CASE REPORTS

CONGENITAL ATRESIA OF THE FORAMINA OF LUSCHKA AND MAGENDIE WITH HYDROCEPHALUS

REPORT OF A CASE IN AN ADULT

CAPTAIN HAL C. HOLLAND (MC), USAF,* AND
COLONEL WISTAR L. GRAHAM (MC), USAF†

Armed Forces Institute of Pathology, Washington, D.C.

(Received for publication August 6, 1957)

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. The subsequent reports and evaluation by Taggart and Walker 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. 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.

* Present address: Branch, School of Aviation Medicine, Gunter AFB, Montgomery, Alabama.
† 3810 USAF Hospital, Maxwell Airforce Base, Montgomery, Alabama.
‡ Armed Forces Institute of Pathology Accession No. 618800.
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