Published in final edited form as:

Childs Nerv Syst. 2011 July ; 27(7): 1145–1148. doi:10.1007/s00381-011-1444-7.

Unique challenges faced by pediatric neurosurgeon Harvey Cushing in 1909 at Johns Hopkins: a choroid plexus tumor of the lateral ventricle mimicking a cerebellar lesion

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Abstract

Introduction—At the turn of the twentieth century, diagnosis of pediatric intracranial lesions frequently relied exclusively on the subtle, nonspecific signs associated with these pathologies. In absence of detailed neuroimaging, erroneous diagnoses and misdirected operative explorations were common within pediatric populations.

Methods—Following institutional review board approval, and through the courtesy of the Alan Mason Chesney Archives, we reviewed the surgical files from the Johns Hopkins Hospital for the years 1896–1912. Harvey Cushing's neurosurgical cases were collected and further analyzed.

Results—A 9-year-old boy presented with a year-long history of headaches and blindness. His neurological examination was remarkable for focal cranial nerve deficits and cerebellar signs. Cushing diagnosed a cerebellar tumor with hydrocephalus and performed two suboccipital explorations with negative results. The patient died prior to further operative intervention, and the postmortem examination revealed a large choroid plexus tumor within the left lateral ventricle.

Conclusions—Early neurosurgical diagnosis and operative planning was fraught with difficulties, and surgeons relied solely on history and physical examination for localization of intracranial lesions. Here we report a case of a choroid plexus tumor of the lateral ventricle, which was misdiagnosed by Cushing. This case serves to emphasize the unique challenges faced by

pediatric neurosurgeons at the turn of the twentieth century, who relied solely on history and examination to localize intracranial lesions.

Keywords

Harvey Cushing; Pediatric neurosurgery; Choroid plexus tumor

Introduction

Choroid plexus tumors are rare primary intracranial tumors, comprising less than 1% of all brain tumors [8]. The majority of these tumors occur in children and infants, with 74% diagnosed within the first decade of life [8, 9]. Signs and symptoms of increased intracranial pressure, including headache, irritability, nausea, vomiting, and changes in vision, are commonly seen at presentation [7, 8]. However, within a pediatric population, these symptoms may be subtle. Therefore, the modern diagnosis of choroid plexus tumors within the pediatric population relies on neuroimaging to supplement the history and physical examination findings. At the turn of the twentieth century, neurosurgeons operated without the benefit of neuroimaging, relying on detailed history taking, and thorough neurological examination for lesion localization and operative planning.

Harvey Cushing had extensive experience treating intracranial lesions in pediatric populations [10, 11], and by the end of his early career at the Johns Hopkins Hospital had refined his neurological examination to detect even subtle signs of increased intracranial pressure. However, in the absence of neuroimaging techniques, diagnosis of intracranial lesions remained fraught with difficulty. Here we report a case of a 9-year-old boy with a choroid plexus papilloma of the left lateral ventricle, who was misdiagnosed with a cerebellar tumor, and underwent two posterior fossa surgical interventions by Cushing, with negative findings.

Methods

Following institutional review board approval, and through the courtesy of the Alan Mason Chesney Archives, we reviewed the surgical files from the Johns Hopkins Hospital for the years 1896–1912. Cushing's neurosurgical cases were collected and further analyzed.

Case report

On January 22, 1909, a 9-year-old boy presented to the Johns Hopkins Hospital complaining of “loss of sight.” His past medical history was remarkable for two falls resulting in brief loss of consciousness, occurring at the ages of 4 and 5 years. His family history was unremarkable.

Ten months prior to admission, the patient developed severe headaches; the parents reported at the time of admission that “these were general, not confined to any portion of head at first, but gradually became almost entirely frontal in character. No occipital headaches. These headaches would last 2–3 days and then disappear for several days.” A lumbar puncture was performed at an outside practice and did not improve his symptoms. The headaches were

worse at night and accompanied by nausea and vomiting. During the first month of these symptoms, the parents reported the patient experienced two “convulsive attacks” that were “not spastic or rigid, both sides seemed equally affected. Pt lost consciousness and ability to talk to some extent.”

