Nasal Encephaloceles presenting at later ages: Experience of Otorhinolaryngology Department at a tertiary care center in Karachi, Pakistan

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Abstract

Encephaloceles are anomalous herniations of the meninges, with or without brain matter. Globally the incidence of encephalocele is about 1 per 35,000 births, but it is more frequently reported in Southeast Asia. As the defect is more pertinent to embryological development, an encephalocele is a more common entity in an infant with a mean age of presentation ranging between 15.5 and 21 months; making an encephalocele presenting for the first time in a relatively older individual a rare occurrence. Consequently a surgeon might not consider an encephalocele among his differentials.

Here we present a series of encephaloceles that presented at a later than usual age as nasal masses to the otorhinolaryngology department of our hospital, and recommend that the differential of encephalocele be entertained for nasal masses as proceeding with routine procedures may result in potentially lethal complications.

Keywords: Encephalocele, Nasal encephalocele, Nasal masses, Meningioencephalocele.

Introduction

Encephaloceles are anomalous herniations of the meninges, or brainmatter and meninges, beyond the boundaries of the cranium; referred to as meningocele and meningoencephalocele respectively. Globally the incidence of encephalocele is about 1 per 35,000 births, but it is more frequently reported in Southeast Asia; nearly 6 times more at 1 every 6,000 births.[1] The etiology is vague occurring perhaps due to failure of the fonticulusfrontalis to close properly thus culminating in a herniation. Hereditary aspect have been implicated in its pathogenesis as episodically encephaloceles have been observed in families whose members had developmental anomalies of central nervous system (CNS).[2]

As the defect is more pertinent to embryological development, an encephalocele is a much more common entity in an infant and may be considered as a differential diagnosis for any mass related to nose, maxilla and forehead. With a mean age of presentation ranging between 15.5 and 21 months;[3] an encephalocele presenting for the first time in a relatively older individual is a very rare occurrence. Consequently a surgeon might not consider an encephalocele among his differentials and proceed to carry out a diagnostic procedure such as fine needle aspiration (FNA), which could potentially be disastrous; leading to neurological deficits and intracranial infections including meningitis.

Here we present a series of encephaloceles that presented at a later than usual age as nasal masses to the otorhinolaryngology department of our hospital, demonstrating the need to utilize radiological imaging techniques prior to other diagnostic procedures including FNA.

Case-1:

An 11-year-old girl was reffered to our clinic from rural area, with an expansile growth over her nasal bridge present since birth, which was associated with intermittent watery discharge from her right eye. She complained of shortness of breath on exertion; however, there was no difficulty in breathing on moderate activity.

On examination there was a 4x5 cm cystic swelling associated with telecanthus. Her nasal cavities were adequate in size and there was no associated anosmia or discharge.

Magnetic Resonance Imaging (MRI) showed a mixed signal intensity lesion at the nasal bridge measuring about 3.3 x 3.5 cm, with an intact underlying nasal bone architecture; indicating a nasal encephalocele.

She underwent excision of lesion. Intraoperatively a well demarcated cystic lesion of about 3 x 3 cm was noted with loss of the nasal bone inferior to it. The lesion was attached to intercranial structures and its stalk was appreciated, passing through a defect in the left cribriform plate. The lesion was mobilized, ligated and excised. Furthermore, an augmentation rhinoplasty was done in order to improve cosmetic outcome. Post-operatively, patient had no complications

Case-2:

A 50-year-old female, known case of hypertension, diabetes and deviated nasal septum, was referred to our clinic with complains of cerebrospinal fluid (CSF) rhinorroehoea for 6-7 years from her right nostril. This was associated with fever, headache and neck pain for the last 10 days. She had a history of severe pneumococcal meningitis

progressing to coma 4 years ago.

MRI showed post contrast enhancement of her meninges indicating meningitis. Fluid density signals were also noted in her sphenoid sinuses. Computed Tomography (CT) scan showed that her sella was enlarged with a soft tissue density area within it. Her sphenoid sinuses were bilaterally enlarged and a defect was noted in the right cribriform plate. CT cysterogram showed the middle part of right cribriform plate to be deficient with thinning of the middle part of bony cribriform plate on the left. Active secretion of contrast through the defect in the right cribriform plate defect into the middle ethmoidal air cells was noticed with a small amount of contrast pooling in right nasal cavity. No active secretion was noted on the left side.

