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Mixed Mesenchymal and Neuroectodermal Tumor of the Cerebellum

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Summary. An unusual cerebellar tumor composed of mixed mesenchymal and neuroectodermal elements in a 44 year old man is reported. The mesenchymal element is well-differentiated adipose tissue, whereas the neuroectodermal components exhibit a variety of medulloblastomatous, astrocytomatous, oligodendrogliomatous and ependymomatous areas. A possible origin of such unusual mixed neoplasms is briefly discussed.

Key words: Cerebellum – Mixed tumor – Lipoma – Neuroectodermal tumor

Intracranial "lipomas", or lipomatous hamartomas are relatively uncommon. An association with other non-neoplastic lesions such as neurodysraphic conditions and various congenital anomalies in the visceral organs has been noted (Budka, 1974; Koehl et al., 1970; Sperling and Alpers, 1946). However, the concomitant occurrence of lipomatous hamartomas and neuroectodermal neoplasms is decidedly rare (Budka, 1974).

Recently we have experienced a case of a 44-year old man with a large infiltrating tumor which was removed from the right cerebellar hemisphere. Microscopically the tumor was composed of a mixture of mature adipose tissue and a mixed neuroectodermal tumor; the diagnosis was confirmed by postmortem examination.

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Case Report

Clinical History

This 44-year old caucasian male was in good health until January 1973 when he subsequently developed unsteadiness of gait, left facial numbness, slurred speech and occipital headache.

Physical and neurological examination were unremarkable except for bilateral papilledema, rapid horizontal nystagmus with the fast component to the right, left facial weakness of the peripheral type, decreased sensation of the fifth nerve, an ataxic broad based gait and a positive Romberg test. A bilateral carotid arteriogram revealed large ventricles with placement of the basilar artery against the clivus suggesting a posterior fossa mass. A vertebral arteriogram showed a mass lesion in the right posterior fossa which appeared to be extra-axial.

Posterior fossa exploration revealed a mass in the right cerebellar hemisphere. The tumor was soft, relatively avascular, and appeared to extend into the deep white matter. The patient expired 18 h after the operation.

Neuropathologic Examination. Upon opening the skull a large hemorrhagic mass occupied the right posterior fossa producing bulg-

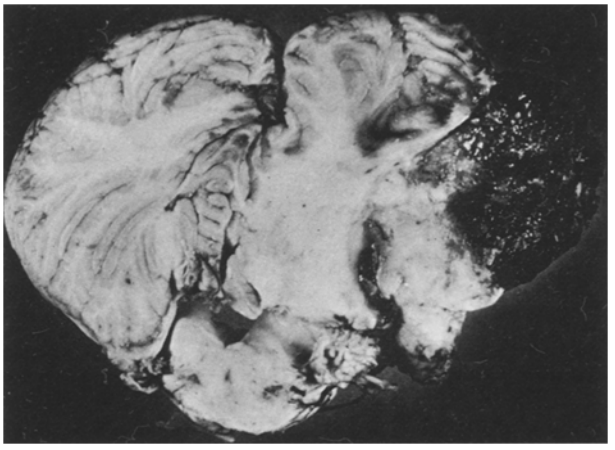

Fig. 1. Horizontal section of gross autopsy specimen demonstrating a mass lesion in the right cerebellar hemisphere, mostly extra-axial, but diffusely infiltrating the cerebellar tissue

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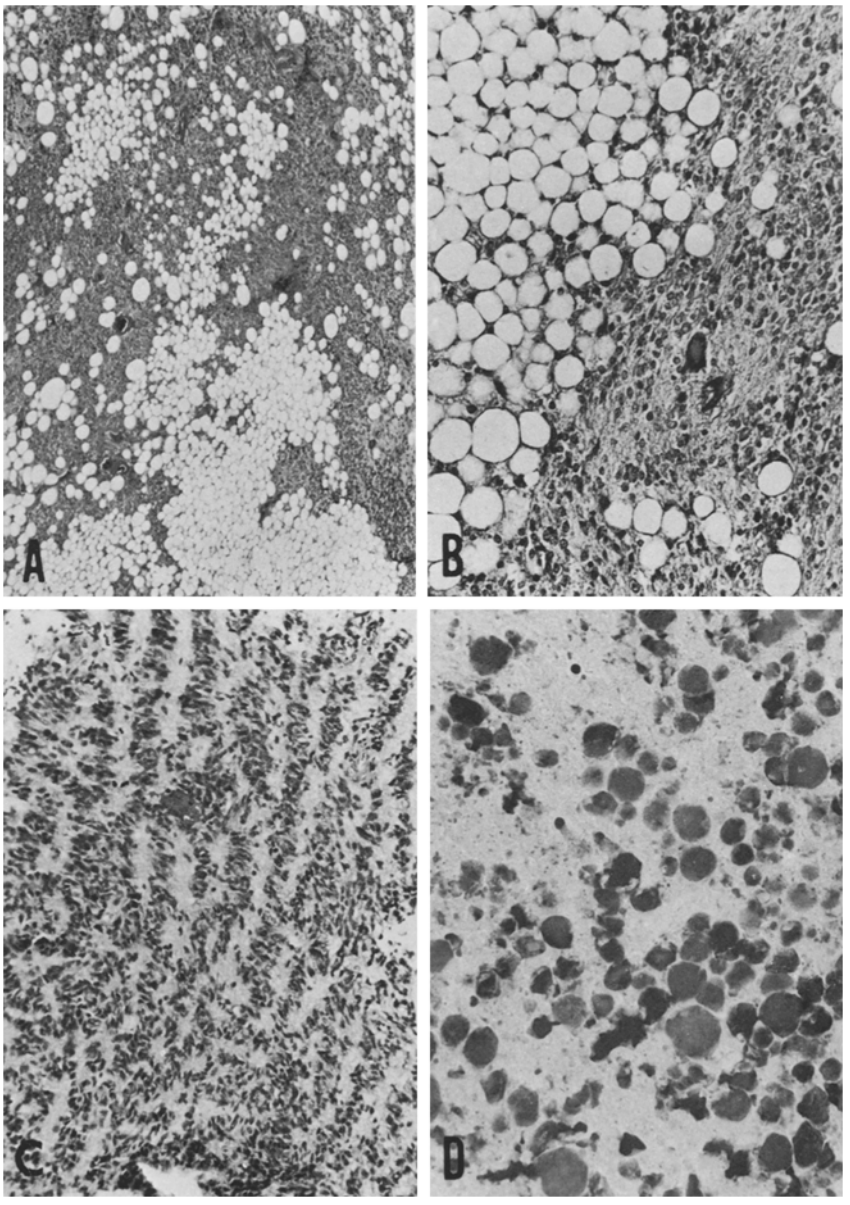

