Malignant Reticulosus Limited to the Central Nervous System

Case Report

J. L. WILLIAMS, M.D., AND H. J. PETERS, M.D.
Department of Surgery (Neurosurgery) and Department of Pathology, Medical College of Georgia, Eugene Talmadge Memorial Hospital, and the Laboratory Division of the Veterans Administration Hospital, Augusta, Georgia

Reticulum cell sarcoma of the brain is a rare tumor usually occurring as a single lesion. We are reporting the unusual instance of widespread lesions in the central nervous system.

Case Report

Examination. This 42-year-old white man was admitted because of a confused mental state and left-sided seizures of 3 months duration. He was stuporous and responded only to painful stimuli. There was no papilledema. The only detectable cranial nerve deficit was a left central facial palsy. There was hyperreflexia of the left arm and both legs, bilateral ankle clonus, and bilateral Babinski signs. Examinations of the blood and urine were normal except for mild proteinuria. Analyses for serum electrolytes, blood urea nitrogen, porphyrin, lead and febrile agglutinins were normal. Lumbar puncture showed a spinal fluid pressure of 270 mm. The fluid was clear, colorless, and contained no cells; the total protein content was 98 mg%, and a colloidal gold curve was 01232100. Spinal fluid cultures were sterile. Bilateral carotid arteriograms were normal; pneumoencephalography suggested a mass involving the corpus callosum and widening of the septum pellucidum (Fig. 1).

The patient remained stuporous and febrile; in spite of a tracheostomy and antibiotic therapy, he continued to deteriorate and died within a few days. No definite clinical diagnosis could be made, except for suspicion of a diffuse infiltrating lesion of the anterior portion of the corpus callosum.

Autopsy. The brain weighed 1570 gm, the gyri were flattened, and the sulci obliterated. There was a pressure cone over both uveal regions, greater on the right. The right mammillary body was markedly flattened and was rotated posteriorly. The pons was edematous and contained occasional petechial hemorrhages. No lesions of the medulla or cerebellum were seen. On midsagittal section, the anterior portion of the corpus callosum was markedly enlarged, the posterior portion being normal (Fig. 2). This was interpreted as a localized glioma of the cor-

Received for publication November 17, 1966.
pus callosum. The rest of the gross examination was normal except for lobular pneumonia.

**Microscopic Examination.** Representative sections of the brain and spinal cord showed a diffuse mononuclear proliferation involving the perivascular spaces and the blood vessel walls (Figs. 3 and 4). The cells were of three types: 1) cells with rod-shaped nuclei and scanty cytoplasm, resembling microglia, 2) larger cells with lobulated vesicular and often multinucleated nuclei, and 3) cells closely resembling normal lymphocytes. Special stains revealed tumor cells lying dispersed between reticulum fibers in the walls of the blood vessel and perivascular spaces (Fig. 5). The proliferations were most pronounced in the corpus callosum, basal ganglia, and pons, but were also present throughout the central nervous system including the hemispheric cortices, cerebellum, medulla, and cervical spinal cord (Fig. 6). No other organs were involved.

**Discussion**

The diagnosis of malignant reticulosis implies that the process is derived from the reticular tissues. Central nervous system cells producing this kind of tumor are probably derived from cells of the perivascular or Virchow-Robin space, adventitial sheaths of the blood vessels, and microglial cells. There is some evidence to indicate that microglia may be derived embryologically from the pia or adventitia of the blood vessels of the brain. Russell, using aseptic puncture wounds in the cerebrum, demonstrated the phagocytic activity of microglia and also that the cells lying in the adventitia of the blood vessels and leptomeninges would incorporate trypan blue dye into their cytoplasm. In tissue cultures utilizing silver-carbonate impregnation of fixed tissue, Dunning and Furth demonstrated that microglia and histiocytes, such as the reticulum cells of the spleen and lymph nodes, and the Kupffer's cells of the liver, are morphologically and functionally identical. It is therefore assumed that they are a single cell type derived from primitive mesenchyma.

In the case we are reporting there were numerous reticulum cells as well as a spectrum of cells resembling lymphocytes and microglia; these cells were numerous in the perivascular spaces and the adventitial layer of the blood vessels, and often formed diffuse proliferative masses in the brain. This observation has led us to believe that the process could have arisen concomitantly from the adventitia of the blood vessels and from cells of the reticuloendothelial system within the brain.

Proliferation of reticulum cells throughout the brain and spinal cord without involvement of other organs has only occasionally been reported. Yuile reported the complete autopsy study of reticulum cell sarcoma arising in the brain. Abbott and Kernohan

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**Fig. 3.** Diffuse mononuclear perivascular proliferation involving the walls of blood vessels and brain substance. ×275

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FIG. 4. High-power view of the proliferation involving one blood vessel wall and its perivascular space. X1100

FIG. 5. Reticulum stain. The reticulin fibers are separated by proliferation in the wall of the blood vessel. X1100
reported 12 cases of primary sarcomas of the brain, only one of which represented diffuse perivascular sarcomatosis involving nearly every portion of the brain and spinal cord. Burnstein, et al., reported 242 cases of primary sarcoma of the brain, found that 41 represented tumors from the reticuloendothelial system.

The mental symptoms in our case could be explained either on the basis of the diffuse perivascular proliferations throughout the cerebral cortex or by the mass in the corpus callosum. The progressive left hemiparesis, focal seizures of the left leg, and bilateral Babinski responses were most likely due to the diffuse cellular proliferations involving the cerebral cortex and descending brain stem pathways. The diffuse changes noted in the electroencephalogram, the widening of the septum pellucidum in the pneumoencephalogram, and the elevated cerebrospinal fluid protein in the absence of increased numbers of cells may be considered evidence of diffuse involvement and should be an indication for brain biopsy.

Edgar and Dutcher reported a case of cerebral reticulum cell sarcoma associated with macroglobulinemia and suggested identity with the Bing-Neel syndrome (macroglobulinemia with central nervous system symptoms). Unfortunately, a serum protein determination was not made in our case. The short survival period in our case has been similarly reported by others.

Summary

We have reported a case of diffuse proliferation of the reticular tissue of the brain (malignant reticulosis). The cerebral hemispheres, corpus callosum, basal ganglia, brain stem, and cervical spinal cord were all involved.

References


FIG. 6. High-power view of the various cells seen in this proliferation within brain substance. X4400