CASE REPORTS

PRIMARY SARCOMA OF THE CEREBELLUM (CEREBELLAR SARCOMA) WITH EXTRACRANIAL METASTASES

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Primary intracranial neoplasms rarely metastasize outside the central nervous system, and reports of such occurrences often are disputed. Winkelman et al.28 reviewed 48 cases reported up to 1952 and concluded that only 9 of these metastatic tumors were primary in the brain: 8 of those they accepted were meningeal sarcomas, and 1 was a malignant hemangiendothelioma. To these they added a fibromeningioma of the cerebrum metastatic to the lung. Since their review there have been reports of metastases from 16 meningiomas11,17,25 and 23 gliomas. Six of the gliomas were ependymomas,14,15,20,21,26 9 were medulloblastomas,4,18,23 7 were undifferentiated gliomas8-12,29,30 and 1 was an oligodendroglioma.16 No attempt is made in this paper to establish the validity of any of the 87 intracranial neoplasms alleged to have metastasized. Significantly only 1 sarcoma of the cerebrum27 (a case not accepted by Winkelman et al.) and no sarcomas of the cerebellum have been reported to metastasize. Hsiu19 stated that although sarcomas of the cerebrum grow rapidly and frequently contain numerous mitotic figures, he had never known one to metastasize, and none of the 10 cases of sarcoma of the cerebellum examined in a review of the literature by Ley and Rosendo19 metastasized.

Zimmerman et al.21 stated that sarcomas of the cerebellum not uncommonly metastasize to lung, lymph nodes and bone marrow. Since we are aware of no such previously reported cases, we are presenting a case of sarcoma primary in the cerebellum with metastases to lung, liver, and bone marrow.

CASE REPORT

A 48-year-old boy began to have bilateral headaches, nocturnal vomiting, anorexia and lethargy approximately 1 month prior to his admission at the University of Chicago Clinics. Several days after the onset of symptoms he was hospitalized and fitted with glasses to correct a left esotropia. After discharge he continued to vomit, lost 18 pounds, became weaker and ataxic, and had paresthesias in the arms and legs.

Examination at this hospital revealed a thin, well-developed boy who was aware of his surroundings but lethargic. Macewen's sign was present and the circumference of the skull was 58 cm. Pupils were equal and reacted to light and in accommodation. The optic fundi showed 14 diopeters of papilledema and mild hemorrhages. Ptosis was evident and outward motion of the left eye was limited. Sustained horizontal nystagmus was present. There were no other abnormalities of the cranial nerves. The patient was right-handed and his strength was equal in both arms. No muscular atrophy or involuntary movements were noted. Finger-to-nose test was performed with slight symmetrical ataxia and alternating movements were slow. Babinski's sign was equivocal on the right. Deep reflexes were difficult to elicit. Truncal ataxia and an unsteady gait were striking. Laboratory findings were normal except for a count of 17,700 white blood cells. Dinstasis of sutures was noted on films of the skull. Ventriculography revealed a posterior-fossa tumor indenting the 4th ventricle posteriorly. The ventricles were dilated proximally.

Operation. Three days after admission a surgical exploration of the posterior fossa was performed. The cerebellar tonsils were below the arch of the atlas and the vermis was occupied by large, solid, firm, grey, vascular tumor approximately the size of an "egg." It extended deep into the mid-cerebellum and right hemisphere, but grossly was not attached to the meninges. A "walnut-sized" portion of the tumor in the vermis was removed. Several episodes of hypotension prevented further operation in this area. The cerebellar tonsils were separated and aspirated to permit decompression, and the tumor was noted to extend to the roof of the 4th ventricle. The bone flap was not replaced, the dura mater was left open and the muscle and skin were closed in layers.

Postoperative Course. The immediate course was uneventful. No definitive histological diagnosis was made by the Department of Neurosurgery, although medulloblastoma was considered. Eight days following operation intermittent vomiting developed which was followed by difficulty in swallowing and by palsy of the 6th nerve several days later. Beginning 2 weeks after operation, 3500 r to the skull and 2300 r of irradiation to the spine were delivered over a 5-week period. The patient was discharged in 4 weeks, slightly improved.

