Primary Sarcoma of the Reticuloendothelial System of the Brain

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This report describes the morphological features of a primary intracerebral neoplasm which warrants classification as a reticulum cell sarcoma. Evidence is presented to demonstrate that this tumor arose from the adventitial histiocytes and provides an exceptionally clear example of the distinctive origin of this type of neoplasm. Criteria useful for recognizing this type of tumor and necessary for its diagnosis are presented.

Case Report

The patient was a 70-year-old white woman who was transferred to Mercy Hospital on March 10, 1964, in coma. There was a past history of gradual mental deterioration, slurring of speech, headaches, and vomiting for several months before her lapse into coma, which occurred 2 weeks before admission.

Examination. The neurological examination showed a comatose elderly woman who responded only to painful stimulation by weakly moving all 4 extremities. There were no lateralizing signs and no papilledema. The Babinski extensor response was present bilaterally. A lumbar puncture done 4 days later, revealed slightly xanthochromic spinal fluid with an opening pressure of 200 mm. of water. The fluid contained 216 mg. of protein per 100 ml. and 3 lymphocytes. Her level of consciousness varied from day to day, but was never normal. Ventriculography revealed a moderate enlargement of the lateral ventricles without displacement. Air was present in the subarachnoid space over the cerebrum and cerebellum but not within the cerebral aqueduct or 4th ventricle. Pneumoencephalography did not visualize the ventricular system.

Course. Coma increased and she developed lateral and vertical nystagmus and quadripareisis more marked on the left side. She died on May 22, 1964, 24 months after admission.

Post Mortem Examination. The cerebral hemispheres appeared roughly symmetrical and edematous. On coronal sections the brain showed marked symmetrical dilatation of both lateral ventricles, and slight dilatation of the 3rd ventricle. In the region of the basal ganglia and hypothalamus there were two independent slightly hemorrhagic tumor nodules 1.5X2.5 cm. on each side. These extended from the anterior cerebral commissure back to the hypothalamus for a distance of 3.5 cm. The cerebellum contained a large, fairly well circumscribed tumor in the left hemisphere. It measured 3X4X3.2 cm. and had almost completely obliterated the 4th ventricle. The tumor appeared to be the result of the union of two smaller tumor nodules. The cerebral aqueduct was patent.

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Fig. 1. Phagocytic tumor cells in the hypothalamus. Note abundance of mitotic figures and prolongation of the cytoplasm in some of these cells. (H. & E. X375.)

Microscopic examination of the various tumor nodules disclosed a similar cytologic pattern. The tumor cells were moderately pleomorphic. Most had an elongated indented nucleus with a prominent and dispersed chromatin network, indistinct nucleoli, and a moderate amount of eosinophilic, sometimes vacuolated, cytoplasm. Frequently irregular, pseudopod-like projections of the cytoplasm were noted which were suggestive of a phagocytic type of activity (Fig. 1), and ingested particulate material was present in an occasional cell. Interspersed between the tumor cells there were numerous rod cells and smaller cells with round hyperchromatic nuclei and small amounts of eosinophilic cytoplasm quite similar to reactive microglial cells present at the periphery of the tumor. There were numerous small areas of necrosis and large numbers of abnormal mitotic figures. In some areas of tumor, the overlying leptomeninges appeared infiltrated by tumor but there was no evidence of diffuse meningeal involvement. Microscopic examination of other areas of the brain showed the tumor to be multifocal, with numerous large and small foci of tumor present in the corpus callosum, medulla, and cerebrum. None of these areas showed abnormalities on gross examination. The different, independent foci of tumor were all similar in cellularity and degree of malignancy. Frequently the tumor cells were seen arising from the adventitial layer of small and medium size blood vessels (Figs. 2 and 3) as well as from capillaries (Fig. 4).
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Tumor cells stained by Penfield's modified silver carbonate method for microglia and oligodendroglia, revealed no cytoplasmic metallophilia, although the nuclei were strongly argyrophilic. No glial fibrils were observed after staining with Mallory's phosphotungstic acid hematoxylin. However, strands of coarse and fine reticulin fibrils generally unrelated to blood vessels were observed in different areas by Gomori's reticulin stain (Fig. 5). There were no foci of tumor elsewhere in the body, and there was no evidence of a primary tumor in other organs which could have given rise to cerebral metastases. Thus, the cerebral origin of the neoplasm was established beyond reasonable doubt. Pathological findings in other organs included bilateral pneumonitis with mild emphysema, fibrous pleural adhesions, pulmonary edema, and arterionephrosclerosis.

Discussion

The reticulum cell sarcoma is a malignant tumor which stems from the reticulum cell or primitive histiocyte. In the central nervous system there are three representatives of this cell series. These are the microglial cells, the cells present in the adventitial sheaths of blood vessels,