CASE REPORT

Encephalcoele- A Cause of Recurrent Meningitis

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Abstract
There are many causes of recurrent meningitis of which anatomical defects such as cephalocele are common. We report a case of a 45 year-old woman who presented with recurrent meningitis and was found to have an underlying naso-ethmoid encephalocele .

Key Words
Naso-ethmoid Encephalocele, Recurrent, Meningitis

Introduction

Recurrent meningitis is defined as two or more discrete episodes of meningitis with normal CSF parameters of inflammation in between the episodes.There are many causes of recurrent meningitis of which anatomical defects such as cephalocele are common. (1)

The meningocele and encephalocele are extracranial herniation of only meninges and meninges with brain tissue respectively through cranial defects. The pathology is usually classified according to its content and location. The most frequent site of an encephalocele is the posterior skull but it may be found in the fronto-orbital region or as a herniation located between the orbits. Nasal encephaloceles are herniations of the intracranial contents through a defect in the anterior skull base.

Case Report

We report a case of a 45 year-old woman who presented with recurrent meningitis and was found to have an underlying naso-ethmoid encephalocele .

Case Report

A 45 year old women presented to the medical emergency with history of high grade fever and altered sensorium for one day. There was no history of any rash over the body, seizures or head trauma. The patient's relatives gave a history of two similar episodes in the past which were diagnosed as bacterial meningitis and that each time patient improved on intravenous antibiotic therapy. There was a history of intermittent episodes of watery discharge from the nose in the past which used to increase on bending the head but these episodes were self limiting and no treatment was ever sought for that.

On examination the patient was aggressive and disoriented in time place and person. The oral cavity examination revealed no fungal involvement. There were no abscess, eczema, warts or rash on the skin. There were no significant findings on systemic examination. CNS examination revealed presence of neck rigidity. Considering the history and examination a provisional diagnosis of acute meningitis was considered.

On investigation her hemoglobin was 11.6gm/dl, TLC - 5600, DLC-P70L36E2M2, Urea-24 mg/dl, creatinine-0.8 mg/dl, S. Bil - 0.5 mg/dl, SGPT- 12 IU, ALP-7 KA, SGOT- 10IU, blood sugar - 90 mg/dl, Na-134meq/l, K-4.9meq/l and urine examination was normal. Chest X-ray was normal. Lumbar puncture was performed and CSF analysis revealed: protein-230 mg%, sugar- 30 mg%, TLC -850 cells/mm3, DLC-P89L11. This was consistent with the diagnosis of acute bacterial meningitis. Patient was started on empirical antibiotic therapy, injection ceftriaxone 2 gm 12 hourly which was later continued after the results of CSF culture and sensitivity were obtained. In view of significant past history of CSF rhinorrhea and similar two prior episodes of meningitis an MRI brain was done. It revealed the herniation of dysplastic brain parenchyma via a defect in the cribriform plate into the region of the ethmoidal air cells and nasal cavity suggestive of naso ethmoidal encephalocele

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Fig.1 Showing the Herniation of Dysplastic Brain Parenchyma via the Defect in the Cribriform Plate into the Region of the Ethmoidal air cells and Nasal Cavity suggestive of Naso Ethmoidal Encephalocele

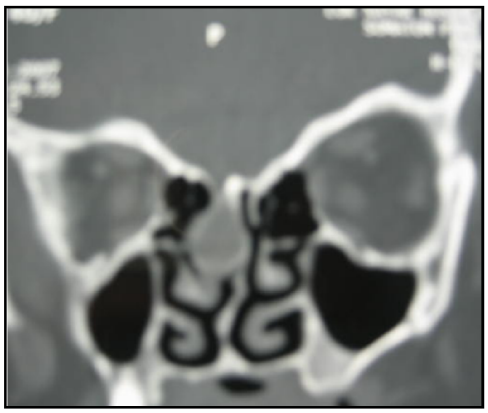

Table. 1 Showing Causes of Recurrent Meningitis

Causes of Recurrent Bacterial Meningitis
Immunodeficiency
a) Congenital
– B cell defect
- Combined B and T cell defect
- Compliment deficiency
- Aspleenia
b) Acquired
- HIV infection
- Aspleenia
CNS Abnormality
a) Congenital
- Intracranial and intraspinal abnormalities with or without dermal connection
- Skull base defect
- Inner ear defect
b) **Acquired**
- Skull fracture
- Shunt surgery
- Instrumentation

(*Fig 1 & 2*). With the appropriate antibiotic and symptomatic treatment patient improved. Patient was referred to neurosurgery department for the repair of the defect.

