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

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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 syndrome 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 undifferentiated 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 episodes 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 impaired and there was definite sensory loss in the left side of his face, with diminution of the corneal reflex. There was no chocking of the optic discs. The left side of the face showed muscular 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

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 3½ × 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 61 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.)
MALIGNANT ROUND-CELL NEOPLASM OF CEREBELLUM

Fig. 1. (Above) There are diffuse sheets of compactly arranged cells with indistinct cytoplasmic borders, showing a superficial resemblance to lymphocytes. (Hematoxylin and eosin, X100.) (Below) The nuclei are hyperchromatic, crowded and irregular. There are no rosettes. The appearance is that of a malignant undifferentiated round-cell tumor. (Hematoxylin and eosin, X400.)

was preceded by at least two other reports. In 1929 Bailey described clearly these malignant intracranial mesodermal tumors. He believed that they arose from the leptomeninges or their derivatives and he considered it advisable to call them sarcomas, for their structure differs considerably from that of rapidly growing intracranial tumors that are certainly of neuro-ectodermal origin. He reported 8 cases in detail and referred briefly to others in his experience. It should be noted, however, that in none of his 8 cases was the lesion a more or less localized cerebellar tumor either in one or both cerebellar hemispheres, as in the case herein reported. They were all either cases of diffuse meningeal involvement so that the meninges grossly
resembled tuberculous infection, or they were masses in the cerebral hemispheres or basal ganglia.

In 1939, Bailey, Buchanan and Bucy\(^4\) described cerebellar tumors in children in whom the gross tumor as exposed at operation was indistinguishable from medulloblastoma and this was also true, to some extent, of the microscopic picture, according to these authorities. These latter statements are also valid for the tumor reported in this paper. They stated, "The course of events in these cases of malignant sarcomas is no different from the medulloblastomas." In describing one of their patients, a boy of 11 with a cerebellar tumor (Case 25, p. 107\(^2\)), they stated, in reference to the microscopic appearance of the tumor: "We believe it to be a sarcomatosis but admit the difficulty of differentiating the two tumors, i.e., sarcoma and medulloblastoma." They also described a 17-month-old girl (Case 26) with a multilocular cystic tumor of the cerebellum (quite similar to that in the case reported herein) which was, microscopically, a sarcoma of the alveolar type. The short postoperative survival of the patient herein reported suggests that he may have had a cerebellar sarcoma, although the insufficient postoperative x-ray therapy was also a major factor in his early death.

The unusual features of the present case were several. First, a verified medulloblastoma or sarcoma cerebelli in an adult beyond the age of 30 is a rather unusual finding. Secondly, to our knowledge, there have been no previous cases reported in the literature in which a histologically verified malignant undifferentiated round-cell tumor of the cerebellum in a child or an adult has produced or has been associated with erosion of the homolateral petrous ridge demonstrated in roentgenograms of the skull, precisely as one sometimes sees in cases of acoustic neurinoma. Hodes, Pendergrass and Dennis\(^5\) reported in 1951 on roentgenographic manifestations in 183 cases of cerebellopontine angle tumors of all types. In 16 of their cases the tumors were gliomas of the angle with cranial nerve involvement; in 1 of these 16 cases, an abnormal "petrous apex" was seen in the roentgenogram but they did not state whether it was a medulloblastoma or other round-cell malignant tumor as in our case, or one of the other types of glioma. Only 1 of their 183 cerebellopontine angle tumors was a medulloblastoma. In view of the unusual clinical features of the case reported in this paper, it is interesting to note that Hodes and his collaborators stated that gliomas in the angle seemed to occur more commonly in the younger age group, the average age of their 16 patients being 18 years, as compared with 40 years for patients with all other forms of angle tumor. Interestingly enough, Hodes and his colleagues found that none of the 16 gliomas in the angle were ependymomas, the microscopic diagnoses being astrocytoma, glioblastoma multiforme, spongiosablastoma polare, and medulloblastoma. In contrast, Kernohan, Woltman and Adson\(^7\) reported 10 cases of glioma in the angle in which 8 were ependymomas and the remaining 2 were fibrous astrocytomas, there being no instance of medulloblastoma or other malignant round-cell tumor (? sarcoma). Thirdly, the final unusual feature of the case herewith reported is that although there was obvious unilateral involvement, clinically, of the 5th, 7th, and 8th cranial nerves, grossly at operation this was not evident in the angle. The involvement must have been more medially or "nuclear" as the lateral angle was grossly free of tumor and the tumor-free nerves were clearly seen in the angle. We have recently had another patient (a 36-year-old white female) with a verified astrocytoma (grade 1) of the pons and cerebellum primarily, in whom there was also questionable erosion of the petrous ridge roentgenologically on
the side of the angle syndrome, which clinical picture was so suggestive of an 8th nerve tumor that operation was similarly performed in the erect or sitting position.

