HEMANGIOBLASTOMA WITH POLYCYTHEMIA


HEMANGIOBLASTOMA OF CEREBELLUM WITH POLYCYTHEMIA

CASE REPORT

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Neurological signs and symptoms are frequently associated with primary polycythemia due to the vascular complications characteristic of this disease. Conversely, instances of intracranial neoplasms with an associated secondary polycythemia have been reported in the literature.

The occurrence of expanding subtentorial lesions with polycythemia is extremely rare. Oppenheimer reported a case of polycythemia and cerebellar medulloblastoma. Two cases of hemangioblastoma of the cerebellum and polycythemia were reported by Carpenter, Schwartz, and Walker. Both of their patients had remission of the polycythemia following removal of the hemangioblastoma. An excellent review of the literature on neurogenic polycythemia was given by Drew and Grant in 1945. They added 1 case, the intracranial lesion being a subdural hematoma. The polycythemia in their patient, however, showed signs of returning after 5 months.

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CASE REPORT

C.M., aged 24, a second lieutenant in the infantry, was admitted to the U. S. Army 70th General Hospital on Aug. 9, 1944. He had been evacuated from the 29th Station Hospital and was flown from Algiers to Oran. On admission, he was conscious but spoke very slowly and in a monotonous tone. P. was 60/min., R. 11/min; B.P. normal. He seemed well oriented but was not alert. When taken by litter to the x-ray department, he began to scream with pain and afterwards complained of persistent severe headache. Later in the evening he became very drowsy and stuporous, but improved slowly after intravenous administration of 50 per cent glucose. The next morning he had projectile vomiting. The officer who had accompanied him from Algiers stated that the patient had deteriorated mentally over a period of about 4 months and particularly during the 7 days before admission.

The patient's illness began in October, 1943, with severe throbbing occipital headaches which persisted. For the last few months he had noticed difficulty in reading, blurring of vision, and diplopia. He had been troubled by dizziness and stumbling, particularly to the right, and there seemed to be some slowing of his speech. He had been in another general hospital in January, 1944, and in a station hospital in February 1944 with the chief complaint of headache. He was discharged both times with a diagnosis of psychoneurosis. He was readmitted to the 29th Station Hospital on Aug. 2, 1944. A lumbar puncture was done there and following this he became much worse, being stuporous a good deal of the time.

Examination. He was a moderately drowsy but easily aroused individual talking in a monotonous tone. There was no aphasia, ataxia, or asynergism. He had nystagmus right and left, and marked bilateral papilledema with exudate. There was a right facial weakness with hypesthesia. The tendon reflexes were slightly hyperactive on the right. Babinski reaction was present bilaterally. There was incoordination of the arm and hand with past pointing with both hands. Heel-to-knee tests were performed fairly well.

RBC was 4,790,000; Hb. 13.5 gm. (90 per cent). Leucocyte and Schilling counts were normal. Clotting time was 7 min.; bleeding time, 3 min. X-rays of skull were negative.

1st Operation. On Aug. 9, 1944, preliminary ventriculography through the right posterior horn (80 ml. of fluid removed and replaced with air) showed a dilated ventricular system with a large 3rd ventricle. Cerebellar craniotomy was then done. After the dura had been opened over both cerebellar hemispheres and the occipital sinus ligated and cut, a cystic tumor was seen in the left hemisphere, extending down into the cerebellar tonsil. The cyst was evacuated. Then, using lighted retractors, it was completely explored, but no nubbin of tumor or evidence of hemangioma was found.

Postoperative Course. The patient did very well. Two lumbar punctures were done because of slightly bloody CSF, but otherwise his course was uneventful. He was evacuated to the zone of interior, and was given a medical discharge from the army.

Subsequent Course. He was asymptomatic in every respect until 1948, when he wrote that his symptoms were beginning to reappear. Accordingly, we advised him to enter the U.S. Veterans Hospital at Jefferson Barracks, Missouri. He was admitted on June 11, 1948. He had been entirely well until about 2 1/2 months previously, when he first suffered persistent, dull headaches, partially controlled by headache powders. Following this he noticed a return of visual disturbances, particularly difficulty in focusing. There was a tendency to stagger on walking and true vertigo on leaning forward. The symptoms were especially prominent during the last 2 1/2 weeks.

Examination. Positive neurological findings were: nystagmus on looking to the right, markedly positive Romberg test, and staggering gait. Fundoscopic examination by the ophthalmological consultant revealed no abnormalities.

RBC was 7,890,000; Hb. 21.1 gm. (136 per cent); hematocrit cell volume per cent 69. Leucocyte count was 6,850, with normal Schilling count; platelet count was 645,000. Repeated RBC was 7,820,000; Hb. 21.9 gm. (136 per cent). Kahn reaction was negative.

Roentgenograms of the skull showed only the old cerebellar craniectomy wound. Films
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of the chest revealed an area of increased opacity in the left apex; the radiologists were unable to determine the state of its activity.

2nd Operation. On June 16, 1948, the cerebellar craniotomy wound was reopened. A large bluish, pulsating mass occupied the whole left half of the posterior fossa. This mass was made up of extremely large, thin-walled blood vessels. It was gradually completely removed, the vessels being coagulated and broken, and the bleeding points secured with electrocautery.

Microscopic Examination. "The section discloses an angiomatous tumor consisting of blood channels ranging in size from immense cavernous structures to minute capillaries or sinus-like vessels. There are relatively solid areas consisting of clumps of cells forming a sort of syncytium. Cytoplasm is clear; the nuclei are round or oval and pale. In some areas slit-like vessels are apparent."

