CASE REPORTS

CONGENITAL ATRESIA OF THE FORAMINA OF LUSCHKA AND MAGENDIE WITH HYDROCEPHALUS
REPORT OF A CASE IN AN ADULT
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Hydrocephalus as a result of congenital atresia of the foramina of Luschka and Magendie of the 4th ventricle was first described adequately by Dandy and Blackfan.3 The subsequent reports and evaluation by Taggart and Walker6 stimulated increasing interest in this disorder as reflected in the more recent literature. In only one fully authenticated case has the patient been an adult.7 We wish to add a second.

CASE REPORT:

A white man, 31 years of age, had been admitted to hospital on numerous occasions because of intermittent headache and blurred vision.

Family History. No pertinent history, although the patient’s mother was said to have “heart trouble.”

Past History. Bouts of pneumonia occurred when the patient was 18 months, 6 years and 9 years of age, respectively. At 9 years he also had diphtheria, and later measles, pertussis, influenza, and possibly mumps. There were no known complications of these diseases. Venereal disease was denied, as was also tuberculosis. At 13 he suffered a blow to the right parietal area in a sledding accident, and was said to have had a fractured skull. After a week of unconsciousness, he noticed tinnitus and marked loss of hearing in the right ear. The tinnitus soon cleared, but the loss of hearing persisted.

After enlisting in the Army in January 1943, the patient had frequent sore throats and occasional exertional dyspnea. He was admitted to hospital on July 3. His tonsils were found hypertrophied and cryptic. The tympanic membranes and the eyegrounds were normal, as was the remainder of the physical findings. The blood leukocytes were 14,200 per c. mm. Leukocyte differential, urinalysis and bleeding and clotting times were within normal limits. The Wassermann test was negative. Audiograms revealed a nerve type of deafness on the right with some loss for high tones on the left. Following tonsillectomy on July 8, there was an elevation of temperature which responded to sulfathiazole. The patient was dismissed from hospital on July 21.

He was admitted again on April 16, 1944, complaining of precordial pain and a slightly productive cough. He had had an upper respiratory infection 2 months previously, following which he had mild exertional dyspnea. His temperature was 101°F. and pulse was 108. Pleuropericardial friction rub and cardiac enlargement were noted. Leukocytes were 12,000 per c. mm. and erythrocyte sedimentation rate was 33 mm. per hour. Roentgenograms showed evidence of copious pericardial effusion. Rheumatic pericarditis being suspected, the patient was kept in bed and given salicylates.

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On being transferred to a convalescent center in Arizona on June 15, the patient's temperature and pulse were normal. No abnormalities were found on physical examination. Leukocyte count was 11,550 per c. mm. (76 per cent polymorphonuclear neutrophils, 5 per cent eosinophils, 5 per cent stab forms and 14 per cent lymphocytes), hemoglobin 85 per cent, erythrocytes 4.42 million, packed cell volume 36 per cent, and sedimentation rate 30 mm. per hour. The urine was normal except for occasional clumps of polymorphonuclear neutrophils and, on one occasion, several waxy casts. The electrocardiogram revealed T-wave changes which were interpreted as evidence of myocardial damage. Roentgenograms of the chest revealed cardiac enlargement and moderate pleural effusion. Nose and throat cultures were repeatedly negative for beta hemolytic streptococci. Bed rest and intensive salicylate therapy were continued for 2 months. By December 20, the electrocardiogram, erythrocyte sedimentation rate and pulse were all normal. At that time there was moderate cardiomegaly, as shown on roentgenokymograms and an esophagogram. The patient was discharged from the hospital and the Army on Jan. 18, 1945. At no time during his illness had he suffered from involvement of joints or had there been evidence of disease of the central nervous system.

Present Illness. In January 1952, when 31 years of age, the patient first noticed periodic blurring of vision in both eyes. The following month he had attacks of diffuse headache associated with weakness and numbness of the right leg. Slight nausea in the morning hours appeared in March.

He was admitted to hospital April 10 because of “sinusitis.” Following submucous resection, the headaches and visual blurring became more severe. After leaving hospital on April 12, the patient continued to have attacks of headache and numbness and weakness of the right leg. New complaints appeared in the form of diplopia, positional vertigo, tinnitus in the left ear, and numbness over the left lower lip and chin.

The patient was readmitted on May 9, 1952, complaining of blurred vision bilaterally. Examination revealed regular but dilated pupils, bilateral papilledema of unspecified degree, and diminished but equal quadriceps-femoris reflexes. Leukocytes were 12,750 per c. mm., with 63 per cent polymorphonuclear neutrophils, 35 per cent lymphocytes and 2 per cent eosinophils. Lumbar puncture revealed clear, colorless, cell-free fluid with an initial pressure of 255 mm. of water. Protein content was not determined.

Further Examination. On May 13, at another hospital, the heart was questionably enlarged to percussion. Hearing in the right ear was found to be reduced about 50 per cent. There was pronounced papilledema bilaterally, and slight nystagmus on lateral gaze, particularly to the left. The abdominal reflex on the right side was absent.

