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Cerebellar Gliosarcoma: Report of a Probable Radiation-Induced Neoplasm

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Gliosarcoma is a primary brain tumor composed of neoplastic glial cells and a spindle-cell sarcomatous component. Supratentorial gliosarcoma has been reported as a complication of radiation therapy for pituitary adenoma and meningioma [1]. This report describes a cerebellar gliosarcoma in a patient who had undergone radiation treatment for parotid carcinoma.

Case Report

A 34-year-old woman presented with a 2-week history of nausea, headaches, blurred vision, and difficulty walking. She had undergone a right parotidectomy for a mucoepidermoid carcinoma 8 years prior to admission, after which she received a course of radiation therapy (50 Gy) to the parotid bed at another institution. A noncontrast CT scan of the head done before admission was limited owing to streak artifact in the posterior fossa. At the time of transfer to our hospital, neurologic examination revealed left gaze nystagmus with an upward rotatory component, papilledema, and ataxia. MR of the brain with and without contrast enhancement confirmed the presence of hydrocephalus and also demonstrated a ring-enhancing mass in the deep right cerebellar hemisphere, compressing and displacing the fourth ventricle (Figs. 1A and 1B). Unenhanced proton-density-weighted (Fig. 1C) and T2-weighted (Fig. 1D) images demonstrated areas of tumoral and peritumoral high signal intensity without evidence of radiation-induced white matter changes. A well-circumscribed, partially necrotic tumor was found at craniotomy that had the appearance of a metastatic lesion rather than a primary neoplasm.

Histopathologic studies included routine H and E stains as well as immunoperoxidase stains for neuron-specific enolase, glial fibrillary acidic protein, keratin, S-100, and special stains for mucin and reticulin. The overall histologic pattern and staining characteristics of two distinct populations of tumor cells was consistent with gliosarcoma. A review of pathologic material from the patient's original parotid carcinoma excluded metastatic disease from this site.

Despite surgery and postoperative radiation therapy, the patient died approximately 5 months after the initial craniotomy. Postmortem was not performed.

Discussion

Gliosarcoma is a high-grade astrocytoma in which the vascular stroma has taken on the appearance of a spindle-cell sarcoma [2, 3]. It is well recognized that gliomas can induce sarcomatous transformation in supporting mesenchymal elements [4]. Feigin et al. [5] proposed that the sarcomatous portion arises directly from neoplastic transformation of the vascular elements within a glioblastoma. A similar pathogenesis has been proposed for sarcoglioma, in which a primary sarcoma induces neoplastic change in adjacent glial cells [6]. About 79 cases of gliosarcoma have been described in the literature as of 1987. In one series, these lesions represented approximately 8% of all anaplastic astrocytomas [7]. The tumors tend to be peripherally located, show a preference for the temporal lobes, and are often mistaken at surgery for meningioma [7]. While selective metastases of the sarcomatous portion of gliosarcoma have been reported [8, 9], survival rates for patients with this tumor are no worse than those for anaplastic or high-grade astrocytomas.

The CT and angiographic characteristics of gliosarcoma have been described previously. Lee et al. [10] described homogeneous tumor enhancement on contrast CT in their series of five patients with gliosarcoma. This was not the experience of Jack et al. [11], who recently described the angiographic and CT appearance of 14 cases. The tumors in their series typically demonstrated an intensely enhancing, irregular rim with a low-density center on contrast CT. In six of nine cases with angiographic correlation, the tumors had a prominent ringlike or nonhomogeneous stain with a well-defined margin and a centrally located area of hypovascularity. On gross examination, the peripheral sarcomatous portions of the tumors were sharply demarcated from adjacent brain tissue, while the central astrocytic components were poorly defined and necrotic.

