Primary intranasal encephalocele

Report of four cases

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Four cases of primary intranasal encephalocele are presented. Three of the patients had been treated for nasal polyps. One of these three patients presented with persistent cerebrospinal fluid (CSF) rhinorrhea after a fourth polypectomy, another with recurrent CSF rhinorrhea and bacterial meningitis following a second polypectomy, and the third case with recurrence of meningitis, also following polypectomy. Recurrent bacterial meningitis was the mode of presentation in the fourth case. Encephalocele was the isolated abnormality in three, but the fourth had a degree of associated hypertelorism. The diagnosis of encephalocele should be considered in any patient with a nasal polyp, especially in children and in patients with recurrent bacterial meningitis, with or without rhinorrhea, in the absence of cranial trauma or surgery, or in the absence of external craniospinal anatomical defects.

Key Words: meningitis • encephalocele • cerebrospinal fluid rhinorrhea • nasal glioma

Cases of intranasal encephalocele are uncommon. In 1943, Ingraham and Swan encountered only one intranasal type among 84 cases of encephalocele. Matson, however, reported 14 cases of the intranasal type in a series of 265 encephaloceles. He also commented that these lesions were apt to be mistaken at first for nasal polyps or some other intranasal tumor obstructing the airway. The prognosis is excellent in these cases and, once recognized, every effort should be made to perform surgical repair promptly through the intracranial approach.

We report here four cases of primary intranasal encephalocele, and discuss the problems in the diagnostic evaluation of these lesions.

Case Reports

Case 1

This 10-year-old girl was referred to us in March, 1968, for the evaluation and treatment of cerebrospinal fluid (CSF) rhinorrhea that had persisted for the past 6 months. Rhinorrhea had developed immediately after she had undergone a fourth left nasal polypectomy in 1967. The previous polypectomies had been carried out at the age of 1½, 6, and 8 years. Histological examination of the operative specimen showed cerebral tissue each time, and a diagnosis of nasal glioma had been made.

Physical examination was normal except for widening of the root of the nose. Nasal examination revealed a large polyp situated behind the left middle turbinate, deviation of the septum to the right, a widened left nasal cavity, and leakage of clear fluid from the left nostril. The nasal discharge contained 60 mg% of sugar. Neurological examination was normal. Plain radiographs of the skull and paranasal sinuses (Fig. 1 left) showed a rounded defect, 2 cm in diameter, in the floor of the anterior cranial fossa anteriorly, increased interorbital distance, deviation of the nasal septum to the right, and soft-tissue shadow in the left side of the nose below the skull defect, with destruction of the left ethmoid sinus and medial wall of the left maxillary antrum. Pneumoencephalography (Fig. 1 right) showed downward extension of the left anterior horn, indicating that the nasal mass was an encephalocele.

At left frontal craniotomy and intradural exploration, a large encephalocele was found passing downward into the nose through a rounded defect in the cribriform plate. The encephalocele was excised, and the bone defect was repaired by means of a Teflon
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FIG. 1. Case 1. Left: Plain radiograph of the skull, occipito-meatal view, showing the bone abnormalities and the soft-tissue shadow in the left side of the nose below the skull defect (arrows). Right: Pneumoencephalogram, hanging head, lateral view, showing the downward extension of the left anterior horn (arrow).

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Postoperatively, the patient did well and has had no subsequent rhinorrhea for the past 14 years.

Case 2
This 20-year-old man was referred to us in June, 1980, for the evaluation and treatment of the cause of recurrent pneumococcal meningitis. He had had four previous attacks of meningitis, occurring at the age of 7, 17, 18, and 19 years, which were treated successfully with penicillin but had left him deaf after the first episode. There was no history of trauma, nor was there any history of cranial surgery or CSF rhinorrhea at any time.

General physical and nasal examinations were normal. Neurological examination was normal except for bilateral sensory neural deafness. Plain radiographs of the skull and paranasal sinuses, computerized tomography (CT), and isotope brain scan were normal. Pneumoencephalography was reported to be normal but, during the procedure, there was for the first time leakage of CSF through the right nostril. Isotope cisternography revealed increased radioactive uptake in the right cribriform area, with an increase in radioactivity in the cotton pledgets inserted into the right nostril.

At right frontal craniotomy and intradural exploration, multiple arachnoidal adhesions were found. After separation of the adhesions, a small encephalocele was found extending downward into the nose through a small osseous defect in the cribriform plate on the right side of the midline. The encephalocele was excised, and the bone defect was repaired with muscle and fascia. Postoperatively, the patient did well and had no subsequent meningitis for the past 16 months.

Case 3
This 35-year-old woman was referred to us in February, 1981, for the evaluation and treatment of recurrent episodes of CSF rhinorrhea and bacterial meningitis. She had undergone right nasal polypectomies at the age of 12 and 13 years.Histological examination of the operative specimens showed cerebral tissue each time, and a diagnosis of nasal glioma was made. Since the second polypectomy in 1968, she had suffered from recurrent episodes of CSF rhinorrhea and meningitis. The latter occurred first in December, 1968, and there were three other episodes during 1980; all were treated successfully.

Physical examination was normal. Nasal examination showed no internal mass lesion but revealed a clear watery discharge from the right nostril. The nasal discharge contained 70 mg% of sugar. Neurological examination was normal. Plain radiographs of the skull and paranasal sinuses, CT scan, and serial tomograms through the anterior cranial fossa and base of the skull were interpreted as normal. Isotope cisternography failed to demonstrate the source of CSF leakage, despite active leakage at the time of examination.