Concerning gliomas extending into the cerebellopontine angle, Kernohan, Woltman and Adson, as mentioned above, reported in 1948 a series of 10 cases of such neoplasms, none of which, however, was a medulloblastoma or other malignant round-cell tumor. According to these writers, to make a pre-operative diagnosis of glioma or other malignant tumor in this location (cerebellopontine angle) would be hazardous; however, several factors would support such a diagnosis. In 7 of the 10 cases of glioma in the angle reported by them, the patients were children and the same number were of the female sex. Neurofibromas and meningiomas, as they pointed out, occur also in the cerebellopontine angle, usually in the later years of life. Two of the 10 patients with glioma in the angle that they reported complained of unilateral deafness, as did our patient, and this finding was also relatively frequent in cases of meningioma of the angle, according to these authors. A number of the illustrations in their paper show large ependymomas confined, for the most part, to the cerebellopontine angle. We have had in our service at the Medical College of Virginia in the last 18 years 2 other cases (a total of 4 at present writing) of gliomas involving the cerebellopontine angle and producing primarily an angle tumor syndrome, including deafness, but all of these except the present case herein reported were characterized by gross invasion of nerves in the angle by the glioma as seen at operation. One was in an adult colored woman, aged 23 years (operated upon by our then resident, Dr. M. McKeel, on Feb. 8, 1954), who had an ependymoma as verified at operation, performed in the sitting position since the pre-operative clinical syndrome strongly resembled that of an acoustic tumor. The other was in a man of 30 operated upon (with recovery) in a 2-stage procedure 18 years ago by Dr. W. G. Crutchfield, then a member of our surgical staff. That patient had a medulloblastoma arising from a cerebellar hemisphere, which grossly invaded the angle and produced a syndrome very like that of the patient reported herein, namely, one highly suggestive of an acoustic neurinoma, with early deafness and unilateral ataxia and 5th nerve involvement. It was accordingly dealt with through a unilateral cerebellar approach, as one would attack an acoustic tumor today. An illustration of a gross brain specimen in Hicks and Warren's Introduction to Neuropathology shows a postmortem specimen very similar to the lesion in our patient. However, that individual was a 20-year-old male with a medulloblastoma in the cerebellum who had progressive right cranial nerve symptoms for 6 months, starting with deafness. It was stated that the symptoms first were very suggestive of an acoustic tumor. In that gross specimen, showing the base of the brain, the tumor is grossly invading the angle also (precisely as in 3 of our 4 cases of glioma of the angle cited above) which was not true of the patient reported in this paper, rendering the present case rather unique in that respect.

SUMMARY

(1) Medulloblastoma or other malignant undifferentiated round-cell tumor of the cerebellum in patients over 50 years of age is a rather rare lesion.

(2) Roentgenographic evidence of erosion of the petrous ridge on the side of the malignant round-cell cerebellar tumor producing the cerebellopontine angle syndrome is entirely unique in our experience, thereby stressing the rarity of this case and suggesting how a cerebellar gliomatous or sarcomatous lesion can, pre-opera-
tively, easily be confused with acoustic tumor, particularly when associated with early unilateral deafness.

(3) The production of an “acoustic neurinoma syndrome” by a malignant round-cell tumor of the cerebellum without gross invasion of any of the nerves in the lateral angle as demonstrated at the time of operation is likewise unusual in our experience and unrecorded, to our knowledge, before the present case. The more usual pathological situation in cases of cerebellar glioma or sarcoma of the angle producing an angle tumor syndrome is for the 8th, 7th, 5th and even additional nerves in the angle to be grossly involved by the glioma or sarcoma, as occurred in the other 3 similar cases of this type from our clinic briefly referred to.

REFERENCES