The association of a tumor of the fourth ventricle with increased intracranial pressure that apparently was relieved spontaneously by persistent cerebrospinal rhinorrhea (Fig. 1) is the subject of this report. The present case serves to re-emphasize the warning of Locke that the sudden unexplained development of cerebrospinal rhinorrhea warrants classification of the patient as a "brain tumor suspect."

Since Miller’s classic paper of 1826 describing spontaneous cerebrospinal rhinorrhea, numerous reports relevant to this subject have appeared. Among these have been accounts of spontaneous discharge of cerebrospinal fluid from the nasal passages in association with brain tumors. Usually these have been pituitary adenomas that had eroded the floor of the skull, permitting egress of fluid into the nasal sinuses, or tumors producing increased intracranial pressure. In 1908 Vigou-
roux\textsuperscript{10} described a case of spontaneous cerebrospinal rhinorrhea in a 28-year-old man, which was associated with a papillomatous tumor of the choroid plexus of the fourth ventricle that produced ventricular enlargement. Necropsy revealed a communication between the subarachnoid space and the nose.

The case to be reported is that of a young woman in whom cerebrospinal rhinorrhea appeared after ventricular obstruction and increase of intracranial pressure had resulted from two large epithelial-lined cysts of the fourth ventricle.

**REPORT OF CASE**

A 22-year-old housewife was referred to the neurosurgical section of the Mayo Clinic in August, 1956 because of persistent cerebrospinal rhinorrhea.

In 1953, while at work, the patient had noted a spontaneous right nasal discharge of clear fluid. This had continued unabated; in December, 1954 and again in February, 1955 she had required lengthy hospitalization for severe attacks of meningitis. Neurosurgical consultation elsewhere had established the diagnosis of cerebrospinal rhinorrhea and the patient had undergone operation in an attempt to correct this condition. Thus, on March \textsuperscript{21}, 1956, a right transfrontal craniotomy had been performed with the repair of a cerebrospinal fistula in the region of the right cribriform plate. At the time of operation the right lateral ventricle had been cannulated and had been found to contain an excess of fluid, indicative of hydrocephalus. Consequently the ventricular system had been inflated with air and routine ventriculograms had been made after closure of the surgical wound. These films had revealed symmetrical dilatation of the ventricular system as far as the aqueduct. No air had been visualized in the fourth ventricle.

Although the patient's postoperative course was uneventful, fluid had continued to drip from her right nostril. One month after operation (April, 1956) she had noted the development of diplopia when looking to the left. Four months after operation (July, 1956) she began to have severe headaches, accentuated by coughing and associated with vomiting; in addition, the patient believed that in July her gait had become unsteady and her vision had decreased.

When examined at the Mayo Clinic in August, 1956, the patient seemed in good health except that clear fluid drained from her right nostril whenever she leaned forward. Findings from routine physical examination and laboratory studies were normal.

Neurologic examination revealed absence of smell in the left nostril, an ataxic gait, slight hyperactivity of muscle stretch reflexes in the left extremities and a minimal bilateral decrease in coordination, but no objective motor weakness. Ophthalmologic examination revealed paralysis of upward gaze, retractive nystagmus, inequality of pupils, weakness of convergence, and skew deviation. There was mild pallor of the optic fundi suggestive of previous papilledema. Visual fields were normal, with enlarged blind spots. These findings were considered consistent with midbrain-pontine involvement.

Although the findings from routine laboratory tests were normal, cranial roentgenograms revealed enlargement of the sella turcica. Metabolic investigation was undertaken to determine the possibility of a pituitary tumor. The results of review of the patient's endocrine history and examination of endocrine systems, including special laboratory tests (urinary 17-ketosteroids and corticoids, plasma corticoids), were within normal limits. The fluid collected from the right nostril was found to contain 74 mg. of sugar per 100 ml.

One of us (JGL) saw this patient in consultation on August 4 and advised further neurosurgical investigation in the form of encephalography, and ventriculography if necessary. The patient and her husband decided to return home and discuss the situation with their home doctor.

On August 10 the patient was admitted to a Rochester hospital in a status of emergency. She was found to be acutely ill with fever, headache and vomiting. Administration of penicillin and sulfadiazine was begun immediately. By August 15 her condition had improved markedly and it was possible to proceed with the diagnostic studies. Lumbar pneumoencephalography
was attempted, but the ventricular system was not visualized. Then ventriculography was performed, utilizing the anterosuperior burr hole in the forehead (present from her original craniotomy). Eighty ml. of ventricular fluid were replaced with 75 ml. of air. The ventriculograms demonstrated a large spherical mass in the fourth ventricle (Fig. 2) with a high degree of obstruction in the aqueduct and dilatation of the lateral and third ventricles. Suboccipital craniotomy was performed forthwith, and two large epithelial-lined cysts (Fig. 3), filling the hugely dilated fourth ventricle and obstructing the aqueduct, were found. One cyst contained an ounce of cerebrospinal fluid and the other contained $\frac{3}{4}$ ounce. There was no evidence of additional aqueductal obstruction. During the operative procedure, which was performed with the patient in the upright position, copious amounts of fluid continued to drain from the right nostril.

The patient’s immediate postoperative period was most gratifying, for during the 1st week she was free of cerebrospinal rhinorrhea and improvement in her visual disturbances (principally nystagmus and diplopia) was recorded. But during the 2nd postoperative week clear fluid began again to drain from the right nostril.