Since the clinical evidence favored an anterior fossa origin for the CSF leakage, bifrontal craniotomy with
intradural exploration was carried out. At operation, multiple subarachnoid adhesions were found on the right side. After these adhesions were released, a small encephalocele was found passing downward into the nose through an osseous fissure in the cribriform plate on the right side of the midline. The encephalocele was excised, and the bone defect was repaired with muscle and fascia. Postoperatively, the patient did well, and at follow-up examination 8 months later had had no subsequent rhinorrhea or meningitis.

Case 4

This 18-year-old girl was referred to us in December, 1981, for the evaluation and treatment of recurrent pneumococcal meningitis and persistent CSF rhinorrhea for the past 18 months. She had had two episodes of meningitis, the first in March, 1980, and the second in December, 1981, following a right nasal polypectomy. Both attacks were treated successfully with penicillin. Histological examination of the operative specimen showed cerebral tissue.

General physical examination was normal. Nasal examination showed no internal mass but revealed a clear watery discharge from the right nostril. Nasal discharge contained 75 mg% of sugar. Neurological examination was normal. Plain radiographs of the skull and paranasal sinuses showed opacity of the right maxillary antrum. Tomograms of the anterior fossa and the sphenoid region showed a defect in the floor of the anterior cranial fossa to the right of the midline, with an associated soft-tissue mass extending down into the nose. A CT scan showed the defect but otherwise normal intracranial anatomy.

At right frontal craniotomy and intradural exploration, a large encephalocele was found passing downward into the nose through a rounded 2.5-cm defect to the right of the midline in the cribriform plate. The crista galli was rudimentary and was thinned to a narrow crest. The encephalocele was excised, and the bone defect was repaired by means of a Teflon sheet. The patient did well, and has had no subsequent rhinorrhea 2 months after operation.

Discussion

The term “primary” is used to clarify the causation of a lesion. The primary encephalocele is a congenital lesion occurring early in embryonic development due to failure of mesodermal ingrowth between the neural tube and the overlying ectoderm, which fail to separate.11 The osseous defect is a secondary occurrence and lies in the cribriform plate on one or other side of the midline, where the encephalocele is covered by layers of meninges. This lesion is rare and may be associated with other lesions, in contrast to “secondary” encephaloceles which are quite common, acquired conditions. Secondary encephaloceles may occur at any age at a bone defect in the cribriform plate resulting from cranial trauma or rhinological operations, and from erosion due to raised intracranial pressure. The osseous lesion is the primary occurrence and the encephalocele extends into the nose through the bone defect, at which level the torn dura mater stops. Leakage of CSF and the chance of infection are common in the “secondary” type.

Primary intranasal encephalocele usually appears as a polypoid mass and presents between the middle turbinate and the nasal septum.1,2 The discovery of a nasal polyp in childhood should arouse suspicion, since nasal polyps are most uncommon in this age group.6,7,13 Our Cases 1, 3, and 4 presented in this way. Associated facial and midline brain defects have been described.9 Hypertelorism was the only associated abnormality in Case 1; it was mild and its significance had been overlooked initially. The encephalocele was the only abnormality in three of our cases, suggesting that the condition can be expressed in a variety of ways. The encephalocele in Case 3 was small and remained unrecognized until exploration at craniotomy. Failure to reveal a definite bone defect radiologically or by isotope cisternography was presumably due to the small size of the defect and the presence of multiple postmeningitic arachnoidal adhesions preventing the flow of isotope.

It is interesting to note in our Case 2 that the conventional radiological examinations, including tomography, CT scanning, and pneumoencephalography, were normal. Leakage of CSF occurred for the first time through the right nostril during encephalography. This may have been the result of stretching and separation of some of the arachnoidal adhesions by the air, thus allowing leakage of CSF. Isotope cisternography, however, revealed the site of the leakage. In Case 1, plain radiographs revealed a large bone defect anteriorly in the floor of the anterior cranial fossa, along with a soft-tissue shadow high up in the nose suggesting an encephalocele, which was confirmed by encephalography. In Case 4, tomograms were required in order to demonstrate the defect in the floor of the anterior fossa and the soft-tissue mass in the nose.

Our cases exemplify the problems in the diagnostic evaluation of intranasal encephaloceles.13,14 The presence of a nasal polyp, especially in children, and recurrent bacterial meningitis, with or without CSF rhinorrhea, again in the younger age group, should alert the physician to an occult basal encephalocele. Presence of associated hypertelorism strongly favors a clinical diagnosis, but its absence does not exclude the possibility of encephalocele. Histological examination of the surgically removed polyps is mandatory. Transcranial intradural exploration is the preferred approach;9,13 the neck of the encephalocele is exposed intradurally by retracting the frontal lobe, the extracranial extension is then removed from within, and the dural defect is repaired directly. Redundant meningeal sac protruding into the nose is left alone, and when empty the sac shrinks quickly to an insignificant remnant. These lesions have also been treated
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successfully by an extracranial approach, but the risk of contamination of intracranial contents is great and the brain tissue extending outside the cranium is difficult to free from the margins of the dural defect in order to close the dura securely.

Matson's observations concerning brain herniation through a defect in the cribiform plate, the clinical presentation, the operative approach, and the excellent prognosis in his patients with this condition, correlate well with our experience. The presentation and operative findings in our cases were also similar to those reported by Smith, et al., in 1963. These authors named these lesions "nasal gliomas," but stated that they had no true neoplastic characteristics. They considered them to be developmental anomalies with the same pathogenesis as nasofrontal encephaloceles. We prefer the term "primary intranasal encephalocele" to clarify the nature of the origin.

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