Diagnosis nearly missed-Recurrent Meningitis

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Abstract:
Recurrent meningitis poses a diagnostic dilemma in which helplessness of the treating physician goes a long way and continues to haunt him until the “bulls eye” is hit. Here we present a case of a young girl with frequent episodes of meningitis in which conventional neuroimaging done each time missed the diagnosis of frontal meningoencephalocele until a very high index of clinical suspicion of a possible anatomical bony defect provoked the enthusiasm of the neuroradiologist for a reconstructed CT imaging thus confirming the diagnosis.

Keywords: Recurrent meningitis, Meningoencephalocele.

Introduction:
Recurrent meningitis is most frequently associated with anatomical defects in the skull base or spine due to genetic defect in the complement system. Recurrent bacterial meningitis is a rare phenomenon and generally poses a considerable diagnostic challenge to the clinician. An intranasal meningoencephalocele or encephalocele consists of a discontinuity of the ethmoidal cribiform plate with protrusion of the meninges and brain tissue into the nose[1]. It may be caused by congenital anomalies, by post traumatic fracture of the ethmoidal cribiform plate, by rhinological operations, or by bone erosion due to raised intracranial pressure. Cerebrospinal fluid rhinorrhoea and recurrent bacterial meningitis can complicate all types of meningoencephalocele[1,2]

Case report:
16 years old girl presented with three recurrent episodes of headache, altered sensorium, high grade fever, nuchal rigidity and vomiting with the interval of 4-5 months. No history of trauma, seizures and any previous intracranial surgery. She had a full term normal delivery.

Examination: She was febrile (T-101 F), pulse rate-110/min regular, blood pressure-100/60mmHg, systemic examination: CVS, RS, PA was normal, CNS-she was drowsy, responding to pain stimuli, neck stiffness was present, Kernigs and Brudzinski signs were positive, pupils were normally reacting to light, fundus examination was within normal limit, no cranial nerve involvement, no motor or sensory deficiency, no sphincter involvement, no rhinorrhoea.

Investigation: immediate computed tomographic(CT) scan of Brain was normal. Haemogram suggestive of leukocytosis(22,000) predominantly polymorphs, Hb-12.5 mg/dl and peripheral blood smear was normal, ESR-30. Cerebro spinal fluid(CSF) picture revealed findings of pyogenic meningitis and CSF cultures suggestive of Streptococcus Pneumoniae. MRI Brain showed leptomeningitis. On second admission with similar complaints CSF showed viral meningitis with normal cultures. Repeated CT Brain was normal.
except meningeal enhancement. On the third admission CT scan of Brain with 3D reconstruction revealed frontal meningoencephalocele due to defect in right cribriform plate. (figure 1).

Treatment: initially patient was treated as pyogenic meningitis and subsequently treated as chronic pyogenic meningitis. Neurosurgery opinion was taken for meningoencephalocele and suggested an endoscopic transnasal approach for the closure of the defect. The defect was closed with the help of surgical glue (1ml) and fascialata incised from the thigh of the same patient.

Discussion

A single episode of bacterial meningitis is often the result of blood borne bacteria. A second episode of meningitis is considered a recurrence if resulting from a different pathogen than the first, or if it occurs from same organism but after 3 weeks of completion of therapy after the initial episode. Bacterial meningitis is a severe potentially life threatening infection associated with high rates of morbidity and significant disability in survivors. Potential long term neurological sequelae include cranial nerve palsies, hemiparesis, hydrocephalus, seizures as well as hearing and visual impairment which can have profound impact on quality of life of survivors.

A large retrospective study by Durand et al, established that as many as 6% patients presenting with community acquired meningitis develop a subsequent recurrence. Streptococcus pneumoniae was the most common isolated organism, consisting of 56.6% of culture confirmed cases of recurrent meningitis. This was followed by Neisseria meningitides, Haemophilus influenza, E.coli, Staph aureus.

