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RESEARCH ARTICLE

FRONTONASAL EPIDERMOID CYST WITH PATENT DERMAL SINUS TRACT OPENING ON THE DORSUM OF NOSE AND INTRACRANIAL EXTENSION THROUGH CRIBRIFORM PLATE DEFECT: A RARE CASE REPORT WITH REVIEW OF LITERATURE

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Abstract

Congenital midface anomalies are rare. Multiple congenital midface anomalies occur in children. Imaging techniques like computed tomography (CT) and magnetic resonance imaging (MRI) help in characterising the lesions, making definite diagnosis and knowing about intracranial extension. We present a case report of 8 year old female child with Frontonasal Epidermoid Cyst with patent dermal sinus tract opening on the dorsum of nose and intracranial extension through cribriform plate defect.

Introduction:-

Congenital midline nasal anomalies are rare with incidence of one in 20000-40000 live births[1,2].Frontonasal Epidermoid with patent dermal sinus tract opening on the dorsum of nose is a rare anomaly and is one of the multiple congenital midline nasal anomalies, other anomalies include dermoid cysts, encephaloceles, gliomas, hemangiomas, teratomas, neurofibromas, lipomas and lymphangiomas. The most commom of all these are dermoid, or epidermoid cyst, encephalocele and glioma[3]. They arise because of failure of normal embryological development. Thorough understanding of the development of this region helps in differention and characterisation of these lesions. CT and MRI plays an important role in characterising the lesions, making definite diagnosis, knowing about extent of lesion and any intracranial component. CT is best for evaluating osseous remodelling , erosions whereas MRI helps in detailed characterisation of the soft tissue and to know extent of lesion and intracranial component so that most appropriate therapeutic intervention can be planned.

Case :

A 8 year old female child presented with history of dorsal nasal pit since birth with intermittent , scanty, serous discharge from this pit for the past 4 years. There was history of broadening of the dorsum of nose. On examination there was broadening of the nasal bridge and dorsum of nose. There was a dorsal nasal pit which was dry and showed dry crusts over it and no secretions at the time of examination. CT brain and frontonasal region was done which showed well defined lesion with CT attenuation upto 15 HU in frontonasal region in midline and towards left side with bone window sections showing defect in left cribriform plate, however definite intracranial extension of the lesion could not be established confidently, so MRI was done for characterisation of lesion, to know about intracranial extension and sinus tract communicating with dorsal nasal pit. MRI showed well circumscribed, encapsulated, fluid signal intensity lesion, showing diffusion restriction on DWI seen in fronto-nasal region in the midline. The lesion showed no signal suppression on T2 fat saturated images. There was a contiguous sinus tract extending to nasal bridge with minimal intracranial extension along the floor of right anterior cranial fossa through a focal bony defect in cribriform plate. Based on the above imaging findings diagnosis of Frontonasal Epidermoid with patent dermal sinus tract opening on the nasal bridge and intracranial extension through cribriform plate defect was made.