Contrast CT and angiography were not performed in our case. Noncontrast CT demonstrated mass effect in the posterior fossa, compression of the fourth ventricle, and hydro-

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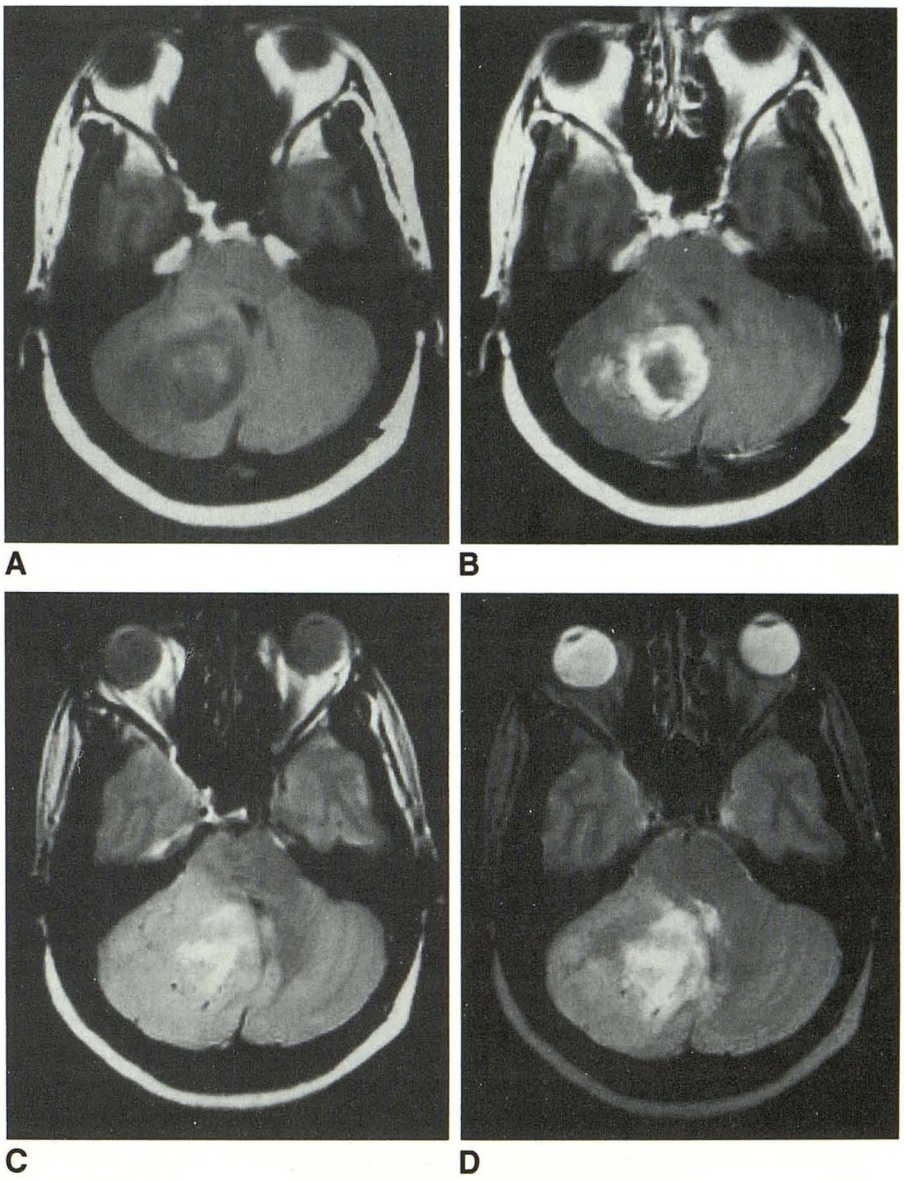

Fig. 1.—34-year-old woman with cerebellar gliosarcoma.

***A* and *B*, Axial T1-weighted (600/20/1) images before (*A*) and after (*B*) contrast enhancement show hydrocephalus and ring-enhancing mass in deep cerebellar hemisphere, compressing and displacing fourth ventricle.**

***C* and *D*, Unenhanced axial proton-density-weighted (2500/25/1) (*C*) and T2-weighted (2500/90/1) images show areas of tumoral and peritumoral high signal intensity without evidence of radiation-induced white matter changes.**

cephalus. Contrast-enhanced MR of the brain demonstrated a ring-enhancing, well demarcated tumor with an avascular central portion. Although nonspecific, the MR appearance of the tumor in this case was similar to the CT and angiographic features of gliosarcoma described previously by Jack et al. [11].

