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Neurocytoma in the cerebellum

Case report

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✓ A neurocytoma is a central nervous system tumor composed of small cells with features of neuronal differentiation; it typically occurs in the periventricular region, close to the septum pellucidum and the foramen of Monro. In this article, the authors report the case of a neurocytoma located in the cerebellum, which to their knowledge is the first reported case of its kind. The finding of a neurocytoma at a nonclassic location refutes the theory that this tumor has its origins in subependymal progenitor cells, unless an ectopic location of progenitor cells is invoked to explain the occurrence of a neurocytoma away from the ventricles. On the basis of this case, the authors suggest that neurocytomas should be added to the differential diagnosis of mass lesions in the supratentorial intraventricular regions as well as in the posterior fossa.

KEY WORDS • brain neoplasm • cerebellum • neurocytoma • posterior fossa

NEUROCYTOMAS were first described by Hassoun, et al.,⁸ after electron microscopic examination of two intraventricular tumors that resembled oligodendroglioma on light microscopy. Neurocytomas are characterized by their intraventricular localization, predominant occurrence in young adults, histological similarity to oligodendrogliomas or ependymomas, expression of neuron-specific antigens, and ultrastructural features of neuronal differentiation.^{1,5,8,16,21,23} Their rate of occurrence has been reported to be 0.1%.²³

Although a few neurocytomas have been noticed incidentally, their clinical presentation is similar to that of any other intraventricular tumor: that is, they manifest signs and symptoms of occlusive hydrocephalus, such as headache, nausea, visual and mental disturbances, and papilledema.^{1,23} Neurocytomas situated within the brain parenchyma may present with epilepsy.^{6,15} Centrally located neurocytomas usually follow a benign course; however, a few cases with malignant histopathology and malignant behavior have also been described.^{8,16,21,23} In cases of intraventricular neurocytoma, an anterior transcallosal micro-neurosurgical approach has been recommended, along with postoperative radiation therapy for malignant or subtotally removed tumors.^{5,23}

In addition to their classic presentation as an intraventricular or periventricular growth, neurocytomas have been found at other sites in the cerebrum and in the spinal cord.^{3,12,14} We report the unusual occurrence of a neurocytoma in the cerebellum.

Case Report

Examination. This 46-year-old woman presented with complaints of neck pain. A magnetic resonance (MR)

image of the patient's cervical spine indicated the presence of a cerebellar lesion. Additional investigation using regular brain MR imaging, using proton-density and T₂-weighted images, revealed the cerebellar lesion as a 1-cm-wide, high-intensity signal, which was situated in the medial inferior portion of the left cerebellar hemisphere and which showed minimum mass effect. The tumor displayed a mild ringlike enhancement after administration of gadolinium (Fig. 1).

Operation and Pathological Findings. The patient underwent suboccipital craniotomy for excision of the tumor. Histologically, the tumor resembled a well-differentiated oligodendroglioma (Fig. 2 upper). The dense uniform cellularity of the tumor was interrupted by less cellular zones of fine fibrillarity. The thin vasculature had a chicken-wire appearance. The tumor cells had round- to oval-shaped nuclei and perinuclear halo formation. Anaplastic features and mitotic figures were not observed. Large mature neurons (Fig. 2 upper) and structures resembling Homer-Wright rosettes were occasionally encountered. Bodian's copper-Protargol staining demonstrated the presence of streams of neurites in the fibrillary areas. Both the tumor cells and the fibrillary zones were immunoreactive for synaptophysin (Fig. 2 lower). Scattered reactive astrocytes that were immunoreactive for glial fibrillary acidic protein (GFAP) were present within the tumor. The proliferative index (MIB-1 labeling index) was 3 to 5%. Ultrastructurally, the tumor had a dense feltwork of cytoplasmic processes containing microtubules and filaments. Synapses were not identified, but astrocytic processes with dense fibers were occasionally seen. Scattered or aggregated neurosecretory granules were apparent either in the processes or in the perinuclear cytoplasm (Fig. 3).

