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Case study

Suprasellar giant cell ependymoma: a rare neoplasm in a unique location

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Summary Ependymomas are glial tumors that usually present in the posterior fossa in children and in the spinal cord in adults. Giant cell ependymoma, a rare ependymal subtype only recently recognized as a diagnostic entity in the last decade, demonstrates pleomorphic giant cells admixed with features of typical ependymoma. Although only 8 giant cell ependymomas have been reported to date, none have been reported in the suprasellar space. Moreover, as these neoplasms demonstrate a high incidence of anaplastic grade, recognition of this ependymal subtype is paramount. We describe the presentation and pertinent radiologic, histologic, immunologic, and ultrastructural findings in conjunction with relevant clinical implications of the first reported case of a suprasellar giant cell ependymoma occurring in a 34-year-old female 7 years after an initial diagnosis of a medullary ependymoma with rare atypical giant cells, a potential tumor seeding culprit.

1. Case report

The patient is a 27-year-old right handed woman who initially presented with 3 months of progressive numbness in her left hand and foot. Examination demonstrated decreased sensation in the left leg and hand, hyperreflexia, and difficulty swallowing. Magnetic resonance imaging (MRI) revealed a 2.5-cm enhancing mass at the medullary brainstem junction, without evidence of other spinal or cranial lesions (Fig. 1A and B). The patient was taken to the operating room, and the tumor was removed via suboccipital approach. A subtotal resection was performed, resulting in the removal of 95% of the tumor. A small portion could not be resected because of the adherence of the tumor to the brainstem. The initial histologic evaluation of 4-μm-thick, formalin-fixed, paraffin-embedded sections stained with hematoxylin and eosin (H&E) demonstrated unipolar processes extending from tumor cells toward blood vessels in a perivascular rosette, typical of ependymoma (Fig. 2A), that were additionally highlighted by immunostaining for glial fibrillary acidic protein (GFAP). Moreover, epithelial membrane antigen (EMA) staining highlighted numerous distended intracytoplasmic lumens as well as classic ependymal perinuclear dot-like staining (Fig. 2B). A Ki-67 stain estimated a proliferative index of 10% to 30%. Although frequent mitoses, microvascular proliferation, and/or pseudopalisading necrosis were not identified to warrant a World Health Organization (WHO) [1]

