MENINGIOMA OF RECORD SIZE WITH UNUSUAL FEATURES

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Intracranial tumours of record size occasionally merit a brief report, especially
if they give rise to unusual complications. The following case history is of interest
because the tumour was first visualized in the lateral ventricle in a child of 5½
years and had reached the enormous size of 1353 grams when finally removed 10
years later. Inconclusive biopsy at the first operation led to failure in establishing
the correct diagnosis, and prolonged refusal on the part of the parents prevented
removal long after its slow growth and deposition of calcium salts had made the
benign character of the tumour evident. Although complete removal of this me-
nangioma was carried out successfully in two stages, epileptic seizures with mental
deterioration followed and the patient finally died 6 months afterwards of hydro-
cephalus secondary to an unusual hygroma which developed in the huge cavity left
after removal of the tumour.

A review of previous accounts of meningiomata of record size shows that the
largest described by Cushing and Eisenhardt1 weighed 810 grams. In their book
they also cited an unpublished 341-gram meningioma removed by Hoen. Pilcher,5
who summarized the literature in 1943, reported one of 374 grams, which he had
removed, and mentioned a still larger one recorded by Davidoff.6 In this case the
weight of 835 grams included not only the tumour proper but an undetermined
amount of cranial exostosis.*

CASE HISTORY

A 5½-year-old boy first entered the Massachusetts General Hospital in 1936, with a his-
tory of intermittent vomiting and headaches which had begun several months after a fall
downstairs at the age of 18 months. The baby had been unconscious for a few minutes, but
had recovered without other immediate sequae.

Examination revealed right internal strabismus and nystagmus. Awkwardness and
dysynergia of the right arm with unsteadiness of gait suggested a cerebellar neoplasn. Motor
power, sensation, and reflexes were normal. Ophthalmological findings were papilloedema of
5 D., reduction of visual acuity to 20/200, and contracted visual fields with a suggestive right
homonymous hemianopsia. Skull films at this time showed evidence of prolonged increase in
intracranial pressure.

At the time of ventriculography combined ventricular-lumbar pressures were recorded:

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<tr>
<th></th>
<th>R. ventricle</th>
<th>L. ventricle</th>
<th>Lumbar space</th>
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<tbody>
<tr>
<td>Initial pressure:</td>
<td>400</td>
<td>—</td>
<td>455</td>
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<tr>
<td>Colour:</td>
<td>Clear &amp; colourless</td>
<td>Deeply xanthochromic</td>
<td>Slightly xanthochromic</td>
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<tr>
<td>Protein:</td>
<td>24</td>
<td>280</td>
<td>708</td>
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<td>Goldsol:</td>
<td>1122210000</td>
<td>3445555555</td>
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* Since this manuscript has been accepted for publication Dr. Eisenhardt has called our attention
to a case report by C. W. Laymon in Minnesota Medicine for November, 1949 (32: 1182-1183). This
records a giant meningioma which had invaded the right frontoparietal portion of the skull and expanded
beneath the scalp to a "size of two grapefruits" and at post mortem weighed 2300 grams. The
Ventriculography showed the right lateral ventricle to be enlarged. Both it and the 3rd ventricle were displaced to the right. Only the posterior horn of the left lateral ventricle filled with air. This appeared to be enlarged with "a sharp concave anterior margin consistent with amputation of the ventricle in this area."

1st Operation. On Nov. 28, 1936 a left occipitoparietal craniotomy and exploration, carried out by Dr. John S. Hodgson, disclosed an encapsulated tumour which blocked the midportion of the lateral ventricle. Owing to the high degree of intracranial pressure and bleeding, it was felt that a radical attack on the tumour was out of the question, so only a biopsy was taken. It was not possible to close the bone flap, which had to be sacrificed to provide for decompression.

Microscopic Report. The specimen proved inadequate for a positive diagnosis. "The findings are probably consistent with polar spongioblastoma, although fibrils are more numer-

![Fig. 1. (A) Appearance of patient and extracranial herniation of tumour just before its removal. (B) Left lateral plate of skull to show size of tumour from area of calcification. Dotted line marks intracranial extent of calcium deposits as seen in x-ray film.](image)

ous and heavier than is usual. The wall of a cyst in an astrocytoma is another possibility."

Course. Postoperative convalescence was uneventful, but a cerebral hernia and a right homonymous hemianopsia rapidly developed. During the ensuing 10 years the hernia slowly increased. In 1937, except for the homonymous field defect, a mild degree of optic atrophy, and a 5-inch herniation, he was a normal appearing boy. Headaches and papilloedema had subsided. In 1942, however, there developed a right hemiparesis and convulsive seizures. These began with an epigastric aura and clonic movements of the right arm, which rapidly became generalized. Although right-handed and temporarily unable to talk after his seizures, there was never persistent aphasia. At our suggestion he was taken by his parents to Montreal, where he was examined by Dr. Wilder Penfield. Despite his advice and repeated efforts on our part, operation was refused by the parents. In 1946 the boy had to be taken out of school because of his seizures and was finally readmitted for operation at his own insistence in September, 1946.