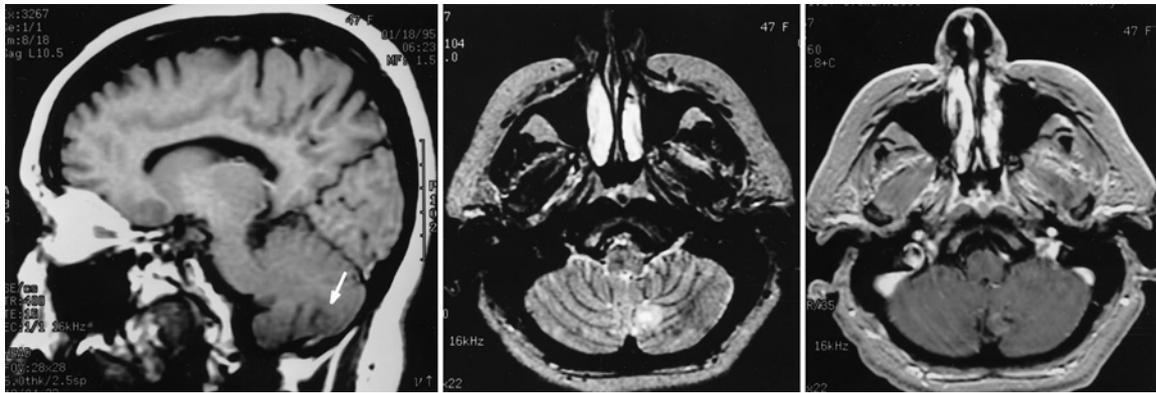


FIG. 1. Magnetic resonance imaging demonstrating the tumor in the medial inferior position of the left cerebellum as a hypointense lesion in a T_1 -weighted image (left, arrow) and as a hyperintense lesion in a T_2 -weighted image (center). Subtle ring enhancement is apparent in a gadolinium-enhanced T_1 -weighted image (right).

Discussion

Neurocytomas are small cell tumors that need to be differentiated from lymphomas, ependymomas, neuroblastomas, dysembryoplastic neuroepithelial tumors, and oligodendrogliomas.^{5,11} A significant number of intraventricular tumors initially diagnosed as oligodendrogliomas have been reclassified as neurocytomas.^{16,21} Neurocytomas typically consist of dense areas of small cells with perinuclear halos, interspersed by a patchy fibrillary network; these tumors demonstrate the immunohistochemical and ultrastructural features of neuronal differentiation. Oligodendrogliomas and ependymomas usually do not demonstrate patchy fibrillary stromata, but rather show a positive reactivity for GFAP and S-100 protein and lack markers of neuronal differentiation.^{5,20,21} Neuroblastomas have Homer–Wright rosettes and ganglion cells, which may occasionally be observed in neurocytomas.¹⁷ However, the perinuclear halo is a feature more characteristic of neurocytomas than of neuroblastomas and more than 80% of neuroblastomas occur in the first decade of life.^{2,5,17,20} Like gangliogliomas or gangliogliomas but unlike neurocytomas, dysembryoplastic neuroepithelial tumors have abundant ganglion cells and characteristically contain a mucoid matrix that is absent in neurocytomas.^{8,11} The neuropathological basis for differentiating neurocytomas from other intraventricular tumors is neuronal differentiation, including immunohistological markers such as neuron-specific enolase and synaptophysin.^{1,3,5,9,13,18,21} However, the usefulness of neuron-specific enolase is limited because of its nonspecificity. Use of antisynaptophysin antibodies to identify neurocytomas, a technique introduced by Barbosa and associates,¹ has provided consistent and specific results and, therefore, has been suggested as the most reliable immunohistological marker for neurocytoma.^{1,3,5,9,13,18,21}

In our patient, the tumor's histological resemblance to an oligodendroglioma, the positive immunoreaction to synaptophysin, and the ultrastructural findings of dense core granules and microtubules are consistent with a neurocytoma. Structures resembling Homer–Wright rosettes and large mature ganglion cells, which are rare in neurocytomas, were also identified in our case. Similar to other neurocytomas, scattered GFAP-positive astrocytes were also seen.

Neurocytomas have been reported to develop typically in the intraventricular location (lateral or third ventricle), near the foramen of Monro, or attached to the septum pellucidum.^{1,5,8,21,23} They also occur at other locations in the cerebrum such as the frontal lobe and in the spinal cord.^{3,12,14,15} Ellison, et al.,⁴ reported the case of a tumor in