Subsequent Course. Three weeks after discharge he

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was readmitted with a 3-day history of vomiting and clonic seizures with opisthotonus. The physical condition was essentially unchanged. One day after admission he became semicomatose and 3 hours later was discovered to be cyanotic and apneic. In spite of artificial respiration and external cardiac massage he expired.

Autopsy. Gross. A 4×4×3 cm. defect was present in the vermis of the cerebellum. The walls were formed by firm, grey-white tissue. The leptomeninges covering the ventral surface of the cerebellar hemispheres were opaque, and in areas fully obscured the folia. Coronal sections of the cerebellum revealed a solid, grey, firm, focally necrotic and hemorrhagic tumor replacing the mid-portion of the cerebellum. The 4th ventricle was compressed into a slit-like opening, but the tumor did not penetrate through the ependymal lining. The medulla beneath the 4th ventricle was compressed and distorted (Fig. 1).

On the surface and in the parenchyma of the liver were 13 firm, grey nodules, 0.4 to 0.6 cm. in greatest diameter. Other organs contained no gross metastases.

Microscopic. The tumor was composed of sheets and cords of cells with relatively small, round to oval, and slightly pleomorphic nuclei with clumped chromatin and prominent nucleoli (Fig. 2). The eosinophilic cytoplasm was scanty and in many areas indistinct. Numerous mitotic figures and occasional multinucleated cells were present. A stroma composed of fibers of reticulin surrounded individual cells and separated cords and parallel rows of neoplastic cells as demonstrated by Wilder's silver stain (Fig. 3). In several areas cells were palisaded around vessels (Fig. 4). Focal zones of hemorrhage and necrosis were present in the center of the tumor, and dense zones of gliosis surrounded the main mass of tumor. Invasion of the meninges covering the cerebellar hemispheres accounted for the grey-white opacity observed grossly. Tumor extended between the folia and in many areas into the molecular layer of the cerebellum (Fig. 5). The muscle and fascia

![Fig. 1](image1.png)

**Fig. 1.** Coronal section through the cerebellum and rostral medulla showing tumor replacing the mid-portion of the cerebellum and compressing the 4th ventricle.

![Fig. 2 (left)](image2_left.png)

**Fig. 2 (left).** Neoplastic cells in single file arranged in parallel rows. In other areas sheets of cells exhibited no definite pattern.

![Fig. 3 (right)](image2_right.png)

**Fig. 3 (right).** Wilder's silver stain demonstrating reticulin fibers separating rows of neoplastic cells.
over the operative site also were invaded. Phosphotungstic acid-hematoxylin and cresyl violet stains failed to reveal glial fibers or neurons within the tumor. The nodules in the liver had a histologic pattern similar to that in the primary brain tumor and were interpreted as metastases (Fig. 6). A small solitary microscopic metastatic nodule was present in the lung (Fig. 7), and several microscopic metastatic lesions were present in...

Fig. 4. Neoplastic cells palisaded around a small blood vessel in the cerebellum.

Fig. 5 (left). Sections of the posterior surface of the cerebellar hemisphere revealed extensive involvement of the leptomeninges and penetration of tumor into the molecular layer.

Fig. 6 (right). Metastatic tumor in the liver with a histologic appearance resembling the primary cerebellar tumor.
the vertebral and sternal bone marrow (Fig. 8). There
was no invasion or destruction of bony trabeculae. Fine
fibers of reticulin were present between neoplastic
cells in the marrow. Histologic examination of the other
organs revealed no significant findings.

DISCUSSION

Prior to 1929 there was considerable confusion in
distinguishing between glial and mesodermal
tumors. Bailey defined the histology of cerebral
sarcomas and classified them into 4 groups: peri-
thelioma, perithelial sarcoma, fibroblastoma and
alveolar sarcoma. The last type was not estab-
lished definitely as a primary intracranial neo-
plasm but was included to aid in the differential
diagnosis of sarcomas and gliomas.

Since that time there have been several modifi-
cations of this classification (Hsi, Nichols and
Wagner, and Abbott and Kernohan). These
authors all included fibrosarcoma and perithelial
sarcoma. Abbott and Kernohan also described an
undifferentiated sarcoma and Hsi retained the
alveolar sarcoma as a distinct type. Abbott and
Kernohan found no tumors that fit the descrip-
tion of perithelioma and suggested that no such
neoplasm exists. Patterns others have described
as reticulum-cell sarcoma, alveolar sarcoma, and
fibromyxoma they considered as variants of fibro-
sarcoma. This classification does not include
malignant meningiomas that arise and grow on the
surface of the brain as opposed to a primary cere-
bral fibrosarcoma that develops within the brain.