Examination on 2nd Admission. The occipitoparietal herniation protruded a good 6 inches (Fig. 1A). There was a moderate degree of right-sided hemiparesis with increase in reflexes

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The specimen included the invaded portion of the skull and no mention is made of its intracranial volume. The patient was a 72-year-old recluse with a 7-year history of frontal swelling, 5 years of deterioration of mental faculties, and 2 years of weakness in the left arm with epileptic seizures.
and a reduction in sense of touch and astereognosis. Eye findings were right homonymous hemianopsia with macula splitting, optic atrophy, and no papilloedema. Visual acuity was reduced to fingers at 2 feet. Lumbar puncture gave an initial pressure of 225 mm.; protein was 124 mg. per cent. X-rays now revealed strands of calcification within the herniated tumour mass, which extended forward and medially nearly to the centre of the skull (Fig. 1B).

2nd Operation. Extirpation of the tumour was undertaken in two stages. On Jan. 14, 1947 the herniating portion of the fibrous calcified meningioma was excised. Except for the vascular capsule, which contained many veins the size of a lead pencil, no unexpected difficulties were encountered. Loss of blood, although profuse, was well compensated for by multiple transfusions. This portion of the tumour weighed 520 gm.

Course. The boy was discharged 3 weeks later and readmitted after 3 months in much the same condition.

3rd Operation. The remainder of the tumour, weighing over 833 gm., was resected on April 8, 1947. By turning down an additional cranial flap anterior to the pre-existent bony defect a good line of cleavage between normal brain and tumour could be developed. Beyond some difficult moments in securing the large vessels at the base of the tumour, the entire mass was resected without difficulty. This left a huge defect in the posterior half of the skull with the falx exposed at the midline and the tentorium at the base. The cavity was filled with Hartman’s solution.

Course. The patient recovered without increase in his hemiparesis or aphasia, but he ran a febrile course (101–103°) for 18 days with a very bloody CSF. At discharge on April 25, 1947, he was continuing to have frequent minor seizures, during which he did not lose consciousness but was unable to speak.

4th Admission. His final hospital admission was on Sept. 10, 1947 in status epilepticus, from which he soon recovered on heavy anticonvulsant medication. At this time he had bulged with a large accumulation of fluid. On tapping the cavity the initial pressure was 140 mm. The fluid was xanthochromic and contained 10 lymphocytes with many crenated rbc.; protein 270; sugar 89 mg. per cent. Neutral phenolsulphonephthalein injected into the cavity was recovered in small amount in the lumbar subarachnoid space. During the following days 400 to 1,700 cc. of fluid were aspirated frequently from the cavity, only to reaccumulate within a few days. On Oct. 4, 1947 a lumbar puncture was done after 2 days of fever spiking to 104°. The protein content had now risen to 600 mg. and the cell count to 750 with 60 per cent polymorphonuclears, 25 per cent lymphocytes, and 15 per cent crenated rbc. The sugar had fallen to 33 mg. per cent. Repeated cultures were sterile. On treatment with penicillin and sulphadiazine the fluid cleared, but the patient continued to run a low-grade febrile course and gradually sank into coma. Death occurred on Oct. 26, 1947.

Pathological Examination. The surgical specimens removed at the last two operations formed a hard, lobulated, roughly ovoid tumour measuring 14×12.5×11 cm. and weighing 1353.5 gm. The outer surface was covered by a glistening, thin, fibrous capsule with numerous large veins. The cut surface was white, avascular, and exceedingly gritty.

Microscopically the tumour was composed of rather straight interlacing bands of slender fusiform cells and dense strands of collagen. There were only a few, widely scattered whorls that were characteristic of meningioma. There were extensive regions of degeneration containing heavy deposits of calcium in the form of fine particles and larger concretions not related to whorls nor resembling the usual psammoma bodies.

Although the weight of the tumour equalled that of a normal brain, its smaller size was due to greater density and the high content of calcium (Fig. 2).

Postmortem Examination. There were discrete ulcers of the oesophagus, cardia of stomach and duodenum, and a perforation of the oesophagus, with extravasation of gastric contents into the peritoneal and both pleural cavities. This was apparently a terminal event, because there was no evidence of mediastinitis or peritonitis.

The cerebral convolutions were moderately flattened. The operative defect in the posterior part of the left hemisphere involved a large portion of occipital and parietal lobes and com-
morphonuclear ofstruction pressed system.

subdural the tile drocephalus. have dipped into and the separated extraeranial great, municated the 458 recent left perivascular placed and the membranes, 0.6 mm. thick, but no adherent to it. This inner membrane dipped into the operative defect, continuing as an unbroken lining of the entire ventricular system. An outpocketing, about 2 cm. in diameter, extended through the tentorial notch, dorsal to the quadrigeminal plate. This and the underlying aqueduct were somewhat compressed and displaced to the right (Fig. 3, insert). Aside from this there was no evident obstruction to the outflow of CSF, as the foramina of Magendie and Luschka were patent.