On September 5 surgical correction of the cerebrospinal rhinorrhea was attempted. A coronal scalp incision was made, utilizing part of the original scalp incision. The bone flap established in March, 1956 was elevated again and adequate exposure was obtained by tapping the right lateral ventricle. This disclosed a small opening in the right frontal sinus which previously had been plugged with wax and two small holes in the dura mater near the right

![Fig. 2. Ventriculogram demonstrating the dilated fourth ventricle and tumor of the fourth ventricle outlined by air. Note also the old surgical defect in the bone.](image)
orbit. Also, there was a fistulous tract through the anterior portion of the cribriform plate on the right (Fig. 1). The dural defects and the fistulous tract (after it had been divided) were closed with dural silk. The cribriform plate and the suture line were reinforced with muscle pledgets and additionally a thin piece of polyvinyl formal (Ivalon) sponge was placed between the muscle pledgets.

Postoperatively the discharge of fluid from the nose was markedly reduced and it was hoped that as the sponge became impregnated with fibrous tissue the rhinorrhea would cease. On September 10 the patient was dismissed to return home, with the surgical wound well healed (Fig. 4).

Fig. 3. Photomicrograph of inner and outer walls of epithelial cyst of fourth ventricle. (Hematoxylin and eosin, ×60)

Fig. 4. Photographs showing the incisions in the skin that were utilized, and the excellent healing of the wound.
CSF RHINORRHEA WITH TUMOR OF FOURTH VENTRICLE

Correspondence with her revealed, however, that clear fluid continued to drain from her right nostril when she leaned forward and into her throat when she lay flat. Consequently the patient was readmitted to the hospital in January, 1957 for reoperation. Neurologic and ophthalmologic examinations attested to the success of the surgical removal of the cysts from the fourth ventricle. Vision and gait were recorded as normal and only a trace of diplopia (on gazing to the left) and nystagmus remained.

On January 9 the old right craniotomy wound was opened again. A 1-inch opening was trephined above the left frontal sinus and to the left of the sagittal suture, thus making possible bilateral exploration of the cribriform plate. Fluid was found escaping through the opening still patent in the right frontal sinus. The dura mater, which was markedly adherent to the cribriform plate bilaterally, was stripped away. No actual fistulous opening could be identified positively. The dura mater from the cribriform plate was closed with continuous dural silk and the dural suture line was reinforced with pledgets of muscle and Gelfoam.

Although the patient’s recovery from operation was satisfactory, on the 8th postoperative day once again clear fluid was dripping from her right nostril and a soft bulge was noted in the middle of her forehead. The amount of this drainage varied from 250 to 400 ml. per day. On January 18 continuous subarachnoid drainage by means of a ureteral 4 catheter was instituted. As a result of this therapy a marked reduction in nasal drainage was accomplished (no drainage, to only a few drops per day). Catheter drainage was maintained for 10 days and terminated January 27. On January 30 general malaise, headache, and stiffness of the neck developed. Immediate lumbar puncture gave evidence of a bacterial infection and the cultures of the spinal fluid showed Staphylococcus faecalis. Vigorous antibiotic treatment, including intrathecal administration of penicillin, soon brought the meningitis under control. When the patient was dismissed to return home on February 23 she had had no nasal drainage since February 1, the longest such period since the symptoms began.

Follow-up correspondence revealed that during the spring and early summer of 1957 the patient noted the appearance of a few drops of fluid in her right nostril during the first half hour she was up in the morning. During the late summer, fall and winter of 1957 even this was an infrequent occurrence. Since March 1958 the patient has been completely free of cerebrospinal rhinorrhea. In July 1958 she completed a normal pregnancy without incident. In every way she has felt normal and healthy.

COMMENT

In 1958 Norsa8 reviewed the published accounts of intracranial tumor associated with spontaneous cerebrospinal rhinorrhea. Reports of 18 verified tumors were found, including 6 pituitary tumors and 12 miscellaneous supratentorial and infratentorial tumors. In Norsa’s list the only tumor of the fourth ventricle associated with cerebrospinal rhinorrhea was a papillomatous lesion of the choroid plexus, originally described by Vigouroux.10 This same case was mentioned by Locke,3 in his review of the problem in 1926. So far as we know, our case is the second of its kind to be reported.

In this case a fistulous communication between the subarachnoid pathways and the nasal passages reduced the excessive intracranial pressure, as was evidenced by the resolution of the optic disks. Just how this method of drainage accomplishes reduction of increased intracranial tension associated with ventricular obstruction is not known. Some cases8 have been reported in which direct communication existed between the ventricular system (usually the anterior horn of the lateral ventricle) and the subarachnoid space. In others,6,8 as in our patient, internal hydrocephalus existed with no abnormal opening present in the cerebral substance to make the ventricular decompression possible. Yet in some way cerebrospinal rhinorrhea contributed to the reduction of excessive intracranial pressure. It has been suggested in the past that ventricular obstruction may be incomplete.3 Alterna-
tively, it may be that so much fluid normally is secreted over the surfaces of the brain that removal of it is sufficient to alter intracranial-pressure relationships favorably.

This case illustrates the importance of eliminating any possibility of a brain tumor in the presence of spontaneous cerebrospinal rhinorrhea, as well as the futility of attempting corrective operation in the face of ventricular obstruction and elevated intracranial pressure. It also emphasizes the usefulness of instituting continuous spinal drainage after repair of cranionasal fistula, so that reduction of subarachnoid pressure may permit occlusive healing.

SUMMARY

In the case reported, spontaneous cerebrospinal rhinorrhea was cured by a corrective operation after the surgical removal of an associated tumor of the fourth ventricle.

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