Brainstem gliomas account for 10%–20% of all pediatric CNS tumors. Although they can occur at any age, brainstem gliomas are most common in children, with the mean age at diagnosis being 7–9 years, with no gender predilection. Children with brainstem gliomas often present with multiple cranial nerve deficits (particularly of the 6th and/or 7th cranial nerves), long tract signs (arm or leg weakness and hyperreflexia), and cerebellar signs (such as truncal ataxia or incoordination). Several classification systems have arisen to determine which tumors may benefit from operative treatment; the simplest system divides brainstem tumors into either focal or diffuse entities. Whereas focal lesions have been shown to benefit from surgery, diffuse gliomas are considered inoperable. This is unfortunate, because diffuse gliomas account for the majority (58%–75%) of all brainstem tumors. Despite advances in imaging and treatment modalities over the past 100 years, most children die within 18 months of diagnosis.

To our knowledge, the earliest written report of a brainstem lesion was published in 1881 by Dr. Charles Mills. He reported a case of a 32-year-old man who was treated with multiple pharmaceutical methods, to no avail. A postmortem examination disclosed a half-inch tumor of the pons, which was determined to be a syphilitic gumma. Reports of brainstem lesions, often definitively diagnosed only on autopsy, are scattered throughout the late 19th and early 20th centuries. Attempts at resection of brainstem tumors were not described until 1909, when Dr. Weisenburg reported on a 46-year-old patient who was treated with a decompressive operation for a suspected cerebellar lesion; 6 months postoperatively the patient succumbed to his illness, and a postmortem examination revealed an extensive tumor of the cerebellum, medulla, pons, and cerebral peduncle. In January 1910, Dr. Philip Zenner reported one of the earliest operative interventions for a suspected cerebellar tumor in a pediatric patient, which on autopsy proved to be a large tumor of the pons.

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In the early 20th century, neurosurgeons documented meticulous efforts to combine information from history and examination to predict locations of brainstem tumor pathology to the best of their knowledge. We report an early case of a pediatric brainstem glioma, which was surgically treated with a suboccipital exploration by Dr. Harvey Cushing at the Johns Hopkins Hospital in January 1910. Cushing’s patient survived 2 days postoperatively, which appears to be the longest survival time of those documented in the literature at that time for pediatric glialomas, highlighting one of Cushing’s many successes within the field of neurosurgery.

Methods

Following institutional review board approval, and through the courtesy of the Alan Mason Chesney Archives, we reviewed the Johns Hopkins Hospital surgical files dating from 1896 to 1912. Our review recovered a single case of a pediatric brainstem glioma, which is described here.

Case Report

On September 20, 1909, a 15-year-old schoolgirl (“M.H.”) presented to Roosevelt Hospital in Springfield, Massachusetts, with a 5-month history of slight mental dulling, numbness in the right cheek, and heaviness of her right eyelid; a 1.5-month history of complete deafness in the right ear, intermittent dizziness and vomiting; a 1-week history of stumbling toward the right side; recent onset of positional headache; and altered sensation of the left hand and foot.

On examination by the house staff at the Roosevelt Hospital, she was found to have right-sided facial anesthesia (including right cornea) and paralysis. Neither eye could be turned toward the right beyond the midline. Upward movement of the eyes was also restricted. Nystagmus was present on upward, downward, or leftward gaze. Babinski reflex was detected in the left foot. There was slight uncoordination in both hands in touching nose and ears.

Her symptoms rapidly progressed over the next 10 days. This resulted in her being completely bedridden, as she was unable to stand or walk. In addition, she was reported to have unintelligible dysarthric speech and a conjugate left-sided ocular deviation.

Pontine involvement was suspected, and Dr. Peterson prescribed large doses of potassium iodide, up to 100 g thrice daily. Radical improvement appeared to be made, as noted by Dr. Peterson at the Roosevelt Hospital: “The improvement was magical. I have never seen such improvement except in specific growths.” Her parents reported her to be walking and playing piano until Christmas Day, when suddenly her symptoms grew worse and she was advised to present urgently to the Johns Hopkins Hospital.

