This is an autobiographical case report of a physician born in 1908 who developed diabetes insipidus in 1920, and in whom a craniopharyngioma was detected in 1938. A unique feature of the case is the spontaneous drainage into the nasopharynx from cystic portions of the craniopharyngioma resulting from tumor penetration through the sphenoid bone. In spite of various medical problems, both directly and indirectly connected with the craniopharyngioma, I have had a successful career as a student, surgeon, and educator.

Case Report

Onset of diabetes insipidus in 1920 at 12 years of age required leaving school for a 5-month period. The polydipsia was severe and the 24-hour urinary output varied between 6 and 8 liters, with a specific gravity of 1.001. The nature of the medical problem was not understood in 1920, according to the patient's father, a physician. No data concerning the skull x-ray films in 1920 are available now, although various radiographic studies were performed without a diagnosis being established. There was gradual spontaneous improvement in the diabetes insipidus which permitted a return to school. Significant polyuria did not recur at any later date. Mild headaches recurred sporadically during the subsequent years, throughout high school, college, medical school, internship, and surgical residencies.

In 1938, headaches became more frequent and severe. It became increasingly difficult to function as a resident surgeon. A bizarre manifestation was a pounding pulsation concomitant with each heartbeat which developed in the head and upper neck after climbing a flight or two of stairs. The diastolic blood pressure was found to drop momentarily to about 20 mm Hg, yielding a pulse pressure of over 100 mm Hg for a few minutes while a prominent carotid pulsation in the neck was evident. During these episodes, the head pulsations were particularly painful. There was no evidence of primary cardiovascular disease. On suspicion of a brain tumor, skull x-ray films were ordered, and revealed bone destruction of the dorsum sellae, the adjoining portion of the clivus, and the floor of the sella (Fig. 1). A tumor mass was demonstrated in the left maxillary sinus. There was never any unequivocal visual field defect.

The neurologists and neurosurgeons who were in charge of the case at that time did not recommend surgical intervention, largely because of the poor surgical results that were being obtained then. Opinion as to the diagnosis was divided between a pituitary tumor and a craniopharyngioma; very little calcification was evident on the roentgenogram at the time. In the 1930's, Dyke and Hare2 were treating pituitary tumors with radiotherapy. Dyke instituted a similar course of treatment in this case, consisting of a total dose of 2400 rads.
Craniopharyngioma with nasopharyngeal drainage

with 200 kV administered through three ports (two lateral and one anterior). The usual treatment plan was for two to three courses of irradiation. Dramatic improvement in symptoms occurred early, even before the therapy was concluded, and therefore the usual second course was not given. About 3 months later, the patient resumed full professional activities and continued in practice for over 24 years. Only at that later time was further radiotherapy required.

At the time that a pituitary tumor was suspected in 1938, there was only slight evidence of calcification in the tumor area. By 1950, extensive calcification was evident, and Dr. Bronson Ray, who was the neurosurgeon in charge then, was sure that the tumor was a craniopharyngioma. There was early evidence of impaired immunity. The patient contracted recurrent scarlet fever and mumps. Impaired pituitary function has required cortisone and thyroid therapy for over 30 years. Infections of various types have occurred. An abscess might develop with little evidence of redness and tenderness.

In the 1950's, it was noted that the patient's bed pillow was occasionally stained with a yellowish fluid which had drained from his mouth during sleep. However, during the last 10 years or more, the drainage into the nasopharynx has increased and is now noted upon awakening or during the day. Drainage is usually evident by the presence of a very characteristic unpleasant taste in the throat, whereupon the motor oil-like fluid can be expectorated. Others nearby may note a pungent, somewhat rancid aroma. Such drainage has occurred on an average of once a week over the years, but sometimes it does not occur for a month or so. About half the time only yellow material, obviously mixed with some pharyngeal mucus, is expectorated; at other times it is mixed with a very small amount of blood. Occasionally, such drainage might occur intermittently during several days in succession. Pain in the head tends to be more prominent just prior to a drainage period, but at other times pain and drainage are not closely related. During a period in 1982 and 1983, the drainage sometimes dropped onto the larynx and caused sudden, unexpected, violent coughing spells.