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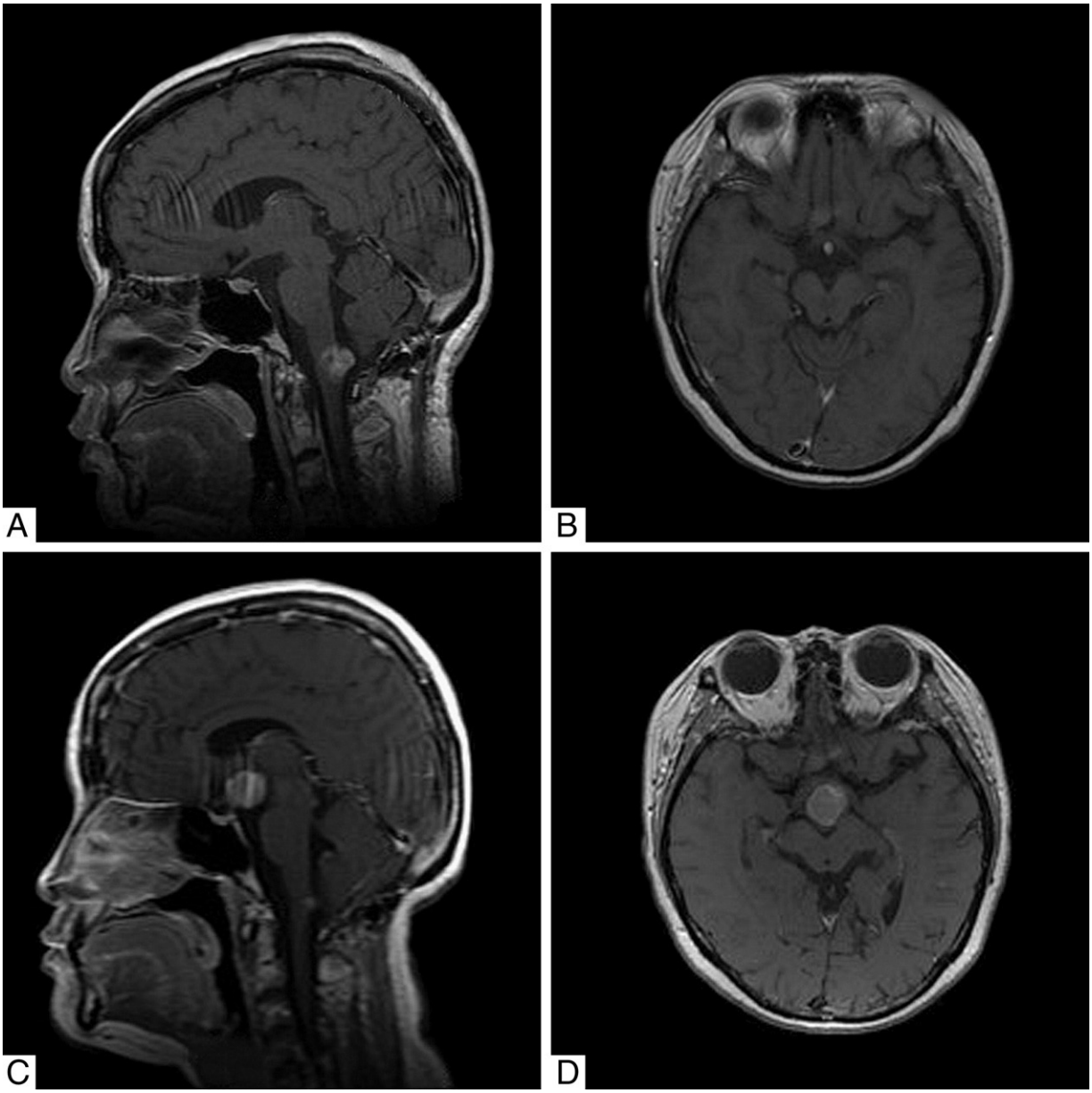

Fig. 1 A, Sagittal T1 with contrast MRI demonstrates an enhancing mass at the medullary junction. B, Axial T1 with contrast MRI demonstrates no signs of a suprasellar mass. Sagittal (C) and axial T1 (D) with contrast MRI demonstrate an enhancing suprasellar mass.

grade III lesion, occasional bizarre, pleomorphic large cells were identified (Fig. 2A). A concurrent cerebrospinal fluid obtained during the operation showed no evidence of malignancy.

The residual lesion was treated with external beam radiotherapy with 21 fractions to a total of 37.8 Gy in 180-cGy fractions. Although originally scheduled to receive a total of 45 Gy, the patient received only 37.8 Gy because of intractable nausea. However, she tolerated a subsequent radiosurgical boost to the tumor bed.

Postoperative surveillance demonstrated no recurrence in the medullary brainstem junction; however, MRI demonstrated a new small area of suprasellar enhancement 5 years later upon follow-up examination. Workup at that time included a lumbar puncture and spine MRI, both of which demonstrated no pathologic findings. Interval MRIs within the next 2 years demonstrated progression of the suprasellar lesion for which she received stereotactic radiosurgery. Despite radiation, the lesion continued to grow (Fig. 1C and D) and caused visual symptoms. The patient was subsequently taken to the operating room for decompression and to obtain a tissue diagnosis.

Standard H&E histologic sections demonstrated a cellular proliferation of nested to somewhat trabecular cohesive tumor cells with a biphasic nuclear appearance ranging from small and round to large, eccentric, and pleomorphic; the latter larger cells constituted approximately 20% of cells and were reminiscent of the bizarre, pleomorphic cells seen in the patient's prior biopsy (Fig. 3A and D). These cells also