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Case Report

Paediatrics Section

Recurrent Meningitis with Upper Airway Obstruction in A Child: Frontonasal Encephalocele- A Case Report

SOUMYA SACHDEVA[1], ROHIT KAPOOR[2], PREMILA PAUL[3], RAKESH YADAV[4]

ABSTRACT

Nasal encephalocele are rare congenital anomalies; these benign masses may be confused with nasal dermoids, hemangiomas, nasal gliomas and anterior skull base masses. These lesions have concomitant defects in the anterior cranial fossa thus this potential communication can cause recurrent episodes of meningitis and/or difficulty in breathing and cosmetic anomalies. We bring a case of a 6-year-old child who presented to the clinic with multiple episodes of meningitis which was associated with nasal discharge. The imaging studies and nasal fluid analysis confirmed it as cerebrospinal fluid; subsequently imaging findings concluded it as frontonasal encephalocele which was later resected and patient showed improvement.

Keywords: Coloboma, Frontonasal encephalocele, Recurrent meningitis

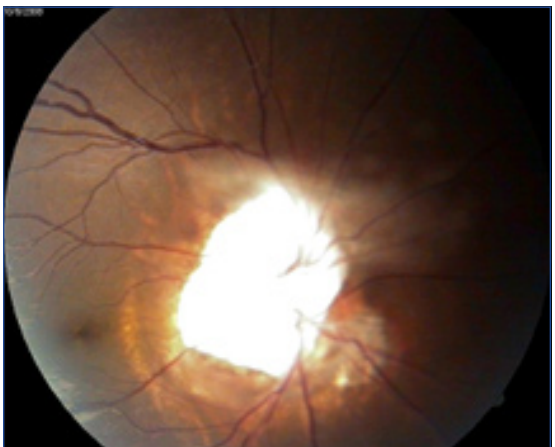
CASE REPORT

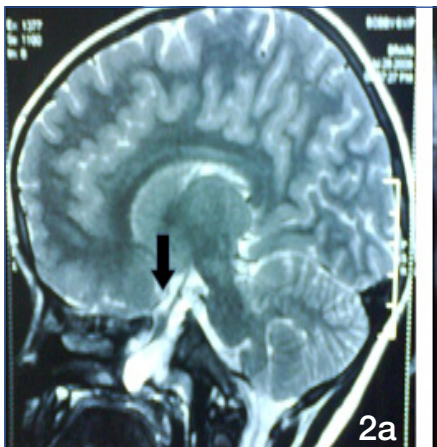
A 6-year-old child presented with fever since 15 days; which was high grade, intermittent, not associated with chills and rigors. The child experienced nasal discharge; which was episodic, watery, odorless, increased by bouts of coughing and vomiting since two days. Headache was throbbing in nature, intermittent and bifrontal. There was associated vomiting; which was non bilious and projectile. On detailed history; multiple episodes of meningitis were elicited. On examination; it was found that the child was febrile, with scar of operated midline cleft lip. ENT examination revealed mass in the upper part of nasopharynx. Central nervous system examination revealed signs of meningitis and fundal examination showed right fundal coloboma [Table/Fig-1]. Rest of the systemic examination was unrevealing. The investigations demonstrated moderate anaemia and increased leukocyte count and the peripheral smear showed neutrophilic leukocytosis. Cerebrospinal fluid examination revealed 50 cells; majority of which were polymorphs and with raised protein of 160mg%. However culture and gram stain was negative. The nasal fluid examination was positive for ß2 transferrin. MRI of the brain showed herniation of larger CSF filled sac into nasopharynx through wide defect measuring 1.1cm in sphenoid body ,sac also contained stretched optic nerves and part of inferior hypothalamus and gliotic septae. The sac measured 2.8 x 1.5 x 2.5 cm in craniocaudal, transverse and anterior dimension ;finding suggestive of nasopharyngeal encephalocele [Table/Fig-2a,b]. Hypertrophy of left maxillary sinus was appreciated as well. The nasopharyngeal encephalocele was subsequently excised and the child showed improvement.

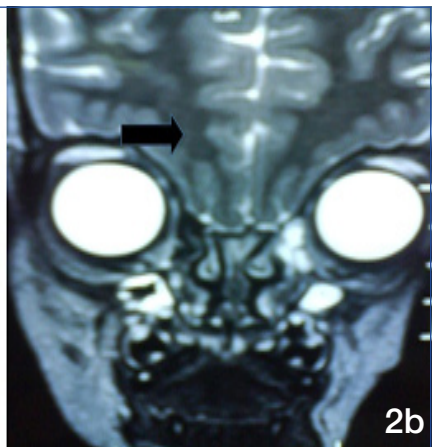
DISCUSSION

According to literature, 1 in 20,000 to 40,000 live births [1] are diagnosed with congenital nasal masses. These lesions present in neonates [2] ; as upper airway obstruction with bony skull defects; with/ without herniation of neuroglial tissue or choroid plexus [3].