Diagnosis: Hemangioblastoma.

Postoperative Course. Immediately after operation, his RBC fell to 5,960,000, with 17.8 gm. Hb. (115 per cent). On June 29, 1948, 13 days postoperative, RBC was 4,800,000; Hb. 16 gm. (103 per cent). His course was uneventful and he was discharged on July 14, 1948.

Subsequent Note. At the present time the patient is in the U.S. Veterans Hospital, Bay Pines, Florida, because of activation of the tuberculous lesion previously noted. Reports from this hospital of the patient's erythrocyte count and hemoglobin are included in Table 1.

<table>
<thead>
<tr>
<th>Date</th>
<th>RBC (c.mm.)</th>
<th>Hb.</th>
<th>WBC (c.mm.)</th>
<th>Differential, Hematocrit, Etc.</th>
</tr>
</thead>
<tbody>
<tr>
<td>8- 8-44</td>
<td>4,790,000</td>
<td>90%</td>
<td>9,000</td>
<td>Polys. 70%, Lymph. 22%, Schilling 100, Baso. 1, Eosin. 2. Clotting, 7 min. Bleeding, 3 min.</td>
</tr>
<tr>
<td>8- 9-44</td>
<td></td>
<td></td>
<td></td>
<td>Operation: removal of cerebellar cyst</td>
</tr>
<tr>
<td>6-11-48</td>
<td>7,890,000</td>
<td>130%</td>
<td>6,850</td>
<td>Neutro. 59, Lymph. 40%, Eosin. 1, Hematocrit 69.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Platelets 640,000</td>
</tr>
<tr>
<td>6-11-48</td>
<td>7,820,000</td>
<td>136%</td>
<td>21.9 gm.</td>
<td></td>
</tr>
<tr>
<td>6-16-48</td>
<td></td>
<td></td>
<td></td>
<td>Operation: removal of hemangioblastoma</td>
</tr>
<tr>
<td>6-10-48</td>
<td>5,900,000</td>
<td>113%</td>
<td>17.8 gm.</td>
<td></td>
</tr>
<tr>
<td>6-17-48</td>
<td>5,680,000</td>
<td>118%</td>
<td>18.3 gm.</td>
<td></td>
</tr>
<tr>
<td>6-29-48</td>
<td>4,800,000</td>
<td>108%</td>
<td>16.0 gm.</td>
<td></td>
</tr>
<tr>
<td>2-10-50</td>
<td>4,270,000</td>
<td>86%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3- 3-50</td>
<td>4,820,000</td>
<td>90%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3-27-50</td>
<td>4,500,000</td>
<td>92%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4- 3-50</td>
<td>4,750,000</td>
<td>94%</td>
<td></td>
<td></td>
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</tbody>
</table>

SUMMARY

The third reported case of hemangioblastoma of the cerebellum with associated polycythemia is presented. The patient had a complete remission of the polycythemia after the removal of a large hemangioblastoma. It will be noted that the polycythemia did not develop until after the large hemangioma was demonstrable. There was no evidence of such lesion at the first operation. His red blood cell count and hemoglobin values have remained normal to the present date.
REFERENCES


A SIMPLIFIED APPARATUS FOR CONSTANT VENTRICULAR DRAINAGE

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Several techniques for effecting constant ventricular drainage have been described.¹ ³⁴⁵ Any successful routine method should be easily available at all times, provide a means of measuring and controlling the intracranial pressure and the amount of ventricular fluid drained, afford maximum protection against infection and be well tolerated by the patient. An apparatus that meets these requirements has been developed by the neurosurgical services of The Peter Bent Brigham Hospital and The Children's Hospital, Boston.

The apparatus consists of the following parts: (A) an ordinary graduated 1 liter Fenwal intravenous bottle which is held inverted by a ring attached to the head of the patient’s bed; (B) a “J”-shaped double lumen glass tube which fits the standard Fenwal single hole rubber stopper and extends into the bottle about 22 cm.; (C) connecting rubber tube attached to the glass tube at one end and a glass adapter and standard intravenous needle at the other; (D) a 12-inch length of small-caliber polyethylene tubing,² which is used as the ventricular catheter. All of this except the polyethylene tubing is sterilized as a unit; several units are kept available at all times.

To institute drainage, a ventricular tap is made in the usual fashion using an open-end needle fitted with a stylet. When a free flow of fluid has been obtained the polyethylene catheter is threaded into the ventricle through the needle and the needle withdrawn, leaving the catheter in place. An intravenous needle of proper size is inserted into the end of the catheter and attached to a 1 cc. syringe. When adequate flow of ventricular fluid through the catheter is assured it is anchored to the skin with stitch and a sterile dressing is applied to the head around the catheter. The needle is then removed from the syringe and attached to the tubing from the drainage bottle. The bottle is usually positioned so that the curved top of the glass tube is about 150 mm. above the former magnum, but any desired pressure can be maintained by raising or lowering the bottle. If the rubber connecting tube is passed over a pulley at the head of the bed and a small weight attached to the bottom of the hanging loop, the patient has considerable freedom of motion without tension on the ventricular catheter. To protect against kinking and tearing, the polyethylene catheter should be reinforced with adhesive tape where it receives the intravenous needle. The system is shown diagrammatically in Fig. 1.

COMMENT

During a 9-month period from June 1949, to April 1950, this type of drainage was used on 26 patients 29 times. The diagnoses in these cases included brain tumor, brain abscess, hydrocephalus of various types, myelomeningocele with Arnold-Chiari malformation, and tuberculous meningitis. The duration of drainage, dictated by the clinical requirements of the patient, was usually 8 days or less (21