The drainage material has repeatedly been cultured, chiefly when blood was apparent. The drainage material was identified as almost pure cholesterol crystals. At rare times, tiny fragments of tissue were seen; these showed cells consistent with a craniopharyngioma (Fig. 2). Occasionally, a moderate increase in leukocytes was noted. No meningeal inflammation has ever occurred.

During 1963, the patient had increasing difficulty in carrying out his professional duties. He lost consciousness on several occasions while he was on a lecture trip and during surgical meetings. In December, 1963, he was hospitalized and, during a long glucose tolerance test, he suffered an epileptic-type convulsion with dislocation of the right shoulder which required reduction. On consultation, further radiotherapy was decided upon. A total of 4100 rads was then administered through two lateral ports in December, 1963, and January, 1964, which was well tolerated. The patient resigned as a Director of Surgery at this time, but continued work at a reduced level until 65 years of age when he ceased to perform surgical operations.

In 1971, stereoscopic radiographic examination of the base of the skull showed destruction and complete erosion of the sella turcica with intrasellar calcification. A mass extended into the sphenoid sinuses with erosive destruction of the petrous pyramid region on the left side. There was also a rounded area of soft-tissue den-
FIG. 3. Tomogram showing the nasopharyngeal aspect of the sphenoid bone.

sity, 1.1 cm in diameter, extending into the posterior nasopharynx to the left of the midline. In 1977, another polytomographic study (Fig. 3) showed an irregular mottled eroded appearance of the basisphenoid adjacent to the nasopharynx with partial erosion of the osseous portion of the nasopharyngeal wall. No clearly defined mass was demonstrated in the nasopharynx. The asymptomatic tumor mass in the maxillary sinus has shown no radiographic change since 1938; it is apparently not an adamantinoma but probably an osteoma and hence an unrelated finding. In 1982, steroid myopathy became evident. Difficulty in rising from a chair was noted and impairment in the full opening of the mouth during eating is a problem. Consultants now believe that no further radiotherapy can be given safely for the continuing pains in the head, which require medication. Symptoms and disabilities were not considered to be of sufficient degree in 1972 to warrant surgical intervention at that late date. At present, at 76 years old he makes only occasional contributions to the scientific literature.

Discussion

Occasionally, a craniopharyngioma may be found largely within the sphenoid sinus, but the very unusual features of this case are the penetration of this craniopharyngioma through the sphenoid bone into the nasopharynx, unassociated with any surgical intervention, and its long persistence without serious complications. The earliest evidence of drainage of cystic portions of the craniopharyngioma into the nasopharynx was with the drooling of yellow fluid from the mouth during sleep in the 1950's. Even in the absence of surgery, the cystic portion of a craniopharyngioma may extend and rupture into the meninges with a resultant aseptic meningitis, which is sometimes recurrent. A craniopharyngioma may also rupture into the nares; most of these rare reports are found in otolaryngological journals. Prolonged drainage of cystic portions of a craniopharyngioma into the nasopharynx unassociated with surgery, as in this case, has apparently not been previously documented. For such a complication to develop, the craniopharyngioma has to penetrate into the sphenoid sinus and through the bone floor of the sphenoid sinus into the nasopharynx. The long survival time in this case probably gave an opportunity for this development.

The first documented leakage of cerebrospinal fluid (CSF) through the nose occurred some time after the first course of radiotherapy when some of the fluid was caught in a test tube and subjected to appropriate laboratory examination. One episode of massive CSF rhinorrhea occurred in 1964 several months after the second course of radiotherapy. At that time, there was intermittent leakage of CSF through both nostrils over a period of 2 days, with a sudden gush at one time. Recently, there has been occasional intermittent CSF rhinorrhea associated with a sneeze or cough. No signs or symptoms of meningeal irritation followed either of the episodes of CSF rhinorrhea or occurred at any other time. Hypothalamic symptoms such as chilly sensations and subnormal temperatures have been common throughout the years. Periorbital edema is usually evident in the morning.

There is no family history of brain tumor or congenital anomalies. There was no impairment in the development of secondary sexual characteristics at the time of the diabetes insipidus in 1920. When the patient was 39 years old, over 10 years after the first course of radiotherapy, he married a physician. Their son is also a physician.

References


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Address reprint requests to: Herbert C. Maier, M.D., 50 East 77 Street, New York, New York 10021.