Discussion

Bacterial meningitis is a relatively common disease entity however recurrent meningitis is relatively uncommon. (1) Kline reviewed the world literature from 1978-1988 and found 47 patients with recurrent meningitis. (2) In his series of 47 patients, the various predisposing factors found were congenital CSF fistula in 55% of cases, traumatic or surgical CSF fistula in 17%,

Fig.2 Showing the Herniation of Dysplastic Brain Parenchyma (arrow marked) via the Defect in the Cribriform Plate into the region of the Ethmoidal air cells and Nasal Cavity suggestive of Naso Ethmoidal Encephalocele

immunodeficiency in 21% and unknown cause in 6%. Therefore every case of recurrent meningitis merits for a search of an underlying abnormality. (2) The abnormality may be anatomical, immunological or a combination of the two. The various causes of recurrent meningitis are summarised in *table 1*.

Recurrent meningitis could be due to immunodeficiency or due to structural defects. Immunological problems in a patient are suggested by history of recurrent episodes of infections at multiple sites. A history of chemotherapy for cancers, immunosuppressive drugs after transplantation, HIV infection suggests immunodeficiency. Appropriate screening tests for immunodeficiency includes quantitative determination of immunoglobulin levels, assessment of immunoglobulin function and estimation of compliment levels. If there is suspicion of infection by staphylococcus as the cause of recurrent meningitis than chemotactic and phagocytic function of leukocytes should also be investigated.

Anatomical abnormalities are those in which an abnormal connection exists between the central nervous system and mucosal surface or the skin2. These can be acquired or congenital. Acquired defects can be either traumatic i.e. anterior skull base fractures or iatrogenic such as following endoscopic sinus surgery. Congenital defects between the cranium and duramater results in herniation of intracranial contents and are referred to as cephalocele. If the herniation contains only meninges it is a meningocele and when both brain and meninges herniated it is called as a meningoencephalocele.

Nasal encephalocele represents a herniation of brain substance into the nasal cavity through a congenital defect in the skull just anterior to the crista galli. This may extend extranasally through the frontonasal suture. If such an encephalocele becomes pinched off and gets isolated from the cranial cavity it is termed as nasal glioma. Nasal encephaloceles are very rare (1:5000) (3) and can be divided into two main groups: frontoethmoidal and basal encephaloceles. Among the frontoethmoidal naso-ethmoidal is the commonest type. Basal encephaloceles are classified as transethmoidal, sphenoethmoidal, transsphenoidal and frontosphenoidal. (4)

In naso-ethmoidal encephlocele the defects lie anteriorly either along the midline or along the cribriform plate and do not involve the sella turcica. The hernial sac extends inferiorly into the sinuses or nasal cavity and typically contains portions of frontal lobes and olfactory apparatus. Naso-ethmoidal encephaloceles usually presents with recurrent episodes of meningitis, intermittent CSF rhinorrhea or sometimes as a nasal mass.

The patient can also presents with unilateral nasal obstruction, chronic nasal discharge and headache after forceful blowing of nose. These can be misdiagnosed as nasal polyps and can be potentially fatal after erroneous polypectomy. (5) Imaging studies should be done in cases of polypoidal nasal mass prior to biopsy. These encephaloceles can be also be associated with other congenital anomalies of midrace, orbits and hydrocephalus. (6)

One of the common manifestation of the naso-ethmoidal encephaloceles is CSF rhinorrhea which can be easily diagnosed by doing an invasive test called the radioisotope cisternography. In this test, an injection of a radioactive tracer is given into the lumbar CSF and some wool pledgets are placed in the roof of both nasal cavities. The anatomic location of leak can be found out by measuring the radioactivity in the pledgets. Immunological tests performed on the nasal fluid like the presence of a positive 2-transferrrin band can also be helpful in diagnosing CSF rhinorrhea. (7)

However with the advent of multislice spiral CT a non-contrast study can non-invasively display the osseous defect beautifully in cases of CSF rhinorrhea. MRI has a superior soft tissue resolution. It can define the nature of the encephalocele and allows an accurate depiction of the olfactory and optic tracts, hypothalamic-pituitary system and agenesis of corpus callosum also. Thus imaging studies such as CT and MRI should be obtained in patients of recurrent meningitis with CSF rhinorrhea.7

Treatment of encephalocele is mainly surgical via a transcranial approach. It is associated with complications like loss of sense of smell, post-operative intracerebral hemorrhage, cerebral edema, epilepsy, frontal lobe dysfunction with memory and concentration deficits. The advent of endoscopic sinus surgery has allowed a new intranasal approach for the treatment of encephaloceles minimizing patient morbidity. (8)

Congenital encephalocele causing recurrent meningitis are usually seen in children. It presenting as a cause of recurrent meningitis at 45 years of age is very unusual. As for the cause of recurrent meningitis in adults, very few case reports are published and are mainly post traumatic in nature.

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