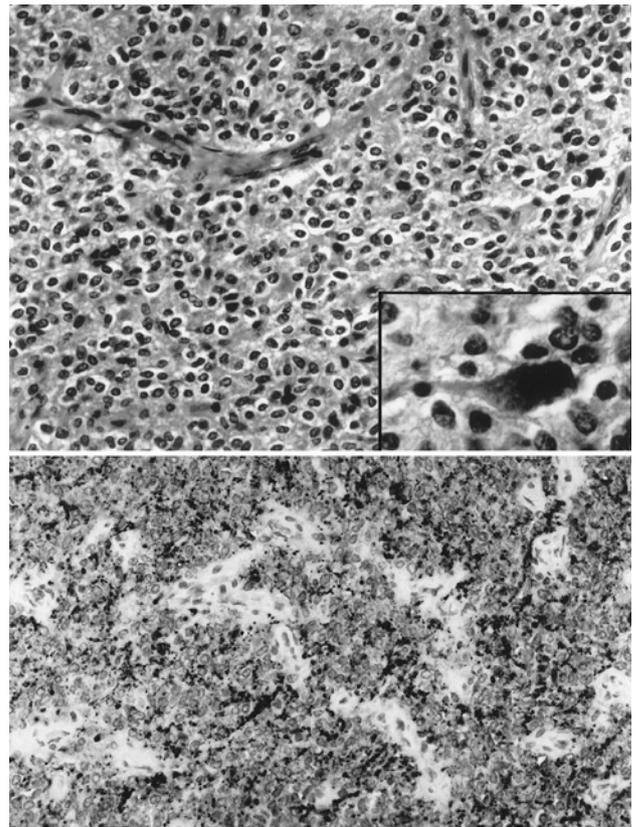


FIG. 2. Photomicrographs showing H & E staining. Upper: Tumor composed of uniform cells with perinuclear halos and delicate vasculatures, which resembles an oligodendroglioma. Original magnification $\times 80$. Upper Inset: A large mature neuron. Original magnification $\times 160$. Lower: Tumor cells displaying immunoreactivity for synaptophysin. Original magnification $\times 80$.

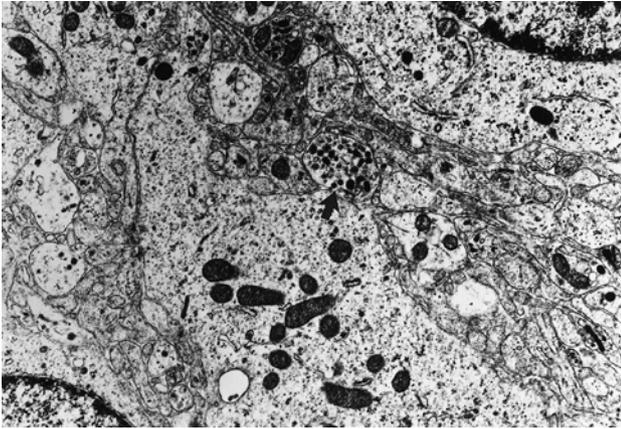


FIG. 3. Electron micrograph showing that the neuropil is composed of densely packed cytoplasmic processes containing dense core secretory granules (arrow) and microtubules. Original magnification $\times 13,700$.

the cerebellum that consisted of cells with features of a neurocytoma as well as cells with features of a lipoma and consequently termed it a neurolipocytoma. To our knowledge, our case is the first report of a neurocytoma in the cerebellum.

Ependymomas and oligodendrogliomas, two tumors whose diagnoses are most frequently confused with that of neurocytomas, are found in significant proportions in the cerebellum. Oligodendrogliomas constitute 5% of intracranial gliomas, with fewer than 10% arising in the posterior fossa; 60 to 70% of ependymomas are infratentorial, with a mean age of presentation in the late teens.⁷ Based on the strong synaptophysin immunoreactivity and the ultrastructural findings of neuronal differentiation in the cerebellar tumor under discussion, we have confidently ruled out the possibility of a cerebellar oligodendroglioma or ependymoma.

Because neurocytomas were originally observed to occur near the foramen of Monro and usually in association with the septum pellucidum, the suggestion proposed by Hassoun, et al.,⁸ that nuclei of the septum pellucidum are the source of neurocytomas was endorsed by several investigators.^{5,10,19,21-23} von Deimling and colleagues²² found expression of synaptophysin and neuron-specific enolase by all neurocytomas and coexpression of synaptophysin and GFAP in several cells. These investigators concluded from immunocytochemical, molecular biological, and ultrastructural evidence that centrally located neurocytomas are neuroectodermal tumors and proposed that they originate from bipotential progenitor cells present in the periventricular matrix of the mammalian brain. However, this speculation comes into conflict with the observation of neurocytomas in the spinal cord^{3,12} or the cerebellum, away from the ventricle. The possibility of dedifferentiation followed by differentiation of resting cells along neuronal lines, or the likelihood of ectopic embryonal matrix cells located away from the subependymal periventricular position, need to be considered.²²

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