1st Operation. On May 20, ventriculograms revealed symmetrical dilatation of the entire ventricular system and absence of air over the cerebral hemispheres. Under general endotracheal anesthesia, a suboccipital craniectomy was then performed. The cerebellar tonsils were found herniated into the foramen magnum. Elevation of the tonsils released copious spinal fluid. The inferior vermis was split, exposing a thin membrane over the roof of the ventricle. The membrane was opened widely, exposing the floor of the ventricle and the caudal end of the aqueduct, which was enlarged. Free communication between the basal cisterns and the 4th ventricle having been established, the incision was closed.

Postoperative Course. No complications developed. Repeated lumbar punctures revealed a gradual decrease of cerebrospinal fluid pressure from 310 mm. water postoperatively to 185 mm. water a month subsequently. Papilledema had receded at the time of the patient's discharge from the hospital on June 19.

Interval Course. After several days of well-being, nausea and vomiting set in, and there were bifrontal headaches, staggering gait, and further decrease in visual acuity.

Further Course. The patient was readmitted on July 1, 1952. He was alert and in no distress. Temperature was normal, blood pressure 118/80, and pulse 88. Papilledema was again evident. The left quadriceps-femoris reflex was decreased, and unsteadiness of gait with a tendency to veer to the left was noted. The Romberg test was negative. The region of suboccipital decompression was soft. The cerebrospinal fluid showed 250 mg. per cent of protein with
a cell count of 44 per c. mm., practically all lymphocytes. No organisms were recovered.

On July 2, ventriculograms were much the same as previously, except that air was present in the cisterna magna. The right lateral ventricle was again punctured. Approximately 80 cc. of air were removed and 1.5 cc. of indigo-carmine dye mixed with warm saline were injected. A Tuohy needle was then inserted into the lumbar sac. Fluid flowed freely, but after 40 minutes no dye had appeared. The needle was replaced by a catheter, which was connected to a syringe with a sterile rubber glove for a reservoir. Seventy minutes after its injection into the ventricle, the blue dye appeared in the spinal fluid draining from the catheter.

Fluid drained from the catheter for 10 days in amounts averaging 60 cc. per day. The fluid was at first dye-stained, then xanthochromic, and finally clear and colorless. The headaches disappeared, but the patient did not notice any substantial improvement in visual acuity. On July 19 the catheter was removed.

Pneumoencephalogram on July 15 revealed air in many of the subarachnoid pathways and a small amount in the right lateral ventricle. Subsequently, the patient remained afebrile, headaches were much less severe, and papilledema had decreased. He was discharged from hospital on July 23.

By December 15, the patient’s vision had improved to an acuity correctible to 20/25 bilaterally. Funduscopic examination showed that “the left optic disc is almost flat, the right disc still evidences residual edema.” Soon thereafter the patient’s vision began to fail, papilledema increased, and vomiting set in.

Further Admission, Jan. 10, 1953. The most outstanding abnormalities were unsteady gait with faulty equilibrium, bilateral old papilledema which was most marked on the right, and nystagmus on lateral gaze, especially to the right. Pulse was 88 and regular; blood pressure 135/80. Cerebrospinal fluid: February 9, total protein 29 mg. per cent, no leukocytes; May 4, total protein 90 mg. per cent, 9 leukocytes per c. mm. The visual fields were moderately constricted.

2nd Operation. On February 9, reexploration of the posterior fossa was performed under general endotracheal anesthesia. The posterior inferior part of the cerebellar vermis was again divided. The 4th ventricle was dilated to much the same degree as at craniotomy in 1952. A thin membrane obstructed the flow of fluid from the ventricle. The cerebellar tonsils had herniated into the foramen magnum and were bound down by adhesions. The cisterna magna and the 4th ventricle were opened widely and the cerebellar tonsils were amputated. Free communication of the ventricular and subarachnoid spaces having been accomplished, the incision was closed.

Course. Several lumbar punctures were performed, with the fluid pressure gradually falling to normal. The patient felt well enough to be released from hospital on March 6. When seen on April 6, he reported that his vision had improved and that he had been free from headache.

Final Admission. On May 9, 1953, the patient returned to hospital complaining of nausea, pronounced blurring of vision, and severe occipital headaches. Improvement had been sufficient for him to resume his job as a truck driver, but after several days of work his symptoms recurred. He walked with a wide-based, staggering gait, and the Romberg test was positive.

The patient rapidly became comatose. On May 11 the right lateral ventricle was punctured and 100 cc. of clear cerebrospinal fluid under high pressure were allowed to escape. The patient became lucid. Indigo-carmine dye was then injected into the ventricle and lumbar puncture was performed. After 25 minutes, only a trace of the dye had appeared in the spinal fluid. Several hours later the patient lost consciousness and death quickly ensued. The illness, from the time of development of symptoms of increased intracranial pressure, had lasted 16 months.