Microscopic examination of the dura was not remarkable. The membranes were composed of dense, vascular, fibroblastic tissue with a slight infiltration of lymphocytes and polymorphonuclear leucocytes. There were a few deposits of blood pigment in the space between

![Image](https://example.com/image.png)

Fig. 2. Size of 1353-gram meningioma compared with a normal adult brain. In the specimen at the left the upper portion represents the 883-gram intracranial extension, the lower portion the 592-gram extracranial extension of the tumour removed at the first stage.

the membranes, but no exudate. On the surface of the brain at the site of operation there were recent haemorrhages, collections of phagocytes filled with blood pigment, and areas of exudate and lymphocytic infiltration. The ependyma of the ventricles was partly destroyed and replaced by a dense layer of fibrous tissue with underlying glial proliferation. This layer was 0.6 mm. in thickness, its surface coated with an exudate of neutrophilic leucocytes and beneath, in the subependymal zone, underlain by proliferating adventitial connective tissue and perivascular infiltrations of lymphocytes.

The protein-rich fluid surrounded by a membrane resembled a subdural hygroma. We have observed a very similar hygroma at autopsy after an operation for obstructive hydrocephalus. This communicated with the 3rd ventricle through a ventriculostomy opening in the lamina terminalis. We believe that the presence of large quantities of bloody CSF in the subdural space after operation led to the formation of membranes with encapsulation, as is the case in the development of subdural haematomas.

**DISCUSSION**

This case is reported not only because the tumour was so much larger than any meningioma of previous record, but because of several other unusual features. These include:
Time of Onset. Meningiomas rarely occur in childhood. In this case signs of increased intracranial pressure first developed in a child of 5½ years. In the series of 313 meningiomas reported by Cushing and Eisenhardt only 6 were found in preadolescents, and the youngest arising within the skull occurred in a child of 8.

Production of Cerebellar Signs by Compression of the Tentorium from Above. Nystagmus, clumsiness suggesting adiadochokinesis, and unsteadiness of gait at the time of the patient’s first admission led to a tentative diagnosis of tumour beneath the tentorium. In Pilcher’s case, where a 374-gram meningioma occupied the occipitoparietal region, the patient also had nystagmus and unsteadiness of gait, which together with deafness on the side of the growth raised the question of an acoustic tumour. Negative posterior fossa exploration in each case was averted by ventriculography.

Late Complications Following Removal of Tumour. The extensive subdural hygroma which filled the postoperative defect appears to have progressively interfered with the circulation of CSF by compression of the quadrigeminal plate and Sylvian aqueduct. As these membranes communicated with the ventricular system, fluid containing erythrocytes, white cells, and a rising concentration of protein accumulated within the cavity and ventricles in increasing amounts. As suggested by Finlayson and Penfield and recently confirmed by Jackson in animal experiments, the discharge of decomposition products of red blood cells in the subarachnoid space and ventricles may produce an aseptic meningitis and ventriculitis. These findings were features of unusual interest in the terminal illness 6 months after successful surgical removal of the tumour.
The tragic features in this case were the misleading biopsy at the primary operation and the prolonged refusal on the part of the parents to permit reexploration. If this could have been carried out within the first 6 years, before the onset of hemiparesis and convulsions, the patient might have made a satisfactory recovery with only a moderate reduction in visual acuity and a right homonymous hemianopsia.

SUMMARY

A meningioma of over 10 years' growth is reported with a record weight of 1853 grams.

Removal of the 520-gram extradural herniation and secondary resection of the 833-gram intracranial mass from the parieto-occipital portion of the brain were accomplished successfully, but too late to prevent deterioration from cerebral atrophy and convulsive seizures. The boy died 6 months later of progressive hydrocephalus. An unusual subdural hygroma, which filled the postoperative cavity and compressed the Sylvian aqueduct, communicated with the ventricles.

REFERENCES


DUANE'S RETRACTION SYNDROME

A CASE REPORT*

John W. Chambers, M.D.

Veterans Administration Hospital, Perry Point, Maryland

(Received for publication February 16, 1950)

This case exemplifies a particular congenital anomaly involving extraocular movements and is reported because of the great confusion in diagnosis which could have been avoided by careful observation, despite the misleading history. Errors in diagnosis of ocular anomalies are frequent as a result of failure to realize that congenital anomalies of the eyes are not uncommon and are often unnoticed by the individual patient—i.e. congenital Horner's syndrome, hyaline bodies of the nerve (Drusen) suggesting choked disks, tilting of the optic disks with bitemporal field defects suggesting a chiasmal lesion, pseudopapilloedema, pseudoneuritis and so forth.

REPORT OF CASE

W.E.S. (P.P.V.H. #20784), a white male aged 19 years, first complained in the fall of 1947, while in the army, of double vision on looking to the left and of inability to move either

* Sponsored by the Veterans Administration and published with the approval of the Chief Medical Director. The statements and conclusions published by the author are the result of his own study and do not necessarily reflect the opinion of the Veterans Administration.