Fibrillary Astrocytomas, Cerebellar (1940) and Spinal Medullary (1966)

Case Report

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We are reporting a case in which sensory ataxia returned 25 years after removal of a cerebellar astrocytoma. Ultimately this was proved due to an astrocytoma of the lower cervical cord.

Case Report

In 1940 at the age of 18 years, this young woman was admitted with a chief complaint of left facial pain and instability of gait. She had suffered intermittent headache, neck pain, nausea, and vomiting for 15 months, and had noted diplopia on fixed gaze. She had had a normal menarche but subsequent menstrual irregularity.

Examination. There was no evidence of von Rechlinghausen's disease. The visual acuity was 17/30 bilaterally with bilateral papilledema. There was hypalgesia in the left trigeminal distribution with a depressed corneal reflex. The extraocular movements were conjugate, and there was no nystagmus. The gait was ataxic, more so with the eyes closed, with a tendency to veer to the left. The Romberg and coordination tests were satisfactorily performed. X-rays of the skull showed pressure atrophy of the dorsum sellae with a thinning of the floor of the sella (Fig. 1) and cephalic bulge of the clivus. In the midline of the posterior fossa approximately 2½ cm anterior to the internal occipital protuberance, there were several short, thick, curvilinear streaks of calcium clumped together in a disorderly fashion. In the Towne position, these could be seen as an irregularly rounded area of calcification that projected through the foramen magnum to the right of the midline.

Operation. A suboccipital decompression was performed with removal of the arch of the atlas and freeing of the cerebellar tonsils from the cervical canal. A cyst in the left cerebellar hemisphere was encountered with a ventricular needle at a depth of 1 cm. The 45 cc of xanthochromic fluid which escaped clotted on standing. Deep and medial within the cavity of the cyst, a mural nodule could be seen. Dissection of the cyst wall and the mural nodule led to the incisura of the tentorium. The operative impression was that of total excision of the tumor. Pathologically it was described as an astrocytoma made up of fibrillary astrocytes and "piloid" astroglia in the mural nodule and the cyst wall (Fig. 2). There were large lamellated calcospherites in one portion of the tumor, and adjacent to this portion there was arachnoidal fibrosis and pial vascular proliferation.

Postoperative Course. The convalescence was principally a matter of recovery from an initial accentuation of the preoperative symptoms. Secondary optic atrophy became apparent in both fundi. A paresis of the left external rectus appeared 4 years postoperatively and was increasingly apparent with near amblyopia of that eye. The vision in the right eye remained useful. Her principal difficulty was cerebellar dysfunction affecting the

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FIG. 1. Lateral x-ray of the skull, 1940, showing signs of increased intracranial pressure due to a partially calcified mass in the posterior fossa (arrow).
left arm and leg, but she quickly compensated for it. She married and raised three children.

Second Admission. In 1965, the patient became less stable on her feet and on a few occasions fell backwards. This difficulty was often precipitated while turning to the left and was particularly bad in the dark. She soon required assistance in walking, and was admitted to the hospital.

Examination. There was ataxia of both legs, with a broad-based gait, although she complained that she was having more trouble with the right leg than with the left. Vibratory and position senses were poor in both legs but intact in the arms. Touch and pinprick perceptions were normal. There was ankle clonus on the right, and the deep tendon reflexes in both legs were more active than those in the arms. The response to plantar stimulation was normal.

A mercury brain scan was negative. The spinal fluid protein was 56 mg\% with no elevation in the colloidal gold curve. There was free hydrochloric acid in the stomach, and the fasting blood sugar was 101 mg\%. There was no anemia, and the VDRL was non-reactive. Skull x-rays demonstrated the craniectomy defect and the clips from the previous operation. X-rays of the dorsal spine showed only intervertebral disc calcification at the ninth and tenth interspaces.

After the lumbar puncture, the patient described a sense of numbness and coldness in the left leg, and there seemed to be a reduction in the perception of pinprick in both arms and legs. Vibratory sense became lessened, and there was difficulty with stereognostic tests in the left hand. Fasciculations were visible in the thenar and hypothenar eminences of the left hand, and she described a sense of fatigue in the left arm. She began dragging the right foot when walking.

Third Admission. In 1966, a cervical and posterior fossa myelogram showed no evidence of a recurrent mass in the posterior fossa, but suggested a widening of the spinal cord in the cervicothoracic region. The protein was 50 mg\%. After this procedure there was hypalgesia in the ophthalmic divisions of the trigeminal nerves and in all segmental nerves below the head. The vibratory sense was reduced on all spinous processes below the C-5 level, and there was a partial Horner’s syndrome on the left. She then developed urinary incontinence and bilateral clonus at the knees and ankles. A Babinski sign developed on the right.

Myelography was repeated and confirmed the previous suspicion of an intramedullary lesion in the high thoracic and low cervical cord (Fig. 3). The spinal fluid protein was 46 mg\%.

