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ScienceDirect
Surgical Neurology 70 (2008) 414–420
SURGICAL NEUROLOGY
www.surgicalneurology-online.com

Neoplasm

Cerebral ependymoma in a patient with multiple sclerosis: case report and critical review of the literature

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Received 22 March 2007; accepted 24 April 2007

Abstract

Background: The concurrence of multiple sclerosis (MS) and brain tumors is a rare but well-recognized condition. The radiologic evidence of the progressive evolution of a mega-plaque in a tumor has never been described. We report the first case of such an occurrence.

Methods: A 27-year-old woman with a diagnosis of MS was referred to us for an intense frontal headache. Magnetic resonance imaging showed a mass lesion in correspondence of a black hole lesion previously diagnosed. The patient was operated on, with complete removal of the tumor documented by an intraoperative MRI. The histologic examination evidenced an ependymoma. Postoperative radiotherapy was performed.

Results: The patient is well and recurrence-free at 2 years follow-up.

Conclusions: The present case, documenting the transformation of a mega-plaque into a tumor, suggests a cause-effect relationship between MS and brain tumors.

Keywords: Black hole; Ependymoma; Intraoperative MRI; MRI; Multiple sclerosis

1. Introduction

The concurrence of cerebral tumor and MS is very rare [3,16,18,19,23,24,29,52,59,61,66]. The first case was described by Bosch [10] in 1912. Since then, about 40 cases have been reported. The tumor was an ependymoma in 1 case only [15].

The aim of this article is to report another case of ependymoma in a patient with MS and discuss the etiopathogenesis through an analysis of the literature available to us. The presented case is particularly interesting because radiologic imaging documented the progressive transformation of right frontal MS plaque to an ependymoma.

2. Case report

A 14-year-old adolescent girl presented with vertigo, diplopia, and ataxia. An MRI showed multiple hyperintense lesions on T2-weighted images and on DP images in the right subcortical frontal region, corpus callosum, left corona radiata, pons, mesencephalon, and right capsula interna (Fig. 1A-C). Therapy with interferon and cortisone was started on the basis of MS diagnosis. There was a regression of the symptomatology, but 6 years after diagnosis, the patient presented with a spastic tetraparesis. A second MRI (Fig. 2A) showed an enlargement of the previous plaques. The previous hyperintense plaque in the frontal area became hypointense on T1-weighted images without contrast enhancement. This feature was interpreted as a "black hole" mega-plaque. At 8 (Fig. 2B) and 10 (Fig. 2C) years after diagnosis, MRI black hole and periventricular plaques increased in volume. Therapy with mitoxantrone 7.5 mg IV every 2 months for 2 years was administered. Clinical picture remained stable for 3 years. When the patient was 27 years

Abbreviations: 3D, 3-dimensional; GFAP, glial fibrillary acidic protein; HPF, high-power field; MR, magnetic resonance; MRI, magnetic resonance imaging; MS, multiplesclerosis.

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doi:10.1016/j.surneu.2007.04.015

