Reticuloendothelial Sarcoma of the Brain

Report of Four Cases

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In 1963 Burstein, et al., reported 41 cases of a group of tumors originating from the reticuloendothelial system of the brain. The histological picture of these neoplasms indicated that the cells originated from microglial, leptomeningeal, and perivascular histiocytes. Although these neoplasms have been described in the medical literature under many different titles, such as perithelioma, primary mesenchymal tumor, and microgliomatosis, their histologic picture is the same, and the argument as to the proper classification has been considered one of semantics rather than true differences in histological character. Recent comparative electronmicroscopic studies of tumor cells from a case of reticuloendothelial sarcoma and non-neoplastic microglia revealed many mutual subcellular characteristics.

A review of the autopsy files of the Department of Pathology of the General Hospital of the Medical Center of the Mexican Social Security revealed four cases of these tumors in a period of 4 years (1963 to 1966). They represent 4% of a total of 100 primary intracranial tumors seen in the same period. The diagnosis of reticuloendothelial sarcoma was based upon a complete postmortem examination.

The attention that has recently been given to these relatively uncommon neoplasms prompted us to report the following cases in an attempt to contribute to a better understanding of the true incidence of these tumors.

Case Reports

Case 1. A 27-year-old man was admitted to the hospital because of headaches, double vision, dizziness, and unsteady gait of 6 weeks duration. The patient had been an alcoholic for the past 10 years and his illness began after a severe alcoholic bout.

Examination. The patient was thin, dehydrated, stuporous, and disoriented. There was slight blurring of the optic discs in both eyes although physiological cupping was present and the blood vessels were normal. The pupils were miotic, slightly irregular, and did not respond to either light or accommodation. The patient had almost complete bilateral ptosis of the eye lids; it was impossible for him to open his eyes voluntarily. Both eyes were fixed in abduction, and he could not move them in any direction. Upon stimulation of the cornea, both eyes lacrimated. He perceived painful stimuli on the face, but facial movements were diminished on both sides. Swallowing was difficult. There was a spastic quadripareisis. Reflexes were equally exaggerated in all four extremities. There was an extensor plantar response on both sides. Sensitivity to painful stimuli was preserved.

Laboratory examinations of blood and urine were normal; Wasserman test was negative. Spinal fluid pressure was 150 mm of water, and chemical and cytological studies of the spinal fluid were normal. Skull and chest x-ray films were normal. An electroencephalogram showed marked alterations over the cortex and elements of lateralization to the left hemisphere. There was no response to optokinetic nystagmus, and a caloric test at 30°C showed horizontal nystagmus of increased amplitude and duration. The patient rapidly became stuporous and died 8 days after admission.

Autopsy. The brain weighed 1330 gm and there was flattening of the gyri, narrowing of the sulci, and bilateral uncal herniation. There was a firm mass, 1.5 cm in diameter, in the right frontal lobe and attached to the falx cerebri. Coronal sections of the cerebral hemispheres revealed symmetrical dilatation of ventricles. Horizontal sections of the brain stem at the level of the aqueduct of Sylvius revealed a gray tumor mass that originated from the lamina quadrigemina, infiltrated the brain stem, and reached the lower end of the
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cerebral peduncles (Fig. 1). The pons was not grossly affected, and the spinal cord was normal.

The neoplasm was composed of small round cells with hyperchromatic nuclei and scarce cytoplasm; the cells were arranged around blood vessels and filled the Virchow-Robin space. In some areas, definite neoplastic cells were seen away from the perivascular space infiltrating the adjacent brain tissue. The tumor filled the perivascular spaces near the ependymal layer of the aqueduct (Fig. 2 left). The pineal body was also involved by the neoplasm, and the neurons of the brain stem were spread apart by the cells (Fig. 2 right). Reticulum stains revealed abundance of fine and coarse reticulum fibers, mainly in the perivascular spaces but also within the rest of the tumor mass. Occasional calcific deposits were present.

The small round tumor nodule located in the falx cerebri contained nests of meningothelial cells characteristic of meningioma. The rest of the necropsy was not significant.

Case 2. A 39-year-old man was admitted to the hospital in a comatose state.

His illness had begun 2½ months prior to hospitalization with left-sided headache, progressive weakness of the right leg and arm, and partial aphasia. His physician noted spastic right-sided hemiparesis, normal fundi, 5/10 visual acuity bilaterally, and normal confrontation visual fields. The spinal fluid then showed 40 cells, normal proteins and glucose. Skull x-ray films were normal.

Examination. Upon arrival at our emergency room the patient was comatose; the pulse was 60, blood pressure 120/80 mm Hg,