Cushing's physical exam documented:

Head—Seems distinctly enlarged. Marked dilation of superficial vessels especially evident in frontal region where I can feel groove made by them [...] Coarse nystagmoid movements especially when looking to left. Easily fatigued in upright posture. Tends to hold head in left hand.

A thorough cranial nerve exam was performed by Cushing in an attempt to localize the lesion:

Cranial Nerves

I. Evidently marked diminution in sense of smell. Unable to smell flower (carnation), lemon, etc. Aromatic salts of ammonia irritated but gives no olfactory impression.

II. Opticus

Complete blindness. No appreciation of bright light. Pupillary reflexes gone. Double choked disks (about 5 D). Evidently trophic glistening nerve.

No hemorrhages.

III—IV—VI

Ocular movements normal. No pupillary reaction. Nystagmoid movements more often accompanying uncertain chance movements than definite efforts to look to right or left. Palpebral clefts equal.

V. Trigeminus

No motor or sensory disturbance evident. No deflection of jaw. No disturbance of taste. Salt and lemon promptly recognized.

VII—IX—X—XI—XII

Other cranial nerves likewise negative except acusticus [sic].

VIII. Possibly hears less well on left side (subjective as well as objective).

An exam of cerebral and cerebellar function was conducted also:

Cerebrum.

Apparently normal mentality perhaps rather too jocular. Memory seems unaffected.

[...] No evidence of pyramidal tract disturbance. No aphasia, no stereognosis.

Cerebellum—

Some unsteadiness in erect posture, more than can be attributed to blindness. Some tremor and ataxia especially in left hand. Ataxia brought out in all extremities.

Tends to swerve to left in walking. Does not stand well on either foot alone. More unsteady on right (?) Definite nystagmus with coarse movements to left.

No definite weakness or inability to execute rapidly repeated movements on left more than right.

Cushing reached the diagnosis of “left cerebellar lesion (tumor)—(cyst) with low grade of internal hydrocephalus (note loss of sense of smell).”

Cushing brought the patient to the operating room on January 25, 1909 for “suboccipital decompression and exploration for presumed left cerebellar tumor.” His operative note documents the procedure:

The shaving of the head brought into view an evident cranial asymmetry, shown by the greater prominence of the left occipito-mastoid region over the right.

The child was placed on the table and anaesthetized in position. Instead of the usual cross-bow incision the operator attempted to carry out the exploration by the simple transverse incision. it was, however, a mistake to do so, for though the bone was successfully rongeured away so as to include the posterior half of the foramen magnum, the depth of the wound was such, even with retraction of the flap, that manipulations were carried out with difficulty.

[...] The cerebellum was so tense that lateral exploration was difficult. There was no evidence on the surface of any growth nor any evident abnormalities [...] the mainly unsatisfactory part of the exploration was the impossibility of getting any sort of view into the left lateral recess where it was presumed the tumor lay (cf acousticus symptoms).

After this unfruitful exploration, the patient's hospital course was not documented, except for a brief note written by Cushing on February 1, stating that “the child has been dull and stupid since operation [...]. Child yesterday was so dull as not to recognize parents.” He returned to the operating room on February 1, 1909 for an aspiration of the right ventricle, under primary chloroform anesthesia.

Cushing brought the patient to the operating room on February 9 for “Suboccipital exploration; second stage operation. Negative findings.” His operative note documents the procedure, as well as his frustration and confusion in not locating the lesion.

The left cerebellar hemisphere in particular was exposed thoroughly and a search made in all directions for growth. It was possible to explore over the superior surface all the way to the median line and to the anterior margin of tentorium. The operator had never succeeded in so thoroughly exposing and exploring the entire field about the cerebellum. On the right the exposure was a little bit less satisfactory, but nevertheless was sufficient to throw out the possibility of growth being in the immediate neighborhood of the cerebellum or on its surface [...] the right cerebellar lobe was consequently incised and an exploratory wound carried to a depth possibly of 2–3 cm. without disclosing any lesion.