Bailey originally suggested that cerebral sar-
comas arise from the leptomeninges. Although the
histogenesis of the leptomeninges is disputed, the
tumors arising from this tissue reproduce the
structures of mesodermal tumors elsewhere, and
Abbott and Kernohan stated emphatically that
these cerebral tumors of the meninges "are sar-
comas, regardless of the germ layer from which
they supposedly arose." Deep intracerebral
tumors may be accounted for by origin from the
meningeal covering which has been shown to
accompany blood vessels at least as far as the
tue capillaries.

Sarcomas arise in the cerebellum much less fre-
quently than in the cerebrum. Bailey et al. re-
ported 4 cases, Abbott and Kernohan 3, and
Ley and Rosendo. The average age of the pa-
tients was 17.7 years. In 6 of the 10 cases the
tumors occurred in children under 10, and males
predominated 4:1. The vermis and cerebellar
hemispheres were involved with almost equal fre-
quency.

Prognosis generally is poor because of the usual
deep location in the posterior cranial fossa and
frequent invasion of the 4th ventricle. Sarcomas
in the more superficial portions of the cerebellum may offer a better prognosis.  
These tumors, like sarcomas of the cerebrum, are grey-white, firm or fleshy and often well-circumscribed. Their histologic appearance is also similar to that of cerebral sarcomas. They are composed of round or spindle cells with small dark nuclei. Cytoplasm often is fragmented and indistinct, and a stroma of reticulin identifiable by silver stain sets the cerebral and cerebellar sarcomas apart from glial tumors of the central nervous system. They may exhibit the characteristic pattern of fibrosarcoma with a fibrillar collagenous stroma and spindle cells arranged in whorls or fascicles (Kernohan) or may assume a perivascular arrangement with small dark cells situated within rings of reticulin.

Zimmerman et al. discussed the cerebellar sarcomas separately in their Atlas of Tumors of the Nervous System. They considered the characteristic pattern to be cells with small, round, dark nuclei arranged in parallel rows separated by fibers of reticulin. Perivascular palisading and glomerular-like structures composed of clumps of tumor cells often are present. No reticulin is present within the glomerular structures but frequently surrounds the clumps of tumor cells. These glomerular structures may correspond to the alveolar forms reported by other authors.

CONCLUSIONS

Cross and Cooper observed microscopic metastases in the lungs from an undifferentiated glioma and from a malignant meningioma in 2 of 6 autopsied patients with primary intracranial tumors. From these results they suggested that perhaps the incidence of metastases from intracranial neoplasms is higher than is generally appreciated. At best metastases to sites outside the central nervous system are rare and one is constantly suspicious that the visceral tumor associated with a brain tumor represents an extracranial primary growth with a metastasis to the brain, or is a second primary neoplasm independent of the intracranial tumor.

Because of these uncertainties it is essential to the interpretation that the intracranial neoplasm be a recognizable brain tumor, and that the metastases are histologically identical with, or a usual variant of, the primary growth.

The histologic pattern of the cerebellar tumor in the present case fits the description of primary sarcomas of the brain. Its deep location and separation from the superficial meninges militates against the diagnosis of malignant meningioma. The location of the largest mass of tumor in the cerebellum, small circumscribed secondary deposits in the liver, and microscopic foci of tumor in the lung and bone marrow support the diagnosis of primary sarcoma of the cerebellum. The presence of only discrete foci of tumor in the bone marrow uniformly exhibiting the pattern of metastases argues against an interpretation of reticulum sarcoma of bone.

Russell and Rubinstein observed that distinguishing between polymorphic-celled sarcomas and poorly differentiated neuroblastomas may be difficult since they are similar histologically and both may contain abundant fibers of reticulin and collagen in the stroma. However, the neoplastic cells in our case did not resemble cells of neuroblastic lineage, appeared to possess no fibrillary processes after staining by the Holmes silver method, and characteristic rosettes were absent.

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REFERENCES


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