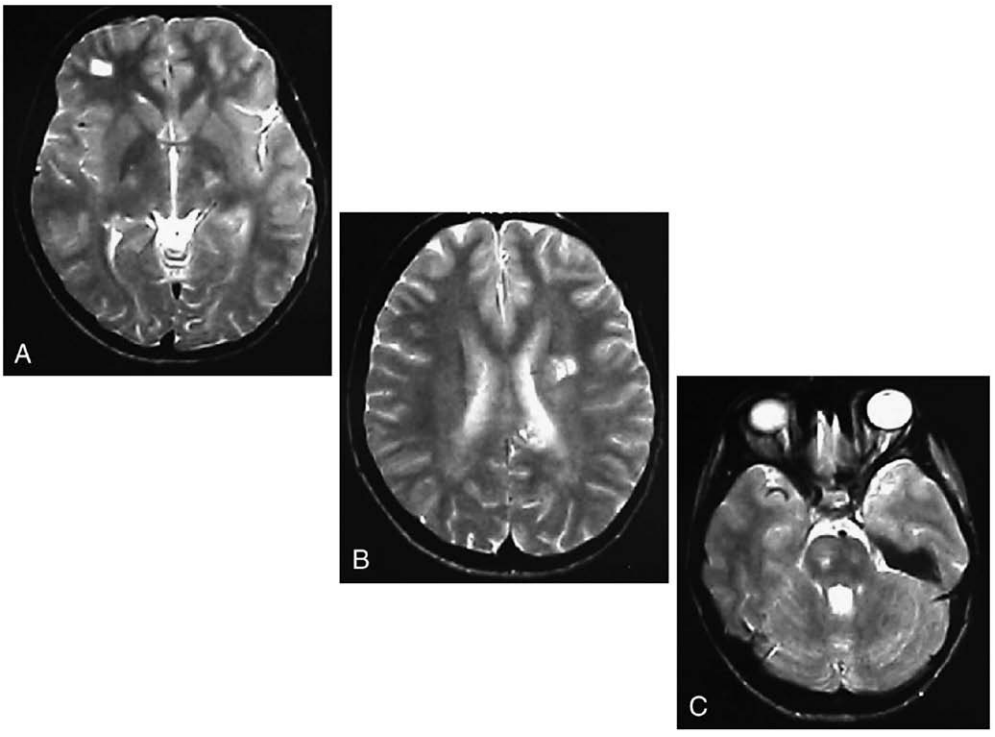

Fig. 1. Magnetic resonance imaging illustrating multiple hyperintense lesions on T2-weighted images in the right subcortical frontal region (A), capsula interna (B), and pons (C).

old, she experienced intense headache. A new MRI showed a slightly hyperintense mass lesion on T1- and T2-weighted images in correspondence of the black hole, with scarce contrast enhancement and midline structure shift (Fig. 3A-C). An MRI spectroscopy (Fig. 3D) evidenced a choline peak within the frontal lesion. A frontal craniotomy was performed, and the lesion was totally removed. The patient was operated on in our operative room, BrainSuite (BrainLab®) and provided with an intraoperative MRI (1.5T-Magnetom Sonata; Siemens) integrated with a Neuronavigator (BrainLab's Vector Vision system) and operative microscope (NC4; Zeiss). Intraoperative MRI showed a small residue of the lesion in the posterolateral portion of the operative field (Fig. 4A). This residue was removed. A second intraoperative MRI documented a complete removal of the lesion. (Fig. 4B).

Microscopic examination demonstrated a glial tumor with moderate cellularity and pleomorphism mainly represented by cells with round to oval, dark nuclei with regular borders intermingled with a minor component represented by cells with oligolike clear cytoplasm and gemistocytes (Fig. 5A).

The neoplasia showed an evident pattern in perivascular pseudorosettes (Fig. 5A) and areas more destructurated with microcysts (Fig. 5B). Focally, in an area rich in pseudorosettes, different sections showed the presence of microfoci of tumor necrosis with palisading and prevalence of astrocytic features with gemistocyticlike cells (Fig. 5C).

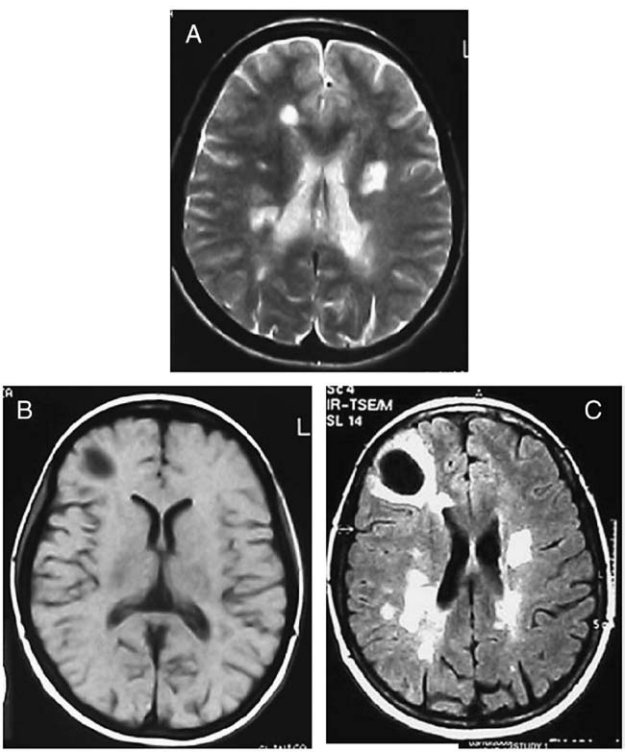

Fig. 2. A: Second MRI, 6 years after diagnosis, showing an enlargement of the frontal plaque. Eight (B) and 10 (C) years after diagnosis, black hole becomes evident, and periventricular plaques increase in volume.