated with a poor prognosis [1]. Hypothalamic hamartoma (HH) is a benign congenital malformation of the brain containing heterotopic nervous tissue. Magnetic resonance imaging (MRI) of HH reveals a sessile hypothalamic mass suspended from the floor of the third ventricle, isointense in T1-weighted images (T1WI) and iso or hyperintense in T2-weighted images (T2WI) to gray matter [2].

Here we present a case of an intra-axial diffuse infiltrating glial neoplasm whose MR spectroscopy (MRS) appearance was consistent with a GC but revealed a prolonged stable or slow growing course followed by sudden symptomatic development of a high grade glioma combined with an HH compatible mass.

This study was performed with the approval of the institutional ethics committee of our university, and after informed consent was obtained from the patient.

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2. Case report

An 8-year-old male patient without any history of remarkable neurological disorders from birth was evaluated at a pediatric endocrinology department because of signs of precocious puberty (enlarged penis and muscular build). His family history was not contributory. MRI revealed a hypothalamic mass involving the mammillary body and showing isointensity in all the MR pulse sequence images that was diagnosed as hypothalamic hamartoma. Simultaneously, an abnormal signal lesion showing homogeneous hyperintensity on fluid attenuated inversion recovery (FLAIR) images, and T2WI, and hypointensity on T1WI with no contrast enhancement was noted to spread in the right temporal, parietal and occipital lobes. Although GC or a diffuse astrocytoma accompanying HH

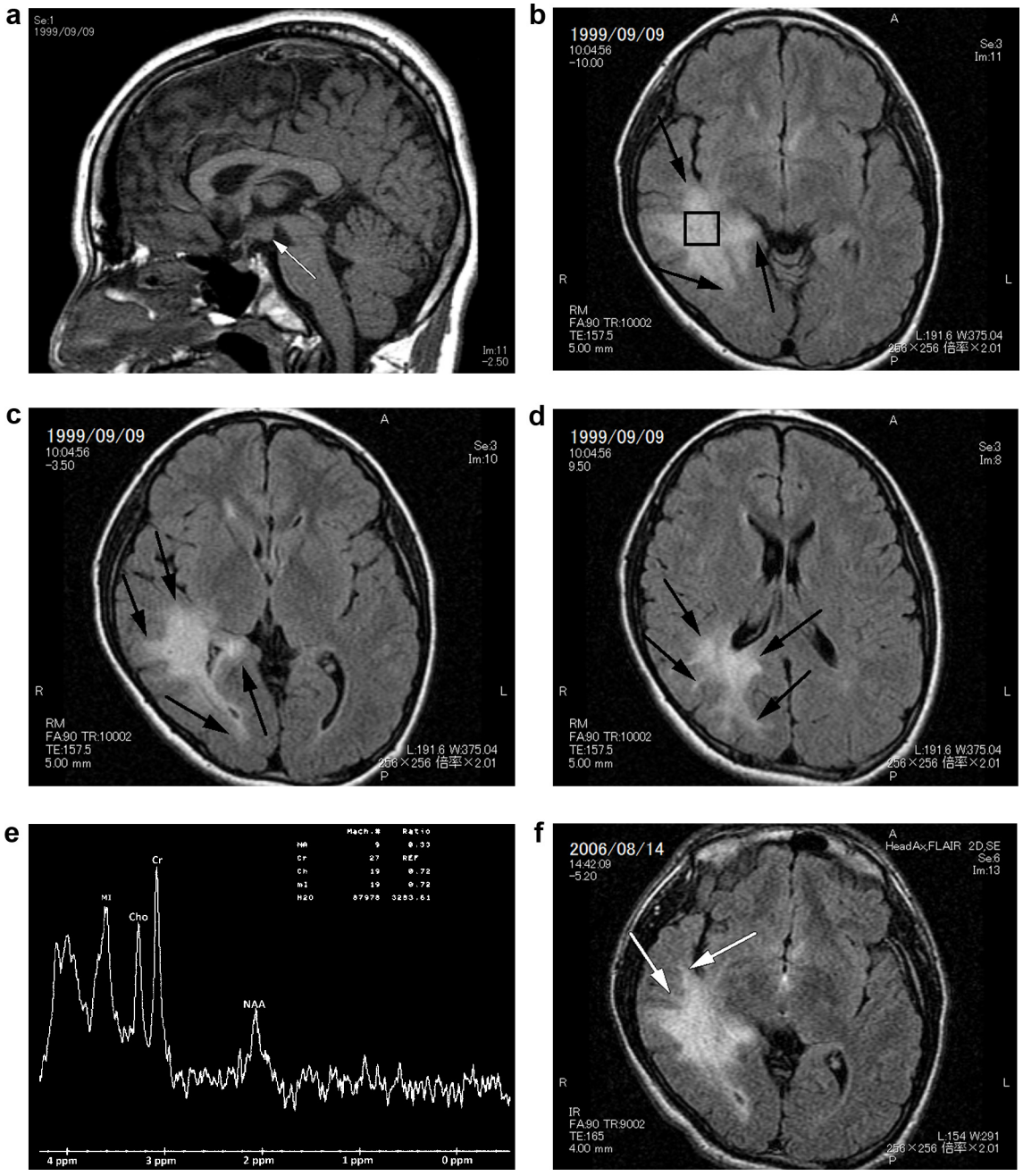

Fig. 1 – Magnetic resonance imaging and spectroscopy (MRS) examined at ages 15 and 21 years. (a). Sagittal T1 weighted image shows a sessile isointense hypothalamic mass with gray matter that was suspended from the floor of the third ventricle (arrow). (b)–(d). Axial fluid attenuated inversion recovery (FLAIR) images show extensive white matter hyperintensity of the right temporal, parietal and occipital lobes surrounding the trigon and posterior horn of right lateral ventricle (black arrows). Voxel indicates sampling area of MRS. (e). MRS reveals reduction of N-acetylaspartate, and elevation of myoinositol without choline elevation. (f). Axial FLAIR image examined when the patient was 21 years reveals anterior extension of the hyperintensity (arrows).

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