The defect was repaired using a combined endoscopic and transcranial approach. Per-operatively a meningiocele was noted medial to the anterior attachment of the middle turbinate which was excised and pushed into cranium. Bone flap was elevated, both frontal sinuses were opened and internalized. A large defect was also noted in the posterior part of cribriform plate. This defect was closed using a peritoneal flap. No complications occurred post-operatively.

Case-3:

A 10-year-old female presented to our clinic with history of fall after which she developed intermittent high grade fever and CSF rhinorrhoea. She had approximately 5 episodes of CSF rhinnorhoea in last 5 months, which were associated with recurrent attacks of fever. She had undergone a previous crainiotomy with reconstruction at another centre, however this failed to resolve her symptoms.

On examination, she was alert and following commands. Anterior rhinoscopy revealed sodden nasal mucosa on the left side. No swelling or abnormality was detected.

CT scan revealed a defect in the left cribriform plate associated with a CSF link in the roof of the left nasal cavity. MRI showed similar findings.

She was initially planned for an endoscopic transnasal approach with the help of a neuronavigation system, to repair the CSF leak. Intraoperatively brain matter was seen herniating through a defect in left cribriform plate. The initial attempt at transnasal endoscopic repair failed and a transcranial approach was employed. The defect was identified intracranially and covered with temporalis fascia, fibronectin glue and a bonegraft. No complications occurred post-operatively and patient remained stable.

Case-4:

A 12-year-old female presented to our clinic with complaint of a swelling over her nose since few years. The swelling was initially about the size of a lemon but progressively increased in size as she grew. On examination a red, mobile, soft and non tender mass of about the size of a tennis ball was noted (Figure-1 A,B). The swelling was not associated with any discharge.

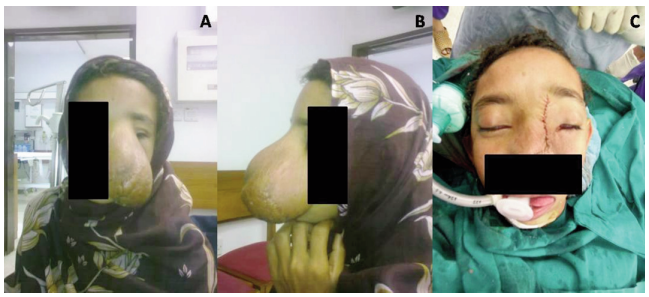

Figure-1: Frontal (A) and lateral (B) views of a mobile, soft and non tender mass of about the size of a tennis ball noted on physical examination. (C) Patient seen immediately after the completion of surgery.

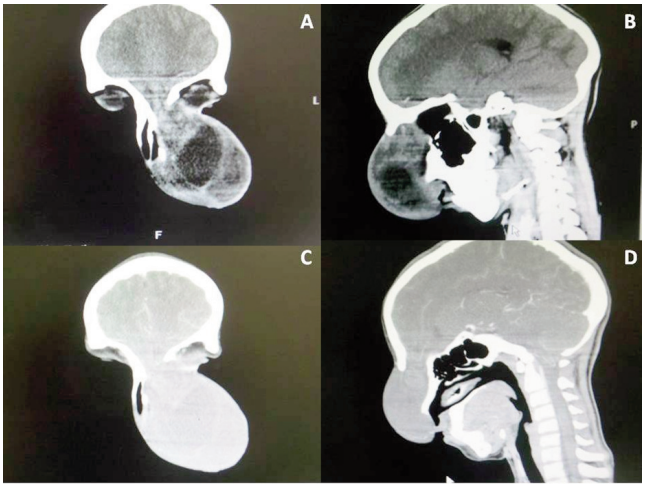

Figure-2: The lesion in patient 4 along with a large bony defect in the left fronto-ethmoidal region as seen on (A), (B)CT scan and (C), (D) MRI.