Autopsy. Dense fibrous pleural adhesions were noted over the upper lobes of the lungs, and there were fibrinous adhesions at the base of the lungs and between the heart and pericardium. The lungs were moderately edematous. The heart weighed about 500 gm. and was slightly dilated. The wall of the left ventricle was about 1.75 cm. and that of the right ventricle was 0.5 cm. at the points of their greatest diameter. The cardiac valves were normally
formed and without gross lesions. Although there was an acute inflammatory exudate about one of the larger coronary arteries, no evidence of rheumatic fever was found on either gross or microscopic examination of the heart.

**Brain, Gross.** The cerebrum appeared somewhat swollen; its surface was dry. The entire ventricular system was uniformly dilated. On removal of the brain the cerebellum appeared swollen. Abundant fibrous tissue was present over the roof of the 4th ventricle (Fig. 1). The foramina of Magendie could not be identified.

**Fig. 1.** Medulla oblongata and cerebellum, showing pigmented cyst over the roof of the 4th ventricle and part of the cyst wall attached to the cerebellum on each side. (AFIP neg. #54 8197)

Emanating from the cerebellopontile angle and attached firmly to the medulla oblongata and cerebellum on each side was a fluid-filled cystic structure measuring approximately 2 cm. in diameter. Superiorly, each extended to the level of the facial nerves, to which they were adherent, while inferiorly each reached the uppermost spinal cord. On sectioning, both cystic structures were found continuous with the foramina of Luschka, and, through them, with the much dilated 4th ventricle (Fig. 2). Within them was clear dye-stained liquid which obviously was cerebrospinal fluid. Tufts of choroid plexus were found in the roof of the ventricle and along the inner wall of both of the cystic structures. Small hemorrhagic areas were present in the wall of the ventricle, especially on the right side. The block of tissue containing the lateral recesses of both sides was lost subsequently.

**Microscopic Examination.** Sections from several areas of the cerebral hemispheres showed slight thickening of the leptomeninges and occasional hemosiderin-filled macrophages. Scanty
hemorrhage was noted at the site of ventricular puncture, and a small hematoma as well as parenchymal softening in the region of the needle track. The choroid plexuses of the lateral ventricles were unaltered.

Section through the medulla oblongata near the level of the lateral recesses revealed moderate subependymal edema and several small hemorrhages in the ventricular wall. Ependymal epithelium on the part of the lateral recesses available was continuous peripher-

![Image](AFIP neg. #54 8199)

ally and completely lined the cyst walls (Fig. 3). Much of the wall proper was composed of neuroglial tissue having the same structure as that of the wall of the lateral recesses. Externally, the cyst walls were fused with thickened arachnoidal tissue. The foramina of Magendie could not be identified. Because of the dense fibrous tissue resulting from the two operations performed in that area, what remained of the roof of the 4th ventricle was composed almost entirely of fibrous tissue.

**DISCUSSION**

The ependymal and glial nature of the cyst walls and the direct continuity of the cysts with the lateral recesses and 4th ventricle, as noted grossly, seem to constitute sufficient evidence that the cysts represent congenital anomalies. These features rule out the possibility that the cysts arose as a consequence of basal cisternal arachnoiditis secondary to the cranial trauma or the rheumatic disease from
which our patient allegedly suffered. Basal cisternal arachnoiditis, as discussed by Horrax,\(^5\) is easily differentiated from the lesion in our case.

The assumption is that atresia of the outlets of the 4th ventricle existed congenitally and that as a consequence of intraventricular pressure the atretic ends of the lateral recesses gradually ballooned. Porosity of the cyst wall must have been such that ample fluid filtered through it during most of the patient’s life. The eventual internal hydrocephalus may have been caused by increasing leptomeningeal fibrosis about the cysts, as well as over the foramina of Magendie.

We could find no evidence that the vermis was maldeveloped. As a rule, congenital atresia of the outlets of the 4th ventricle with ensuing hydrocephalus (with or without cyst formation as in our case) is associated with dysplasia of the inferior vermis of the cerebellum, a form of rachischisis. As brought out by Taggart and Walker,\(^6\) this disorder usually occurs in early youth and is characterized roentgenographically by enlargement of the posterior fossa, elevation of the transverse dural sinuses, and cystic dilatation of the 4th ventricle. There may also be papilledema. Cases of this kind have also been described by Benda\(^4\) and more recently by Gibson.\(^4\) Occasionally the dural sinuses are not displaced\(^3\) and the vermis of the cerebellum is normally formed, as demonstrated in our case.

### SUMMARY AND CONCLUSIONS

This report concerns a case of hydrocephalus in an adult, associated with large cystic structures in the cerebellopontile angles, the walls of which were continuous
with the 4th ventricle through the lateral recesses. From the histological structure of the cyst wall, as well as for other reasons, the cysts were regarded as outpouchings of congenitally atretic foramina of Luschka. It is assumed that the foramina of Magendie were also congenitally atretic.

REFERENCES


NOTE

Reprints may be obtained from Armed Forces Institute of Pathology, Washington 25, D. C.