Encephaloecle is diagnosed in 1 of every 4000 live births; with an equal incidence in both males and females. The first case can be dated back to the 16th century [4]. Occipital encephalocele is the most common type (75%); followed by Frontal encephalocele (25%). Among the Frontal encephalocele; there are two categories; namely the sincipital (60%) and the basal (40%) type. Transethmoidal, sphenoethmoidal,transsphenoidal,sphenomaxillary,nasopharyngeal and pterygopalatine fossa type are the types of basal encephalocele







[Table/Fig-1]: Optic Nerve dysplasia and right fundal coloboma. **[Table/Fig-2a]:** Herniation of large CSF filled sac into nasopharynx through wide defect measuring 1.1 cm in sphenoid body,sac also contains stretched optic nerves,part of inferior hypothalamus and gliotic septae.(arrow) **[Table/Fig-2b]** Sac measures 2.8 x 1.5 x 2.5 cm in craniocaudal, transverse and anteroposterior dimesnsion suggestive of nasopharyngeal encephalocoele(arrow); hypertrophy of left maxillary sinus can also be appreciated

and present as intranasal masses. The external nasal masses are the sincipital type and based on their location; are divided into nasofrontal, nasoethmoidal, and naso-orbital [5].

Nasal gliomas, encephaloceles, and dermoids result from abnormal embryological development [6,7]. The literature suggests many mechanisms of development of nasal glioma and encephalocele. The most widely held view is the Encephalocele Theory, which suggests that these lesions develop as a protrusion of forebrain secondary to faulty closure of the anterior neuropore [8].

Our case presented as multiple episodes of meningitis and nasal discharge; diagnosed as Nasopharyngeal Encephalocele. These lesions can mimic a variety of different problems causing nasal obstruction like common cold,sinusitis and allergic rhinitis in pediatric population [9]. The basal encephalocele and morning glory syndrome should always be kept as a possibility in cases of midline defects; when ophthalmological finding of strabismus and poor visison are present [10]. Some rare cases of extramedullary hematopoiesis have also been reported as frontoethmoidal enchephalocele [11] and a case of buccal angiomatosis has been reported in transalar sphenoidal meningoenchephalocele [12]. Cryptophthalmos syndrome has been described in the literature along with the basal enchephaloceles [13]. Pitutary and hypothalamic dysfuction has been reported by different authors in basal enchephalocele [4]; which was absent in our case. The common lesions with which nasopharyngeal encephalocele is confused include dermoid, hemangioma and glioma [4]. In a patient with optic nerve anomalies like megalopapilla and optic nerve dysplasia; possibility of basal enchephalocele should be kept [13] in mind. However, our patient presented with optic nerve coloboma, as has also been seen in various studies [13]. These cases must be treated early in order to prevent cerebrospinal fluid leak and meningitis.

The main modality of management of enchephalocele is surgery. It is recommended that the earliest treatment be instituted to reduce cosmetic deformity and chances of infection. The procedure adopted is a frontal craniotomy with resection of intracranial component with the repair of dural covering and the skull base. The size of the nasal mass determines whether surgeon has to adopt extracranial approach. In extracranial-extranasal approach; the bicoronal incision can be extended by pulling the bicoronal flap till the bony cartilage junction and further the intracranial extension can be determined. For the excision of the extracranial intranasal enchephalocele; transnasal endoscopic approach is made use of, which has good results.

CONCLUSION

In a patient presenting with recurrent episodes of meningitis , nasal discharge, upper airway obstruction, midline oral cleft anomalies and optic nerve anomalies; the physician must be prompted to look for enchephalocele.

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PARTICULARS OF CONTRIBUTORS:
1. Intern, Department of Pediatrics, Vardhman Mahavir Medical College and Safdarjang Hospital, New Delhi, India.
2. Medical Officer, Department of Pediatrics, Government of India.
3. Ex Associate Professor, Department of Pediatrics, Vardhman Mahavir Medical College and Safdarjang Hospital, New Delhi, India.
4. Medical Officer, Department of Pediatrics, GDMOII,Government of India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Soumya Sachdeva,
108 Model Town, Janta Sewak Hospital, 8 Marla, Gurgaon, Haryana-122001, India.
Phone: +917838159168, E-mail: soumyasachd@gmail.com

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