MALIGNANT ROUND-CELL NEOPLASM OF CEREBELLM
SIMULATING AN ANGLE TUMOR WITH PETROUS RIDGE EROSION*

JOHN M. MEREDITH, M.D.
Department of Neurological Surgery, Medical College of Virginia Hospital,
Richmond, Virginia

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The following case is unusual in that a malignant undifferentiated round-cell
tumor of the cerebellum in a man aged 33 years produced a typical angle tumor syn-
drome with unilateral 5th, 7th, and 8th cranial nerve involvement and ataxia on
the same side. It was thought pre-operatively that he probably had an acoustic
neurinoma, particularly as roentgenograms (Towne position) of the skull showed
erosion of the petrous ridge on the side of the cranial nerve involvement. However,
at operation, he proved to have a medulloblastoma or other malignant un-
differentiated round-cell tumor of the cerebellum originating on the side of the
cranial nerves affected. The tumor had scarcely, if at all, invaded the angle and its
nerves grossly, but seemed to cause the symptoms that it did by intrinsic cerebellar
and brain-stem involvement, more than by direct extension into the lateral angle.

CASE REPORT

R. T., a 33-year-old white man, was admitted to the Medical College of Virginia Hospital
on July 31, 1953, with a complaint that he had noticed deafness beginning in his left ear 3
years previously. This had gradually become more marked and 6 months before admission
headache had developed and unsteadiness with falling particularly to the left side was noticed.
For the last month or more, he had observed weakness of facial muscles and marked sensory
diminution in the left side of his face, and loss of taste in the left side of his tongue. These
complaints had all been progressive. In recent weeks he had been unable to walk without
fixing his eyes directly on the path in front of him or on his feet. There had been several epis-
odes of projectile vomiting during the 3 months before admission. He had fallen on several
occasions and had noticed occasional amblyopia which was of only transitory duration.

Examination. Nystagmus was present in all directions. There was a 6th nerve weakness
of the left eye, shown by inward squint of that eye. Hearing in the left ear was markedly im-
paired and there was definite sensory loss in the left side of his face, with diminution of the
corneal reflex. There was no choking of the optic discs. The left side of the face showed muscu-
lar weakness of peripheral type. There was a definite tendency to fall to the left side when
walking or standing. There was marked adiadokokinesis of the left hand and obvious ataxia
in the left heel-to-shin test.

The clinical diagnosis was probable acoustic neurinoma in the left cerebellopontine angle
with nearly total destruction of the function of the left 8th nerve. The otologist was unable to
instigate any vertigo with an unlimited amount of ice water in the left ear; on the right side,
20 cc. of ice water produced vertigo and nystagmus. Roentgenograms of the skull on Aug. 1,
1953, including a Towne position film, showed erosion of the left petrous ridge, suggesting an
acoustic neurinoma.

Operation, Aug. 3, 1953. Under endotracheal anesthesia, with the patient in the sitting

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position, a hemicerebellar approach was made. The cerebellum was moderately tense even though a cannula had been introduced into the posterior horn of the left lateral ventricle. A moderate amount of fluid was evacuated from the cisterna magna when the dura mater was first opened. A brain retractor was placed laterally under the left cerebellar hemisphere and this structure was then lifted upward and medially to expose the jugular foramen and the 9th, 10th and 11th nerves, which appeared normal. The 8th nerve was then seen clearly and there was no tumor present in that region. Because the cerebellum was rather full, it was decided to amputate the lateral third of the left hemisphere and in the process of carrying this out with a cautery and suction, a cavity (3 × 4 cm.) was entered into in the left cerebellar hemisphere from which exuded clear, yellow fluid, at least an ounce in volume. No tumor nodule such as an 8th nerve neoplasm could be palpated. After the cystic cavity had been evacuated, the cerebellum tended to fall away from the lateral sinus above; two large veins were coagulated immediately beneath the tentorium. The cerebellum was then retracted upward and medially and there was exposed a fleshy, somewhat fibrous but vascular tumor, about \(3\frac{1}{2} \times 4\) cm. in size, bluish-purple in color, which apparently arose in the cerebellar hemisphere itself and appeared grossly to be a glioma or other malignant invasive tumor. It definitely seemed to be contiguous with the subcortical cerebellar tissues rather than arising extrinsically (so far as the cerebellum was concerned) in the cerebellopontine angle. Several large portions of the tumor were removed for histopathological study and additional tumor was aspirated away from the site of the neoplasm. The dura mater was left open and closure was made in the usual manner without drainage.

Microscopic Examination. The tumor was a very cellular growth composed of small, dark-staining cells. It was quite vascular and hemorrhagic in one area. The diagnosis was malignant undifferentiated round-cell tumor: medulloblastoma or sarcoma (Fig. 1).

The original microscopic hematoxylin and eosin preparation of this tumor was kindly reviewed by Dr. J. W. Kernohan of the Mayo Clinic, who considered the tumor to be probably an atypical medulloblastoma. He stated, "It is my opinion that it is closer to a medulloblastoma than any other tumor in the nervous system with which I am familiar."

The postoperative course was stormy, possibly because of the fact that the bony and dural decompression had been carried out only over the left cerebellar hemisphere. Respiration was precarious for several days in spite of frequent ventricular taps. His temperature reached 103° and 104° on several occasions, but with the administration of roentgen therapy his condition markedly improved and he left the hospital 25 days after operation in good condition. Because of the precarious immediate postoperative state of the patient, only 500 r of x-ray therapy was delivered to the tumor itself, additional therapy being administered in another state 2 months later; a total of 1,900 r was given in October, 1953 and in January, 1954. Thus a total of only 1,700 r was given to the tumor in the first 5 postoperative months.*

We did not see or hear from him until, in response to our letter of inquiry, it was learned from his family that he had succumbed on Feb. 20, 1954, approximately 63 months after operative verification of the highly malignant undifferentiated round-cell tumor in the left cerebellar hemisphere.

COMMENT

Freid and Davidoff in 1951 described a pathological entity: primary cerebellar sarcoma, easily mistaken for medulloblastoma microscopically. The sarcoma may also "seed" into the spinal meninges and even metastasize, i.e., involve distant viscera outside the central nervous system, even the skeletal or lymphatic systems. They reported 4 such cases. However, their description of these sarcomatous tumors

* Preferred dosage: Using 4 circular skull and 2 long spinal portals and doses of \(250\) r (air) to 2 portals daily, totals of 1,000 r (air) to each portal in a series delivers approximately 2,000 r to the tumor bed. These series are repeated at 6-week and later at 3-month intervals over a 1-year period using the usual 1 mm. copper half-value layer. (Technique of X-ray Department, Medical College of Virginia, Richmond, Va.)