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Brief Note

Central Neurocytoma: A Newly Recognized Brain Tumor

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The concept and classification of brain tumors may change as time passes. The oligodendrogliomas were first discovered and described as one distinct type of gliomatous brain tumor by Bailey and Cushing in 1926.[1] In 1931, Martin[2] described a subtype of oligodendroglioma growing exclusively in the third and lateral ventricles. This type of the oligodendroglioma has been recognized as a midline oligodendroglioma or supratentorial intraventricular oligodendroglioma. It has received attention mainly from the clinical point of view because it shows the symptoms and signs of increased intracranial pressure from the early stages of the illness, such as a choked disc, divergence ocular palsy, character change, organic dementia, and ataxia, without any focal signs.[2,3] Recently, Hassoun and his co-workers[4] showed in an electron microscopic study that this type of oligodendroglioma may be derived from neuronal elements and proposed the name "central neurocytoma" as a newly recognized brain tumor.

The immunohistochemical and electron microscopic reevaluations of seven cases previously diagnosed as oligodendroglioma in our institute revealed two cases that coincide with the neurocytoma advocated by Hassoun *et al.*

The first case (A 89979) was a 33-year-old woman with an 8-month-course of the illness. Computed tomography (CT) demonstrated a left intraventricular tumor of 4.0 cm in size (Fig. 1A), which was completely removed surgically (S 80-556). The second (A 6795) was a 24-year-old man with a calcified midline tumor with a 4-month-course. The patient died of cerebral edema 2 days after partial removal of the tumor (S 684). An autopsy (A 11) demonstrated a left intraventricular tumor of $5.5 \times 3.0 \times 7.5$ cm in size (Fig. 1B). Other clinicopathological details of the second case have been described elsewhere.[5]

The histopathologic and electron microscopic findings of the two cases were quite similar. The tumor cells were small and round with clear cytoplasm and a round nucleus, which occasionally showed a perinuclear halo. These tumor cells presented a uniform proliferation with vascularized septa subdividing groups of cells into lobules and took a honeycombed structure. Patchy fibrillary stroma were scattered in the tumor tissue (Fig. 2). Cellular atypism and pleomorphism of the tumor cells or endothelial proliferation of the blood vessels were not seen. Numerous calcifications were observed in the latter case but not in the former. Immunohistochemically, the tumor cells did not react to antibodies of the glial fibrillary acidic protein (GFAP), S-100 protein, myelin basic protein (MBP) and neuron specific enolase (NSE) by the peroxidase-antiperoxidase method. Electron microscopically, the tumor cells had a clear cytoplasm with many ribosomes and occasional mitochondria and microtubules.

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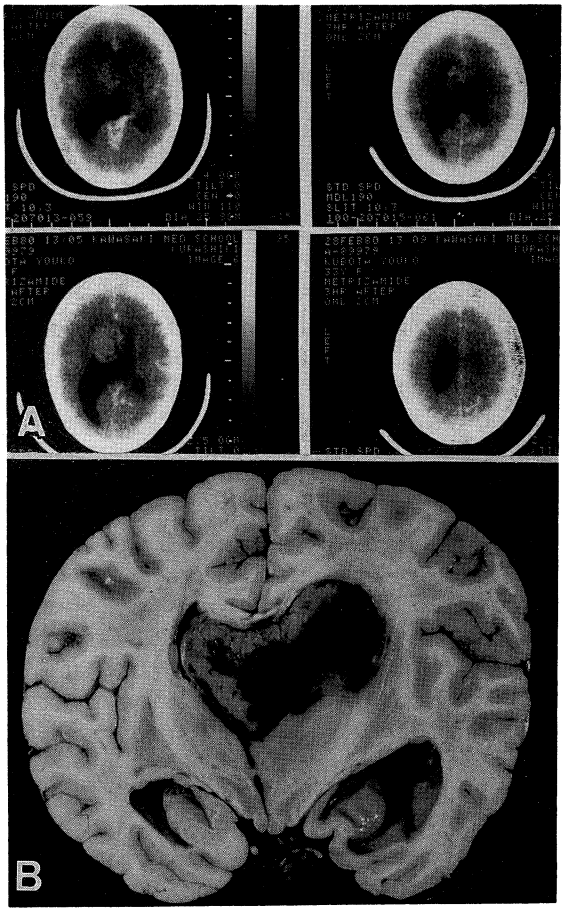

Fig. 1. A: CT of case 1 showing a left intraventricular tumor. B: Coronal section of the cerebral hemispheres of case 2 through the mammillary bodies showing a left intraventricular tumor.

They formed many entangled processes which contained numerous microtubules, dense-cored and clear vesicles, and synapse-like devices (Fig. 3).

These histopathologic and electron microscopic features are compatible with the central neurocytoma described by Hassoun *et al.* Its clinicopathological characteristics are as follows.[6] It occurs as a supratentorial intraventricular tumor in young adults. Hassoun *et al.* pointed out that its clinical course is longer than traditional oligodendrogliomas of the cerebral hemispheres. However, it may be shorter because of its intraventricular growth and the subsequent rapid progression of increased intracranial pressure, as Martin has indicated.