Operation. Laminectomy of the lower two cervical and the first thoracic segments exposed a distended nonpulsating dura. Caudally the exposed cord was thinned by an intramedullary cyst which, when aspirated, yielded xanthochromic fluid (Fig. 4) that clotted on standing. The cyst was opened, and a mural nodule could be seen in its cephalad portion. With evacuation of the cyst there was still a widened appearance of the cord through the midportion of the exposure. The myelotomy was extended cephalad for 3 cm and the nodule removed. At either end the tumor appeared to be attenuated in a tubular strand which was centrally oriented to the region of the central canal. Silver clips were applied at the limits of the resection. Microscopically the tissue at these limits as well as in the cystic mass was interpreted as astrocytoma. Ventrally the tumor had surfaced to the pia in the midportion of the resection, and here it seemed to receive a vascular pedicle. This ventral tissue was actually white matter of the cord. There was fibrosis of the adjoining meninges, spinal nerves, and blood vessels. No neurons were

FIG. 4. Operative exposure in 1966 shows the intramedullary cyst caudally (arrow), and the cord widened by tumor in the midportion of the field.

FIG. 5. Photomicrograph of the fibrillary astrocytoma removed from the cervicothoracic cord in 1966 (H. & E., ×250).

seen, and tumor cells infiltrated these portions of the specimen. The tumor cells had small, round, vesicular nuclei while the cytoplasm was scant but homogeneous and formed a syncytium of closely arranged fibrillary matrix (Fig. 5).

**Postoperative Course.** The patient was immediately aware of more sensation in the trunk and legs and improvement in the appearance of the left eye. Urinary bladder function returned. Two months later she could walk with support and had position sense in both great toes. At 4 months she could walk with a cane. Vibratory sense remained absent, but the clonus was diminished and the right-sided Babinski less conclusive. She had been given 4000 r of cobalt therapy through a single posterior port measuring 12 cm in length, centered on the operative site.

**Discussion**

Multiple fibrillary astrocytomas and metastases from a fibrillary astrocytoma are rare. In the years 1928 to 1967 at the Yale-New Haven Medical Center there have been only two cases of cerebellar astrocytomas in which astrocytomas in other sites have also been encountered.

Dr. Gilbert Solitaire has reported the other case from our clinic; it was more protoplasmic and probably metastatic. The patient was a 15-year-old girl in whom a right temporal lobe tumor appeared 16 months following an
original operation for a small cyst and a large tumor nodule in the vermis of the cerebellum. The cerebral and cerebellar specimens showed similar neoplastic cells and architecture; in some regions of the cerebral tumor, protoplasmic astrocytes were the predominant cells.

In the Cushing series of 98 cerebellar astrocytomas, 12 were protoplasmic, but no multiplicity of site was noted. Among the 187 spongioblastomas of the cerebellum (the so-called "cerebellar astrocytomas") tabulated by Zülch, there was one case "of an astrocytoma-like glioma of the cerebellum... (which) spread diffusely via the cerebrospinal fluid. The tumor did not have the typical appearance of a spongioblastoma but more that of the protoplasmic astrocytoma." Zülch did not believe there was such a thing as true metastasis of an astrocytoma. Neither Earle at the Armed Forces Institute of Pathology nor Sayre at the Mayo Clinic indicated any such association of tumors.

Russell and Cairns reported a case of spinal metastases of an unoperated astrocytoma of the optic thalamus. Metastatic nodules were described in the arachnoid around the pineal gland, corpus callosum, and spinal cord. They were of identical structure with the primary fibrillary astrocytoma. In 1959, Perese, et al., described diffuse and intraventricular postoperative dissemination of a Grade I-II astrocytoma which was originally removed from the conus medullaris.

All three cases of multiple astrocytomas involving the cerebellum, described by Courville, were identified at postmortem examination. The history in each case was that of only a few months. The occurrence of a malignant astrocytoma of the frontal lobe, causing death 7 years after the removal of a cerebellar astrocytoma, opened the consideration of multiple astrocytomas separated in time.

In his tabulated review, Courville listed no case of multiple gliomas appearing in the cerebellum and the neuraaxis. The 20-year survival of 50% of Cushing's astrocytoma patients provided one of the best recorded series in which to witness the development of multiple tumors, and it is significant that none is recorded.

Astrocytomas of the spinal cord most commonly originate in the posterior aspects of the cervical cord. In our patient, the cervical tumor occurring 26 years after a cerebellar tumor removal and located within the substance of the cord was, in all probability, a second primary astrocytoma. The apparent extensions cephalad and caudad in the anatomical location of the central canal might raise the question of extension from the posterior fossa. The microscopic appearances of the cerebellar and the spinal tumors were not identical, although both were fibrillary astrocytomas.

**Summary**

In 1940, a girl then 18 years old had a huge, cystic cerebellar astrocytoma successfully removed. It was identified as a fibrillary astrocytoma, containing large calcospherites. She later married and raised three children; she was examined at intervals, without evidence of tumor recurrence.

In 1965, she complained of returning ataxia, and symptomatic progression led to the successful removal of a comparable fibrillary astrocytoma from the region of the central canal in the cervical cord.

**References**


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