Encephalocele Extending into the Sphenoid Sinus

Report of a Case

DAVID DANOFF, M.D., JOHN SERBU, M.D., AND LYLE A. FRENCH, M.D.

Division of Neurosurgery, Department of Surgery, University of Minnesota, Minneapolis, Minnesota

Congenital herniations of brain tissue through the base of the skull have been described since the mid-nineteenth century. Virchow in 1863 referred to a museum case in which such a herniation presented in the pharynx of a stillborn. A sufficient number of cases had been reported by 1882 for Heinecke to delineate 3 types of basal encephaloceles according to their anatomical locations. These were (a) spheno-pharyngeal, in which the hernia passes through an opening in the ethmoid or sphenoid bone or through the suture between them and presents in the nasal or naso-pharyngeal cavity; (b) spheno-orbital in which the mass protrudes through the superior orbital fissure, and (c) spheno-maxillary in which the encephalocele passes through both the superior and inferior orbital fissures into the pterygopalatine fossa. Apparently Heinecke considered these developmental abnormalities to be no more than anatomical curios. Fenger quotes him: "Cephalocele basalis is of no surgical importance as it has been found only in nonviable monsters." However, 13 years later a case was recognized in a 29-year-old man and successfully repaired by an intranasal approach. Subsequently basal herniations of the brain became recognized as occasional causes of nasal obstruction and of cerebrospinal rhinorrhea. By 1938 Mood was able to collect 40 cases of anterior encephalocele and references to 30–32 others in older literature. Approximately half of these were of the basal variety. In spite of the number of articles dealing with procedures to repair such defects, basal encephaloceles actually are comparatively uncommon lesions. Ingraham and Matson in a review of 20 years of experience at Boston Children’s Hospital, found 546 children with meningoceles. Eighty-four of these had encephaloceles, only 6 of which presented as intranasal masses and only 1 had herniated through a defect in the cribiform plate. A review of the literature in 1964 found 70 case reports and an estimated incidence of one basal herniation in every 35,000 live births.

On reviewing the indications for the various types of intracranial and intranasal repairs for these lesions, McCoy presented another variant of basal encephalocele, a thin walled meningocele protruding through a .5X1 cm. dehiscence in the lateral wall of the sphenoid sinus. The patient (Case 3) was a 40-year-old housewife who had had recurrent attacks of rhinorrhea. McCoy resected the lesion via an intranasal approach. The bony defect was closed in a second procedure with a flap of mucoperichondrium.

Von Nothuis and Bruyn in 1964 presented a case of a 4-month-old boy with still another variant. In this patient there was a large midline defect in the sphenoid bone and several cerebral abnormalities. The patient survived operation by the intracranial route but died from intracranial hypertension and tentorial herniation 1 week later.

The purpose of this paper is to present a second case of encephalocele of the sphenoid sinus and to review briefly the embryologic factors which allow such a malformation to occur. The case under discussion was successfully repaired via an intracranial route.

Case Report

U.M.H. #1092221, L.J., a 41-year-old white housewife, was admitted to the University Hospital on April 24, 1963, for evaluation of persistent rhinorrhea. The first attack of rhinorrhea had occurred the preceding October, in conjunction with her menstrual period. Nasal drainage appeared the following month with the onset of menstruation but continued beyond the end of the menstrual period. The rhinorrhea was so profuse that the patient slept with a bath towel under her head to prevent soaking her pillow. The patient described the fluid as clear in color and salty in taste, and stated that the drainage, which came almost exclusively from the left nostril, increased when she bent her head forward. She had seen an otorhinologist in February, 1963, and after diagnostic x-ray studies was treated with various drugs for "sinusitis." There was no other history of sinus infection nor of allergy, epistaxis, tinnitus, deafness or otolaryngologic surgical procedure. The patient had hit her head on a fence about 10 years prior to admission but this was not severe enough for her to lose consciousness nor was there otorrhea or rhinorrhea at the time. A possibly significant feature is that 2 years prior to her admission to the University Hospital she seemed, for no apparent reason, to smell an ether-like odor. She experienced a similar episode in April, 1963. She denied any loss of her sense of smell, visual disturbance, headaches or seizures.

Examination. General physical and neurological examination at the time of admission were normal except for the presence of clear cerebrospinal fluid draining from the left nostril. No evidence of intranasal mass or old nasal fracture was found. Skull roentgenograms on May 3, 1963, demonstrated a normal sella but there was

Received for publication June 14, 1965.
a rounded mass-like shadow, with sharp margins, visible in the left side of the sphenoid sinus (Fig. 1). The left anterior clinoid was small and less well demarcated than the right. The radiologist suggested that this appearance was more indicative of a meningioma invading the sinus than of a primary lesion of the sphenoid sinus. Laminograms confirmed the presence of the mass. Free fluid was demonstrable in the sinus cavity. Radioisotope scan, electroencephalography with nasopharyngeal leads, visual fields, and a left carotid angiogram were normal.

Operation. A left frontotemporal craniotomy was performed on May 8, 1963. The dura over the cribriform plate was visualized and found to be intact. The tip of the temporal lobe was retracted and a small hole was found in the anteromedial wall of the middle fossa connecting the middle fossa with the sphenoid sinus (Fig. 4). Pia-arachnoid and brain tissue from the anteromedial part of the temporal lobe protruded through this bony defect. A silver clip was placed across the neck of the protrusion and it was then severed by electrocoagulation. The mass was removed from the sphenoid sinus (Fig. 8), the sinus plugged with muscle and a plastic closure of the dural defect was carried out, using temporalis fascia. Microscopic examination of the herniated mass showed "normal-appearing nerve cells and glia but the usual cortical architecture was distorted. There was

Discussion

Since the earliest reports of encephaloceles, there has been considerable speculation on the anatomical and embryological factors responsible for such lesions. In 1837, Geoffroy-St.-Hillaire published his "adhesive theory" attributing the bony defect to adhesions between the brain and dura on one side and the skin overlying the cranium on the other. This allegedly resulted in a localized arrest of development of the bony vault with subsequent outpouching of the brain. As knowledge of the development of the human skull increased, the theory was modified to state that these hernias can occur because of clefts in the skull which are present either normally or pathologically between the various bones during their development. The case presented above does not violate this rule.

The body of the sphenoid bone, as outlined by DeBeer, develops in an orderly fashion by the fusion of basi- and presphenoid elements. The basisphenoid, which ossifies first, arises from the fusion of paired centers in the central cartilaginous stem and another pair of ossification centers in the processus alaris. The presphenoid arises from no less than 5 centers (lateral paired centers which arise in the ala hypochiasmatica, medial paired centers which arise in the central cartilage mass and a median unpaired center which ossifies in the back part of the nasal septum). The medial