Miss M.H. was admitted on January 4, 1910, and was examined by Dr. Cushing. On examination, Cushing reported the following:

Palsy Right abducens—not total. Pupils unequal. Left possibly slightly [as written] > right. Prompt reaction to light and accom. Nystagmus in all directions most marked on looking upward in which position eyes swing to left owning to trace of bilateral abducens palsy. Rhythm rapid to left and especially so to right where fine movements are brought out.

Dr. Cushing conducted an extensive neurological examination the following day.

Olfactory: mother has no sense of smell also grand-father. [olfaction has] Been lost early in other members of family. Patient has had sense of smell but at present does not recognize difference between orange and lemon. […] Taste: 1) Orange not recognized right. immed. recognized on left

2) Syrup called sour—right. recognized immed on left

3) Salt: recognized left—not right (called sour)

4) Acetic acid: recognized but more definite left than right.

Auditory: Has bone conduction in right ear, apparently no air conduction. Lesion would seem to be in middle ear. Forehead test with fork—positive right, negative left (i.e. sound not + left on closing external ear). Air conduction much better left than right to fork and voice.

CN X: Apparently not involved. Change in voice probably not due to laryngeal involvement.

CN XII: Tongue in median line

Cerebrum: Considerable dullness. Tendency to sleep a great deal. Mentally inactive though responds with fair intelligence. No astereognosis—though left is worse than right.

Cerebellum: History of definite tilting of head to right. In erect posture causes usual spinal discomfort—much accentuated by bending head forward. Definite characteristic Kernig.

Tenderness on pressure partic. on right (suboccipital).

Dizziness: subjective—brought out by erect posture Rotation of objects to right has been observed not present now.

Coarse ataxia in both hands—worse in Rt. Present also in mov. of legs. History of tendency to fall to Rt. (unable to walk at present)

Diadoconesis [diadochokinesis]—present

[…] Medulla—evid. involv. from disturb. of speech—slow and thick—also disturb. of deglutition [deglutition]

Dr. Cushing performed a suboccipital exploration for presumed cerebral tumor that took place on January 6, 1910. The operative note documents the procedure:

The usual bilateral approach with a wide exposure. No complications. Bone much thinned particularly on the right side and easily removed well towards each mastoid process. The emissary vein was torn on the left but easily controlled by the use of wax.

Half of the foramen magnum removed and dura exposed down to the arch of the first vertebra. Dura very tense. Primary opening made on the left side towards the upper part of the spinal meninges. Some fluid encountered which sprung from the primary minute opening. It was impossible to expose the lower border of the hernia in the cerebellum which extended almost down to the first vertebra. Finally the operator desisted from the local attack and opened the dura widely on each side. Cerebellum bulged through these openings but fortunately no lesion of either hemisphere was produced.

Although a thorough exploration was made on both
sides (particularly on the right side), Cushing was unable to locate a tumor. Furthermore, “a free transverse incision was made in the right hemisphere and carried well forward toward the anterior part of the cerebellum. Tumor not disclosed.” Closure was performed “as usual” in layers without drainage, and the dura was left wide open. In view of the negative findings in the lateral recess:

It is probable that the tumor actually is a Pontial tumor, cerebellar symptoms due to involvement of the superior cerebellar peduncles. This accounted for the unusual state of sensory disturbances and for the right conjugate paralysis of the ocular movements.

Despite the operation being “well borne” and anesthesia being “well taken,” a few hours after the operation Miss M.H.’s temperature rose suddenly to 105.7°F, with a very feeble, irregular pulse and labored breathing. On January 7, 1910, she became comatose, and on January 8, 1910, she suffered sudden death from “respiratory failure.”

An autopsy was performed and an “infiltrating (?) glioma of the pons” was detected. A portion of the tumor extended out along the sensory root of the trigeminal nerve almost to the ganglion. Cushing’s autopsy note, and the accompanying pathology images (Figs. 1 and 2) describe the lesion:

There is a soft gelatinous mass occupying the cerebello-pontine region on the right extending from the mid-line and including the triangle, measuring 5 cm in antero-posterior diameters. There is a cyst at the outer angle. The cranial nerves from the third to the eighth inclusive are definitely involved in the growth.

