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

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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, pro-
gressive 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 disturb-
anee 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 ex-
aggerated, 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 cerebel-
lar 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 88, 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 estab-
lished.

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.

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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 38th 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
cranial nerves were essentially negative. She was able to write her name, and the motor power in the right side definitely had improved. However, the right hemianesthesia persisted in all modalities. The operative area was well healed and not bulging.

*Fourth Admission.* The patient returned to the hospital on October 21, 1942 for further radiation therapy. The neurologic status was essentially the same. Her facies was pale and gave the impression of chronic ill health. After every radiation treatment she had a respiratory crisis characterized as follows: Within a few hours she began to breathe rapidly. The respirations were shallow, often simulating the panting of a dog, and varied from 60 to 112 a minute. The breathing was so labored that each inspiration and expiration was attended by a simultaneous lateral propulsion of the head as the accessory muscles of respiration came into play. This hyperpneic phase, which lasted from a few hours to as long as forty hours, then subsided until the respirations were down to 8 to 10 a minute. During this apneic phase, which often persisted for as long as sixteen hours, she was comatose.

Roentgenologic examination of the chest on October 23 revealed changes simulating a pneumonitis, although physical examination failed to disclose any positive signs of involvement of the lung, and the patient's temperature was normal. Because of the respiratory embarrassment, which apparently was due to radiation therapy, smaller doses were attempted but were not tolerated. There was no change in the neurologic status. Roentgenologic examination of the lungs three weeks after admission was essentially the same as on admission. The patient was discharged on November 2, 1942, to the care of her family physician, and further information concerning her course is not available.

**DISCUSSION**

Although tumors of the fourth ventricle do not produce a typical clinical syndrome, they seldom present such a misleading sign as bilateral deafness. In most instances, regardless of the initiating symptoms, signs of increased intracranial pressure, with or without involvement of the pons, medulla or cerebellum, appear. Craig and Kernohan list headache, choked disk, vomiting, ataxia, diplopia, stiff neck and hiccups as predominant in their series of 82 cases. All these symptoms result from either obstruction to the cerebrospinal fluid circulation or direct local involvement of the medulla by the tumor mass itself.

Bilateral nerve deafness, central cough and paroxysmal hyperpnea, without signs of increased intracranial pressure, were the outstanding features in this case. In order fully to appreciate this bizarre picture, one need only recall the compactness of the vital centers and the numerous structures in the floor of the fourth ventricle. In addition to the three important vagal, hypoglossal and acoustic trigones, there are the three cerebellar peduncles, and the nucleus of the sixth nerve (eminencia teres). Adjacent and posteriorly are the expanded parts of the posterior columns named the clavus and cuneus.

Regardless of our knowledge of the anatomic arrangement in this area, the neurophysiologic mechanisms involved are difficult to understand. Brain tumors, other than cerebellopontine angle tumors, do not play an important part in nerve deafness, although it does occur with midbrain lesions, usually late in the course of the disease. Studies by Craig and Kernohan, Gibbs, Drury and Northington fail to mention examples of fourth ventricle tumors as a cause of such deafness, although the position of the acoustic apparatus
in the floor of the fourth ventricle is so superficial as to be vulnerable. Nevertheless, deafness may occur late in chronic, focal pressure on the brain stem, but in these instances it is usually unilateral.

The respiratory and coughing mechanisms are regulated by centers located in the lower part of the medulla, primarily from impulses along the vagus nerve and tractus solitarius. Under normal conditions they respond to variations in the carbon-dioxide content of the circulating blood and stimuli from the respiratory mucosa, as well as impulses set up by the expanding and contracting lung. Theoretically, it is remarkable that disturbances of respiration, such as were present in this case, are not more frequent in cases of fourth ventricle tumors, although they have been noted in cases of encephalitis with acute bulbar symptoms. Usually one encounters the Cheyne-Stokes type due to tonsillar herniation.

In the case reported the breathing was of the Biot type, in which the onset of hyperpnea and apnea is paroxysmal. As a result of this anoxia, the patient was cold, cyanotic and lightheaded. Continued hyperpnea induced an acapnea, and during this phase the patient's respiration frequently was decreased below 8 a minute. Contrary to experimental findings, there was no consistent correlation between the hyperpneic phase and the pulse rate and blood pressure. The blood pressure was maintained on a steady level. At times the pulse rate was increased and at other times it was unchanged.

The secondary lung changes consequent to the protracted hyperpnea, which undoubtedly were due to vascular changes in the parenchyma of the lung, were most interesting. The nature of the lesion and the slow progression of the changes in the lung, as shown by roentgenologic examination, were not characteristic of metastasis but rather of a pneumonitis.

The neurologic signs pointed to direct involvement of the posterior columns, the pyramidal tract and the spinothalamic pathways. The true nature of the lesion was not suspected until these structures became involved.

**SUMMARY**

A case is reported of a tumor of the fourth ventricle which manifested itself by signs of an unusual disturbance in the respiratory centers, together with deafness. Tract signs were not produced until late in the course of the disease. The disturbance in the respiratory center produced paroxysms of the Biot type of breathing and periods of uncontrollable coughing. The changes in the parenchyma of the lung were secondarily induced, probably based upon alterations in the blood vessels of the lung.

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