To our knowledge, gliosarcoma has not been previously described in the posterior fossa. The unusual location of the tumor in this patient caused us to consider the possibility of a radiation-induced neoplasm, given the patient's history of treatment for a mucoepidermoid carcinoma of the parotid gland. Recognized criteria have been established to show a causal relationship between radiation therapy and the occurrence of a subsequent neoplasm [12]. These include: (1) a sufficient delay between irradiation and the detection of the second tumor, (2) the second tumor must be in the irradiated field, (3) the histology of the second tumor must be different from the initial neoplasm, and (4) a family history of tumor diathesis, such as neurofibromatosis or tuberous sclerosis, must be excluded. It has also been noted that radiation-induced neoplasms tend to occur in the periphery of the irradiated field rather than in the maximal dose region [12].

Our patient met all the criteria for presumptive diagnosis of a radiation-induced neoplasm. A total of 50 Gy was administered in fractionated doses to the right parotid region approximately 8 years prior to discovery of the second neoplasm. The gliosarcoma arose in the deep right cerebellar hemisphere, outside the maximal dose region, in the 10–15 Gy isodose field (Fig. 2). A review of the pathologic material from the patient's original parotid malignancy revealed no histologic similarities between the two tumors. There was no family history of tumor diathesis or phakomatosis.

In addition to meeting the established criteria, the occurrence of such a histologically unusual tumor in a previously

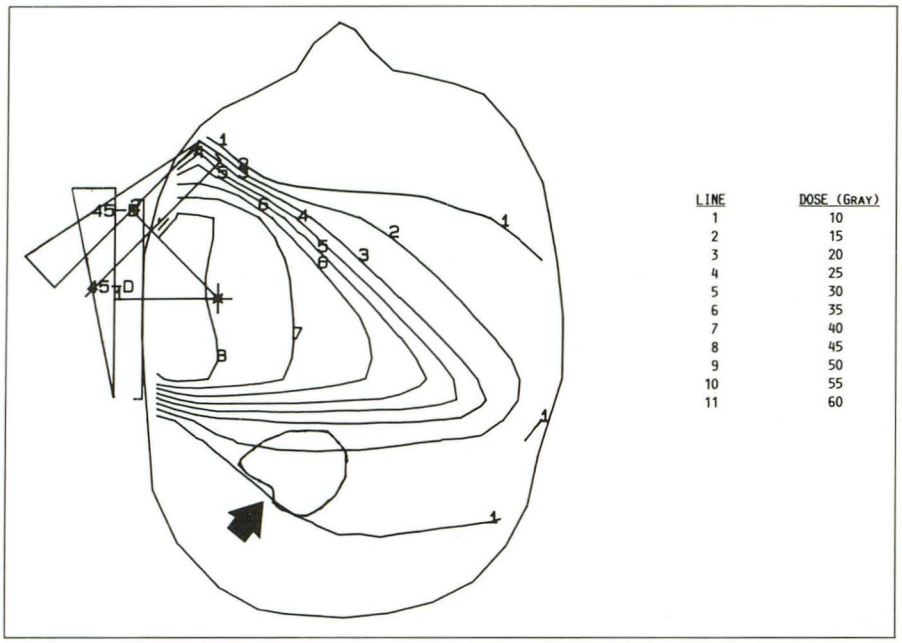

Fig. 2.—After right parotidectomy for mucoepidermoid carcinoma, the patient received 50 Gy in 25 fractions utilizing right anterior oblique and lateral wedged fields with a cobalt-60 teletherapy unit. Reconstructed isodose distribution at central axis is shown. The cerebellar gliosarcoma occurred at periphery of irradiated field (*arrow*) approximately 8 years later.

unreported location certainly supports the proposition of a radiation-induced neoplasm. Cerebellar glioma following radiation therapy for unrelated disorders has been described [13–15], and at least two cases of supratentorial mixed sarcoma-glioblastoma multiforme have been reported following radiation therapy for pituitary adenoma and meningioma [1]. Cerebellar gliosarcoma, however, would seem to be an extremely rare occurrence with or without prior radiation therapy.

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