[…] On removing the hind-brain and cutting across the peduncle the tumor is encountered on the right, passing up and forward, where it has obviously involved the red nucleus and the substantia nigra.

[…] Sections taken through the mid-brain and cerebellum show that the entire right half of the pons is involved in the growth, which extends downward and backward, even to a lower level than the fourth ventricle […] The fourth ventricle is distorted to an extraordinary degree, and here the tumor juts out over the medulla, as shown particularly well in the third section (of diagram). [See Fig. 1.] Here there is lateral expansion out into the apparently involving cerebellum, though this is not definite. The impression is that the cerebellum has escaped from all but pressure. The pons is much distorted and pushed to the left.

The tumor itself is a most variegated affair. In many places it contains gelatinous cysts, in others bright red patches of haemorrhage, many thrombosed vessels, yellowish areas of tissue degeneration, and a dry surrounding zone of grayish, opaque tissue evidently growing edge, which has the appearance of a section of unripe fruit.

**Discussion**

Early pioneers in surgery and medicine were faced with numerous challenges, including precise tumor localization, safe anesthesia, intraoperative hemorrhage, and postoperative infection. Specifically, in pediatric patients with brain tumors, Cushing had noted a great diagnostic challenge for physicians. Even at present, there can be great difficulty in diagnosing pediatric brain tumors because symptoms may mimic other diseases of childhood. With advancements in medicine, such as the introduction of MR imaging, diagnostic hypotheses can be tested and surgical approaches planned. However, at the turn of the 20th century, diagnosis relied solely on careful history taking and meticulous physical examination. Cushing’s strong beliefs in careful problem solving and knowledge acquisition appear to be reflected as part of a letter he wrote to Dr. Henry Christian on November 20, 1911.

I would like to see the day when somebody would be appointed surgeon somewhere who had no hands, for the operative part is the least part of the work.

We report here Cushing’s early experience with a pediatric brainstem glioma while he was at the Johns Hopkins Hospital.

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**Fig. 1.** Cushing’s diagram of the patient’s brain at pathological examination. Lines represent the section of brain as explained in the pathology report. The drawing was obtained from archived microfilm of the handwritten records, courtesy of the Alan Mason Chesney Archives.
Hopkins Hospital. At that time, intracranial neoplasms were broadly diagnosed as a “glioma,” encompassing a wide range of brain tumor pathologies. It was not until 1926 that Cushing later separated gliomas according to pathological findings, which marked the beginning of pathological groupings.\textsuperscript{5,19} Although a definitive diagnosis is not possible, the description of the lesion as “infiltrating” suggests it may have been diffuse in nature.

Although there is evidence in the case notes that this was believed to be a pontine lesion, the reason for Cushing’s limited suboccipital cerebellar exploration are not explained. It is possible that he recognized that exploration of the pons would prove fatal, and thus, a cerebellar exploration for a lesion compressing the pons offered the chance for decompression and improvement in the quality of life, with the potential for postoperative survival. Even today, resection of diffuse pontine tumors is considered nonbeneficial.\textsuperscript{3,13,22,30} In 1969, Matson\textsuperscript{26} suggested that “regardless of specific histology, brainstem gliomas must be classified as malignant tumors since their location in itself renders them inoperable.” One of the first neurosurgeons to strongly support surgery for certain
brainstem and pediatric brainstem glioma

brainstem tumors were Dr. J. L. Pool in the 1960s. He was noted to have performed operations in several children, whose survival ranged from 10 to 25 years. In the early 1980s, more reports of highly favorable surgical outcomes for certain types of brainstem gliomas became available. Since then, multiple classification systems have been developed to help determine which tumors can be resected safely.

Cushing's work appears to predate the earliest published reports of surgical interventions for pediatric brainstem glioma in which the patient survived postoperatively. Furthermore, with pediatric cases such as the brainstem glioma in which the patient survived postoperatively, outcomes for certain types of brainstem gliomas became available. Since then, multiple classification systems have been developed to help determine which tumors can be resected safely.

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