It is difficult to detect the underlying aetiologies for the patients with high risk of recurrence when the bacterial meningitis occurs for the first time unless there is history of skull base injury or the presence CSF leakage. Of the 363 cases of recurrent meningitis in various studies grouped by Nigel Curtis et al, 214 (59%) were related to anatomical defects, 132 (36%) to immunodeficiencies, and 17 (5%) were related to parameningeal infections.

In the series of Lieb et al. the cause of recurrent meningitis in 25 children, was an anatomical lesion with 13 intracranial defects including encephaloceles, skull fractures, Mondindysplasias, neuroenteric cyst, fibrous dysplasia, persistent craniofaryngeal defect. In 24 out of 25 cases the final treatment was surgical intervention. Meningoencephalocele is a herniation of meninges, CSF and brain parenchyma from the skull. It is a rare disease that occurs in 1:6000 to 35,000 newborns. Most cases are congenital but some could be acquired by skull injury. Fronto-ethmoidalencephaloceles are most common type followed by nasopharyngeal and orbital types.

A radiological anatomic study of the cribriform plate compared with constant structures shows that frontal bone is thick and dense in the ethmoid roof area. But medially, the transition from thick bony part of frontal bone to thinner lamellae of ethmoid is observed. The intranasal post traumatic meningoencephalocele is caused by a discontinuity of the ethmoidal cribriform plate as a consequence of fracture at the base of the skull with herniation of the dura, arachnoid and brain. Recurrent meningitis usually develops within 3 months. Encephaloceles are usually congenital, and have been less commonly described as being acquired caused by rhinological operations or traumatic head injuries. Basal encephaloceles may protrude through cribriform plate into superior meatus (transethmoidal), posterior
ethmoid and sphenoid sinuses (sphenoethmoidal) or through a patent craniopharyngeal canal (transsphenoidal) into nasal and pharyngeal spaces\textsuperscript{[i]}

In conditions with abnormal CSF connection in cranial location like head injury, basal skull fractures, congenital basal skull defects, meningoceles/ meningoencephaloceles and inner ear abnormalities), S. pneumoniae was the most frequently isolated organism, accounting for 179 (72\%) of the 247 culture positive episodes of recurrent meningitis\textsuperscript{[6]} in our patient Streptococcus pneumoniae was isolated. History should include that of head injury, otorrhea, rhinorrhea, persistent nasal discharge, anosmia or hyposmia. Our patient presented with headache only.

Drummand et al, proposed that children with recurrent meningitis of unknown etiology should undergo audiological evaluation, CT scan of skull bone and paranasal sinuses\textsuperscript{[12]} CT scans in general and high resolution HRCT in particular produce best definition of bony structures and are considered to be method of choice for investigation of basal skull defects by many authors. Other radiological techniques that can be used to identify include CT cisternography, radionuclide cisternography, and cranial MRI scan\textsuperscript{[13]} In this case, 3D reconstructed CT scan was done to see the defect.

Defects in cribiform plate can be sealed with autologous material such as abdominal fat, nasal septum mucosa, bone, fascia lata and muscle such as our case where fascia lata from thigh was taken. The graft can be attached with fibrin glue or Vaseline gauzes where the fibrin glue simulates thrombin and fibrinogen in the coagulation cascade.\textsuperscript{[14]}

Transnasal endoscopic repair is a challenging procedure. Surgical management now relies on an extracranial approach where the success rate is 90\%, when compared to intracranial approaches where the success rate is generally accepted as being 67 to 73\% after first procedure and upto 90\% in subsequent multiple procedures. Taking into consideration the age of our patient, a transnasal repair of defect was an important decision we chose to make. Our patient is now symptom free with no further episodes of recurrent meningitis.
Figure Legends:

Photograph 1: CT with 3D reconstruction showing defect in right cribriform plate.

Photograph 2: Intra operative repair of the defect (Frontal Meningoencephalocele).
References