Fig. 2 A-D
Photomicrographs of the tumor.
A Autopsy specimen demonstrating the intimate mixture of the lipomatous and neuroectodermal components. H.-E., × 44. **B** Higher power of the same specimen to demonstrate the astrocytic component of the tumor. H.-E., × 177. **C** A medulloblastomatous area of the tumor showing palisading and rosettes. H.-E., × 133. **D** Sudan IV stain clearly indicates the presence of neutral fat in the lipomatous areas. × 177

ing of the tentorium. Prominent tonsillar herniation was attributed as the immediate cause of death. The fresh brain weighed 1525 g. Multiple coronal sections of the brain revealed a large tumor mass in the right cerebellar hemisphere, producing a marked shift to the left. The tumor was well delineated but with focal infiltration of the tumor into the deep white matter (Fig. 1). The tumor was elastic, partially semitransparent, with multiple areas of hemorrhage and necrosis. Microscopic examination of the tumor was accomplished with the help of the following stains: H.-E., Wilder's reticulin, Bodian, Nissl, PTAH, Masson's trichrome and Sudan IV. The tumor was composed of multiple islets of mature adipose tissue, confirmed by Sudan IV stain, separated and intermixed with a neuroectodermal tumor of mixed components (Fig. 2). Small elongated cells with glial processes, confirmed by a negative reticulin and trichrome stain and a positive PTAH stain indicating an astrocytomatous component (Fig. 2B) were noted. Other areas, which were prominent, showed sheets of small cells with hyperchromatic nuclei forming Homer Wright type of rosettes and nuclear palisading characteristic of medulloblastoma (Fig. 2C). Still in other areas, there is an oligodendrogliomatous element exhibiting honeycomblike cell patterns as well as perivascular arrangements of elongated cells with PTAH positive process not unlike those seen in ependymomas. The tumor diffusely infiltrates the right cerebellar hemisphere, and partially the middle cerebellar peduncle. Sections from the surrounding uninvolved cerebellum show only a focal Purkinje cell drop out with torpedo formation and a Bergmann's gliosis. No areas of microdysgenesis or pial aggregates of fat cells are noted. Electron microscopy on formalin fixed material was in agreement with light microscopy findings.

Discussion

Our histopathologic study indicates that the tumor has unusual mixed mesenchymal and neuroectodermal elements (Fig. 2), the former mature adipose tissue

while the latter is composed of medulloblastomatous, astrocytic, oligodendrogliomatous and ependymomatous areas. There is no clear histologic separation between these two main components. A question arises as to whether the tumor arose as a de novo mixed tumor or whether an inert mass of lipomatous hamartoma was subsequently invaded by a neuroectodermal tumor.

Combinations of intracranial lipomatous hamartomas with neuroectodermal tumors, though rare, have been previously described, in which the neuroectodermal tumors have been medulloblastoma (Budka, 1974), spongioblastoma (Henschen, 1955), gliosarcoma (Nippe, 1912) and medulloepithelioma (Treip, 1957). In most of these cases the lipomatous hamartoma and neuroectodermal tumors are described as separate lesions where the latter may infiltrate the inert hamartoma. However, Nippe's case of lipoma-gliosarcoma and several reports of similar mixed tumors (Holimon and Rosenblum, 1971; Shuangshoti, 1973; Shuangshoti and Netsky, 1971) are not described as separate lesions and may suggest that such tumors could arise from areas of dysgenesis of endogenous tissue as de novo mixed tumors. It is interesting to note that some of these tumors have been associated with areas of dysgenesis (Budka, 1974). Considering the observation by Horstadius (1950) and Weston (1970) that the cranial mesenchyma is derived from the neuroectoderm, it is a distinct possibility that mixed mesenchymal and neuroectodermal tumors could originate from the same germ layer, i.e., neuroectoderm. Some investigators, however, consider that displacement of anlage tissue during closure of the neural crest may be responsible for these unusual neoplasms of mixed cellular elements (Budka, 1974; Holimon and Rosenblum, 1971; Sperling and Alpers, 1946).

Although portions of the tumor in our case may be confused with Enterline's group III "lipoma-like" type of liposarcoma (Enterline, 1960) there are no areas of cellular immaturity or nuclear atypia in the lipomatous areas. Instead, mature adipose cells are intimately mixed with neuroectodermal elements. This would suggest that the neoplasm may have originated from an area of dysgenesis in which two distinct cellular elements co-exist in close proximity.

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