On postoperative day 3 the patient was listed as being “in critical condition.” There is no further documentation of the patient's postoperative course. The patient died on February 15, 1909 at 2:50 p.m. A postmortem examination was conducted, and the intracranial findings were documented (Fig. 1):

A series of coronal sections of the brain brought into view an extraordinary cystic tumor, which extended all the way from within 5 mm of the posterior part of the left occipital pole to a level corresponding practically with the tip of the right temporal lobe, making the tumor, including the enveloping cysts, fully 10 cm in its anteroposterior diameter [...]. On the right there is an unusual dilation, brought into view, of the right lateral ventricle; on the left there is a tumor mass the size of a hen's egg, of a curious reddish-gray color. The posterior end of the egg-shaped tumor has been shaved off by the section [...] the tumor is surrounded by a number of cystic cavities of varying size, containing the same yellowish, clouded material and lined by a delicate membrane. The cut surface of the tumor shows it to be somewhat lobulated, of a lightish yellowish-red color with an occasional grayish patch which may possibly indicate areas of necrosis [...] at the upper margin of the tumor itself is seen a structure which apparently connects with the choroid plexus of the left ventricle [...] it is therefore quite possible that the growth may have originated in the tip of the left choroid plexus.

Following the postmortem examination, Cushing modified the patient diagnosis to read “choroid plexus tumor.” Cushing included this patient in his 1925 paper on papillomas of the choroid plexus [6], providing detailed information regarding the pathologic findings:

The tumor showed histologically the characteristic appearance of having originated from the choroid plexus. The villi were arranged in a somewhat compact fashion, and only in places showed typical papillomatous characteristics.

Discussion

Although Cushing described a slightly higher prevalence of 1.5% for choroid plexus tumors in his 1923 publication on intracranial lesions [4], at the turn of the twentieth century, choroids plexus tumors were exceedingly rare. In 1925, Cushing and Davis reported only 13 examples of choroid plexus papillomas in the existing literature [6], and a 1933 review of the literature reported on 54 documented cases [12].

Until the introduction by Bailey of a histological stain separating ependymal tumors from gliomas [1], and later the publication by Cushing and Bailey in of a standardized classification for gliomas [2], parenchymal tumors invading the ventricles were often confused for tumors of the choroid plexus [6]. The advent of histological staining for tumor-specific structures and proteins allowed more specific categorization of intracranial tumors and improved recognition of choroid plexus tumors [1, 3, 5, 6].

In the 1925 article [6], Cushing reported briefly on the case described here, offering a brief summary of the neurological examination and no details of the operative interventions.

Despite the brevity of his own report [6], Cushing conducted an extensive neurological exam, using findings on the cerebellar and cranial nerve examinations to attempt to localize the suspected intracranial lesion. With prominent cerebellar signs and focal deficits in the cranial nerves, Cushing diagnosed the child with a lesion of the left cerebellar hemisphere with accompanying hydrocephalus. Two operative explorations returned negative findings, and the patient died prior to any further interventions. A postmortem examination revealed a large choroid plexus tumor with accompanying cystic structures within the left lateral ventricle and the occipital lobe. This case serves to emphasize the unique challenges faced by pediatric neurosurgeons at the turn of the twentieth century, who relied solely on history and examination to localize intracranial lesions. Cushing understood both these challenges and the obligation of the neurosurgeon, stating:

It should be the ambition of the neurosurgeon to be able not only to localize a tumor with sufficient accuracy to expose it in the way most favorable for operative removal, but also, if possible, to foretell its pathologic nature, for on this the line of attack will largely depend [6].

Although Cushing appeared surprised by the postmortem findings, his other surgical cases nevertheless demonstrated an extremely high mortality rate among patients he operated on for intraventricular tumors; it is possible that the correct diagnosis would have led Cushing to perform a less complex and more anatomically direct decompressive operation, or even to have performed an exploration of the lateral ventricle, with an unlikely impact on the patient's ultimate outcome.

Acknowledgments

CP was supported by a Howard Hughes Medical Institute Student Research Training Grant. AQH was supported by a K08 NIH grant and an HHMI grant.

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Fig. 1.

Left Cushing's schematic demonstrating the sectioning of the brain during the postmortem examination. *Right* photographs of four sections of the brain, documenting a tumor of the left lateral ventricle choroid plexus