BILATERAL NERVE DEAFNESS, PERSISTENT COUGH AND PAROXYSMAL HYPERPNEA DUE TO A TUMOR IN THE FLOOR OF THE FOURTH VENTRICLE

DANIEL WELLER, M.D.*

Neurological Service, Lenox Hill Hospital, New York, New York

(Received for publication June 20, 1944)

A typical cases at times are instructive concerning the physiopathologic mechanisms that come into play as a result of disease or disturbance of certain vital centers. Such cases are of importance in that they confirm or refute accepted physiologic principles based upon observations on animals or normal persons. Because of these facts, the following record is presented.

REPORT OF CASE

A 22-year-old nurse was first admitted to the Lenox Hill Hospital, New York City, on October 20, 1941, because of bilateral deafness. Nine months previously onset of rapid, progressive deafness in the left ear had occurred, and three months later impairment of hearing in the right ear. In addition, for several months she had had frequency of urination and had become fatigued easily. There had been no otorrhea, tinnitus, dizziness, headache or disturbance in equilibrium. The family and past personal histories were noncontributory.

Examination. The patient was ambulatory, afebrile, cooperative and alert. In repose the facial expression was somewhat drawn and blank. The heart and lungs were normal. The ears, nose and throat were essentially negative.

On neurologic examination the fundi and visual fields were normal, as were the other cranial nerves, with the exception of the auditory nerves. Bone and air conduction were poor on both sides. Caloric tests showed active labyrinths bilaterally. The deep reflexes were exaggerated, those on the right side being greater than those on the left. There was transient right ankle clonus. The abdominal reflexes were weak to absent in all four quadrants. There were no pathologic reflexes. There were no motor weaknesses, sensory disturbances or cerebellar signs.

The hemoglobin was 95 per cent; erythrocytes 4,380,000; and leukocytes 9,950, with polymorphonuclears 52, lymphocytes 28 and monocytes 1 per cent. The Wassermann reaction was negative. The creatinine was 0.5, uric acid 3.6, sugar 83, and CO2 48.1 mg. The spinal fluid examination gave the following results: Initial pressure 80 mm. of water, cells 1, protein 18, glucose 59 and chloride 743 mg.; the Wassermann test and the colloidal gold curve were negative. The basal metabolic rate was +9 per cent.

Course. The patient was discharged on October 30, 1941, no diagnosis having been established.

Second Admission. She was readmitted to the hospital on February 9, 1942, complaining of severe, throbbing frontal headaches, sometimes relieved by lying down. Nausea and vomiting occasionally were associated with these headaches. She believed that the headaches followed the original spinal tap. In addition, she occasionally dropped objects from the right hand. The neurologic status was essentially the same, except for questionable nystagmus on lateral gaze. Lumbar puncture showed an initial pressure of 120 mm. of water, normal dynamics and normal chemistry. Roentgenologic examination of the skull was negative. Ventriculograms were reported as normal. She was discharged on March 1, 1942, without a diagnosis.

* Fellow in Neurosurgery, The Lahey Clinic, Boston, Massachusetts.
Interval Note. The patient remained at home and was asymptomatic except for impaired hearing until five weeks later.

Third Admission. When she reentered the hospital on April 22, 1942, she had had severe headaches and vomiting for a week.

The neurologic examination revealed normal fundi, bilateral nerve deafness and occasional nystagmoid jerks on lateral gaze. The deep reflexes were hyperactive, those on the right being greater than those on the left. The abdominal reflexes were absent. There was no Babinski or Hoffmann’s sign, motor weakness or spasticity. Light touch, pinprick, position and vibration sensations were diminished over the right arm, trunk and leg.

One week later she began to complain of a difference in the two sides of the body and stiffness of the right arm and hand. The headache became unbearable. The pupils were moderately dilated and reacted sluggishly to light. Well maintained nystagmus was present on lateral gaze. The hemihypesthesia became more pronounced, and the sensory loss involved the right side of the face.

Within 24 hours the patient developed a dry cough, which was resistant to all types of medication. The lungs were clear to auscultation and percussion. Roentgenograms of the chest were negative, and fluoroscopic examination showed normal movement of both leaves of the diaphragm. The dry, rasping character of the cough and its occurrence at regular intervals during an hour, as well as its resistance to medication, suggested central origin. Cocainization of the posterior pharynx and larynx was tried unsuccessfully in an attempt to break the reflex arc, and basal anesthetics were necessary to prevent exhaustion. The cough developed into a peculiar rasping, expulsive shriek. The patient vomited and retched frequently. Since she was unable to take food by mouth, parenteral feedings were begun. She became hoarse, was able to speak only in a whisper, and also began to have periodic tremors of the entire right side of the body. Bronchoscopy failed to reveal the cause of the cough. At the end of the second week in the hospital she complained of weakness on the right side. There was objective evidence of a right hemiparesis with exaggerated reflexes and a right Hoffmann’s sign.

Operation. Because of the patient’s persistent cough and rapid decline, ventriculograms were again performed, but failed to show any abnormality of the ventricular system. In spite of this, a suboccipital exploration was performed on May 9, eighteen days after admission. Upon opening the posterior cistern, a smooth, bluish, elongated mass was disclosed attached to the posterior aspect of the medulla. This mass extended into and was also attached to the floor of the fourth ventricle, and extended downward to the level of the atlas. It was attached firmly to the cord and was impossible to remove, but some specimens were taken for biopsy. The histologic report was medulloblastoma.

Postoperative Course. As soon as the decompression was accomplished, pronounced improvement in the patient’s respiration and general condition occurred. By the following day the paroxysmal cough had disappeared and, except for a right upper monoplegia, she was fairly comfortable. Ten days postoperatively deep radiation therapy to the suboccipital area was started.

The patient ran a stormy course with paroxysms of hyperpnea and apnea, and at times she was cyanotic, cold and clammy. Carbon-dioxide determinations, following a prolonged attack of hyperpnea, revealed a marked acidosis. Intense alkali, blood, glucose and saline infusions restored the blood chemistry to within normal limits, and on about the 28th day she began to improve. The respiratory crises, although of a milder degree, continued until a few days before dismissal on the 49th postoperative day.

The neurologic examination on discharge showed that she was comfortable and free from cough, respiratory crises, vomiting and nausea. The pupils reacted promptly to light and in accommodation, and the fundi were normal. The only persistent signs were the right-sided hemianesthesia and weakness of the right upper extremity.

Interval Note. When seen in the follow-up clinic two months later she was able to walk, and felt well, except for a persistent headache and occasional bouts of respiratory difficulty. The