CT scan showed a large bony defect in the left fronto-ethmoidal region. A heterogeneous soft tissue density lesion containing meninges and CSF was seen in continuity with the defect. The left superior frontal gyrus was also seen extending into the lesion. There was mild traction on the left frontal horn and pressure over the left lobe of the brain representing a left sided fronto-nasal encephalocele (Figure-2 A, B).

MRI also showed a similar bony defect with herniation of the brain parenchyma along with meninges and CSF. Asymmetry of the frontal horns was noted with the left horn appearing stretched and the genu of the corpus callosum to be deficient (Figure-2 C, D).

She was planned for surgical excision involving a multidisciplinary team including ENT and Neurosurgical specialists. Intra operatively a huge pedicled cystic lesion of

about 5x6 cm was noted exiting from the left side of the cribriform plate in close proximity to the orbital contents. The nasal septum appeared to be absent and left lateral nasal wall was flattened and pushed to the right side.

The pedicle was divided and lesion excised. Brain matter remnants were pushed back and closed using fibroblast glue and remaining mucosa around the lateral wall.

Nose architecture was reconstructed using poly methylmethacrylate. She was vitally stable post procedure but she developed a collection at the wound site. A Penrose drain was applied and the expulsion appeared to be CSF. The involved neurosurgical team was consulted and a lumbar drain was placed. The swelling decreased in size and her lumbar drain was subsequently removed. She was then discharged.

On one week follow up in the ENT clinic she complained of pillow wetting. On examination CSF rhinorrhea was noticed and she was admitted under the same multidisciplinary team. On rigid endoscopy the site of CSF leak could not be identified and a lumbar drain was again placed. On removing the lumbar drain her CSF rhinorrhea redeveloped and she underwent intracranial repair of CSF leak. Her postoperative recovery was uneventful and she was discharged.

Discussion

Encephaloceles can be grouped according to their anatomical location into occipital, sincipital and basal. Herniation through the cribriform plate and fovea ethmoidalis are a few of basal variants.[4] Similarly, they can also be classified into anterior and posterior encephaloceles. Some of the anterior encephaloceles include frontoethmoidal, nasofrontal, nasoethmoidal, nasoorbital.[1] The frontonasal type of frontoethmoidal encephalocele is the subtype that occurs more frequently.[5]

Patients with nasal encephaloceles typically present with a soft compressible mass that transilluminates. However various other nasal masses take precedence in the list of differentials over an encephalocele when patients present at a later age with initial complaints of a nasal mass. Physical examination of the swelling is vital for accurate diagnosis since multiple anomalies simulate an encephalocele. A characteristic test is a positive Furstenberg test i.e. enlargement of the mass with bilateral compression of internal jugular veins. The mass also enlarges on crying or valsalva manoeuvre and may be associated with CSF leak.

Imaging modalities are imperative to the diagnosis of an encephalocele along with providing assistance in determining the extent of the lesion, the type of herniation and any other concomitant deformity that might be missed on physical examination.[6] Computed tomography (CT) scan can be utilized, along with magnetic resonance imaging (MRI) which complements the CT findings and is even superior to it by making vessels discernable and thus illustrating intracranial communication in majority of cases. Lately, CT Cysterogram has also been utilized in diagnosing encephaloceles.

Turgut et al reported mortality of 46% when there is brain tissue in the encephalocele,[7] thus demonstrating that prompt repair is crucial. Repair of fronto-nasal encephalocele, in toddlers, requires a simpler operative technique even for larger and more complex lesions.[8] In comparison to children, fronto-nasal encephaloceles in adults, present a complicated situation for the surgeon,since advancement in age enlarges the size of the defect, more gliotic brain tissue will be present in the herniation, along with an increase in size of the paranasal sinuses,[9] thick calvarial bones, large bony defect and sometimes scars from previous surgeries.[9]

A few large series have been published pertaining to adult encephalocele,[10] making it a rare presentation; however the authors recommend that the differential of encephalocele be entertained for nasal masses as proceeding with routine procedures may result in potentially lethal complications.

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