Light microscopically tumor cells are small, round and clear with occasional calcifications, resembling those of traditional oligodendrogliomas. It is impossible to distinguish a central neurocytoma from an oligodendroglioma except for the presence of patchy fibrillary stroma in the former. There is no astrocytic differentiation of the tumor cells or mixed features with astrocytic cells. Electron microscopic examination reveals numerous synapses or synapse-like devices,

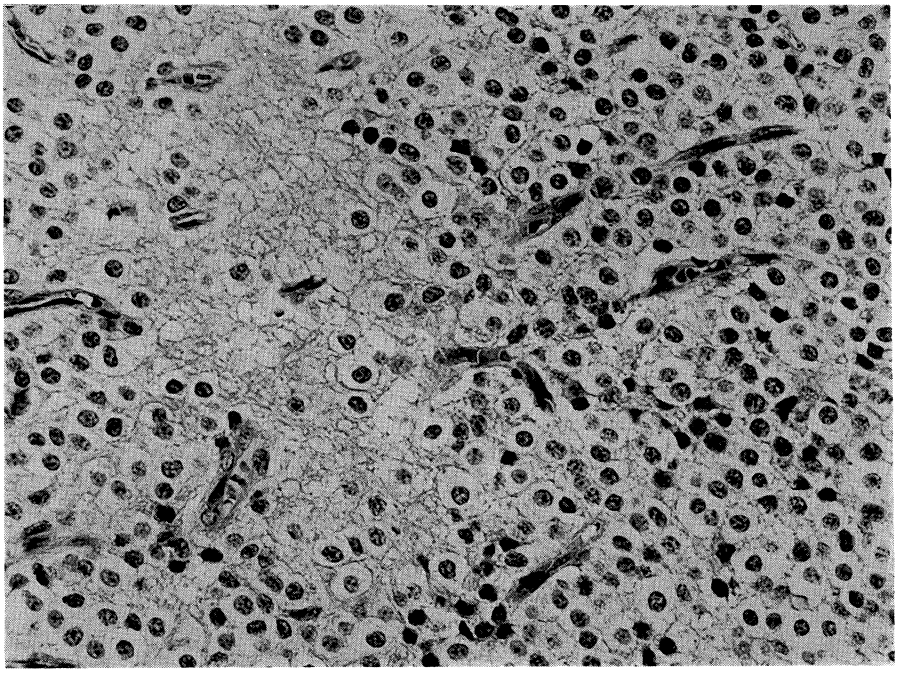

Fig. 2. Photomicrograph of the tumor cells of case 1 showing a honeycombed structure and patchy fibrillary stroma. HE, ×160.

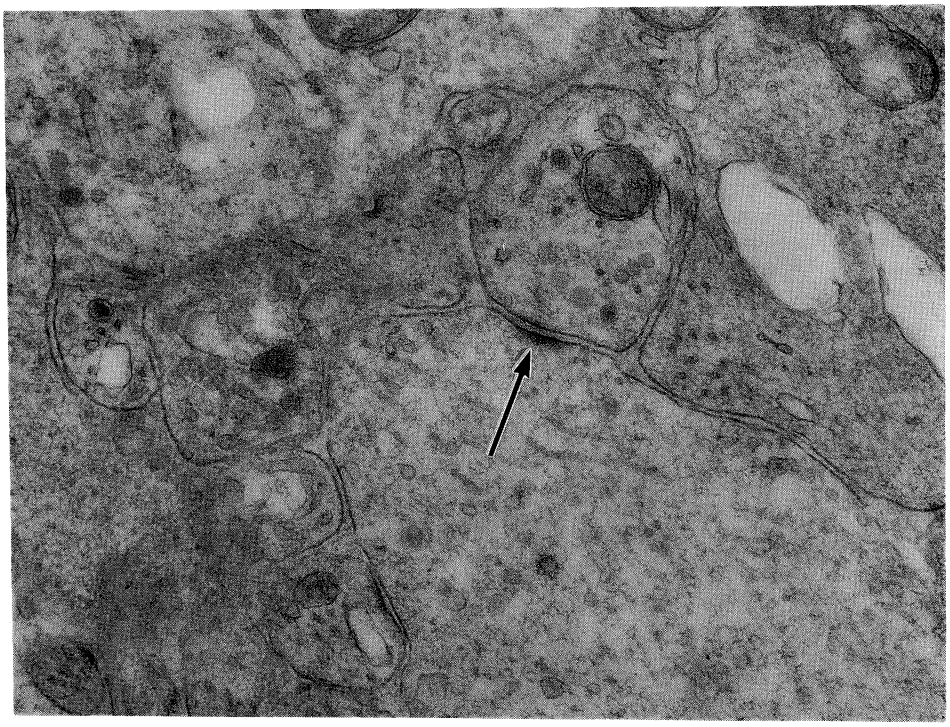

Fig. 3. Electron micrograph of the tumor cells of case 2 showing entangled cell processes with numerous microtubules, dense-cored and clear vesicles, and a synapse-like device (arrow). ×40,000.

axon-like processes containing many microtubules, and dense-cored and clear vesicles. These electron microscopic findings suggest that this tumor is potentially derived from neuronal elements or that it has a tendency towards neuronal differentiation.

Hassoun *et al.* presume that central neurocytomas may originate from the nucleus of the septum pellucidum because they occur in the ventricles around the septum pellucidum or near the foramen of Monro. The incidence of central neurocytomas is not clear, but there may be a number of cases, considering the fact that we found 2 cases of this tumor out of 7 cases previously diagnosed as oligodendroglioma. For an accurate diagnosis, it is essential to examine the tumor electron microscopically and locate the synapses or synapse-like devices, axon-like processes with microtubules, and dense-cored and clear vesicles which are characteristic of central neurocytomas.

That this tumor is derived from neuronal elements as Hassoun *et al.* have suggested, is not certain, since it is questionable whether the synapse-like processes are true synapses of the neurons. Clinicopathologically, however, it is undoubtedly different from traditional oligodendrogliomas of the cerebral hemispheres.

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