Primary Mesenchymal Chondrosarcoma of the Cerebrum

Report of a Case

ROBERT RASKIND, M.D., AND STANLEY GRANT, M.D.

Departments of Neurological Surgery and Pathology, The Permanent Medical Group and Kaiser
Foundation Hospital, Oakland, California

Except for the meninges, the various component tissues of the central nervous system are rarely the site of sarcoma. Davis and Davis calculated the incidence of various types of brain tumors in 3 large series, and noted that among 2996 microscopically verified intracranial tumors (1215 from the Northwestern University Surgical Library; 1781 from the Cushing series), 31 were metastatic sarcomas, 1 meningeal sarcoma, and 25 unclassified tumors; among the 5250 verified brain tumors of Olivecrona's series, no primary intracranial sarcoma is noted; among the 6000 histologically verified brain tumors in Zuleh's series, 2.7 per cent were primary intracranial sarcomas. Zimmerman et al. indicated that malignant tumors of mesenchymal origin rarely arise from the leptomeninges covering the surfaces of the cerebrum or from these membranes in the subleia, but did not mention a specific rate of incidence or site of predilection.

Russell and Rubinstein discussed the controversy respecting the term "primary sarcoma" of the brain. Although there are difficulties and inconsistencies that cannot yet be resolved, it seems evident that parenchymal sarcomas (neoplasms that are neither metastatic sarcomas nor malignant meningiomas) of the cerebral hemispheres do occur. The following case is presented in substantiation of this thesis.

Case Report

A 48-year-old woman had spontaneous subarachnoid hemorrhage without localizing or lateralizing signs. After her recovery from an initial period of lethargy and nuchal rigidity, there were no persistent neurological findings. No abnormality was noted on carotid angiography, and the patient was released from the hospital. Headache recurred and a second angiogram was performed which again showed no evidence of abnormality.

The patient appeared moderately well until 3 months after the first episode, when severe generalized and occipital headache was followed by lethargy and vomiting. A physician who examined her in her home advised hospitalization but the patient declined. Twelve hours later she became unresponsive and was brought to the hospital's emergency room, deeply comatose, with nuchal rigidity (3 plus), general flaccidity and hyporeflexia. Both pupils were fixed; the diameter of the right was 7 mm.; of the left, 4 mm. In the right eye there was papilledema (2 diopters); 2 fresh, flame-shaped retinal hemorrhages were seen, but there was no subhyaloid hemorrhage. The left optic disc could not be clearly seen but there appeared to be some degree of papilledema; no hemorrhages were noted in the retina of the left eye. Angiography gave evidence of a large space-occupying lesion in the right frontal area.

Operation. Emergency craniotomy revealed a large subcortical hemorrhage surrounded by necrotic tissue. A greyish brown, gelatinous mass lined the walls of the cavity from which this clot was evacuated. Several pieces of the tissue were submitted for microscopic examination. The anterior third of the frontal lobe was resected in order to effect internal decompression. The patient failed to rally following the procedure and died on the third postoperative day.

Autopsy. The biopsy specimen showed only a minute portion of cellular tumor, insufficient in amount for definitive classification. At autopsy the skull was unremarkable. In the right frontal area there was a small epidural hematoma. The meninges showed no evidence of tumor. In the right frontal lobe, an irregularly shaped cavity 8 cm. in diameter was filled with blood clot. No tumor was grossly identifiable. Sections from the cavity wall showed multiple areas of tumor, characterized by 2 components (Figs. 1-3): (1) The predominant compo-
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Fig. 2. Spindle cells with adjacent area of astrocytes: ×250.

Fig. 3. More closely packed area of spindle cells: ×250.

ment was of spindle cells, uniform in size and shape, of moderate size, showing no mitotic figures, with elongated moderately chromatic nuclei. The tumor cells were arranged in interlacing bundles. (2) Scattered through the spindle cell stroma were small, round to oval nests of cartilage. Slight transitions were seen between the spindle-shaped cells and the cartilage cells. The tumor merged with cerebral parenchyma. Slight gliosis was present. Masson trichrome stain showed no collagen formation between the spindle cells of the tumor. Phosphotungstic acid-hematoxylin stain revealed no glial component. The appearance of the tumor was considered to be consistent with that of a so-called "mesenchymal chondrosarcoma."

There was, in addition, a 4×4×4 cm. mass in the left lobe of the thyroid gland, having the histologic characteristics of a microfollicular adenoma. The left lower lobe of the lung showed chronic bronchitis with bronchiectasis and cystic changes. The uterus contained small leiomyomata.

Comment

Mesenchymal chondrosarcoma, a recently described malignant neoplasm, usually occurs in bone. The tumors have a distinctive appearance, characterized by a predominant mesenchymal stroma containing small nests of mature cartilage. One such tumor, which occurred intracranially, was attached to the dura and depressed the underlying brain, but did not invade the cranial bone. An intracranial tumor having a fibrochondral sarcomatous component resembling mesenchymal chondrosarcoma together with a gliomatous component (gliosarcoma) has also been described.

Summary

We have reported a case of primary mesenchymal chondrosarcoma, occurring in the frontal lobe of a 38-year-old woman. The meninges were not involved. Although primary sarcoma of the brain is rare and the authenticity of reported cases has been disputed, this instance presents additional evidence that the entity does indeed occur.

Dr. N. Leonard Morgenstern, Chief of the Department of Pathology, and Dr. Melvin Friedman, Senior Consulting Pathologist, Kaiser Foundation Hospital, Oakland, California, reviewed the microscopic sections.

References

4. Lichtenstein, L., and Bernstein, D. Unusual benign and malignant chondroid tumors of bone. A survey of some mesenchymal cartilage tumors and