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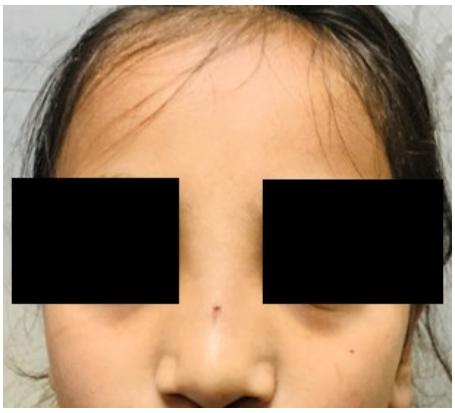
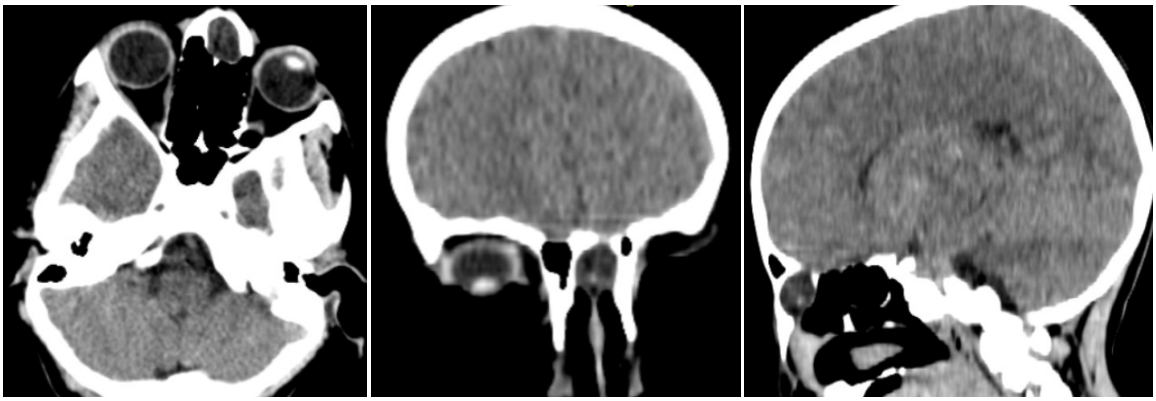
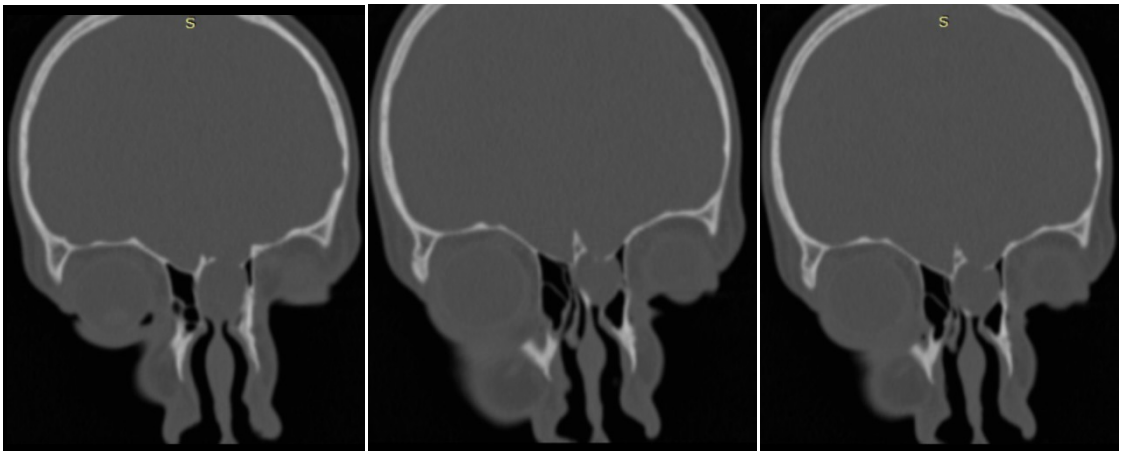


Figure 1:- Photograph showing dorsal nasal pit , broadening of the nasal bridge and dorsum of nose likely due to underlying frontonasal mass.



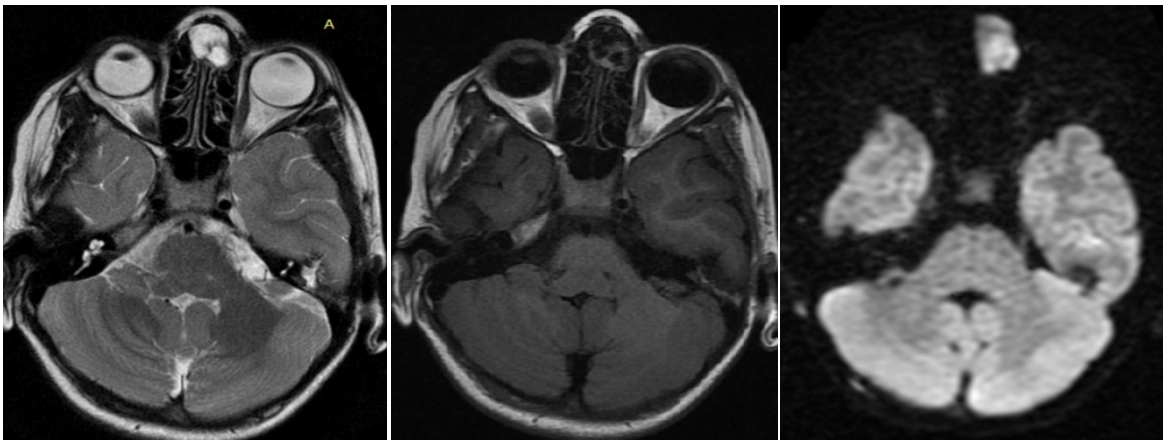
(a) (b) (c)

Figure 2:- CT soft tissue window images(a. axial, b. coronal and c. sagittal) showing well defined lesion in frontonasal region with CT attenuation upto 15 HU. However definite intracranial extension can't be established on CT.



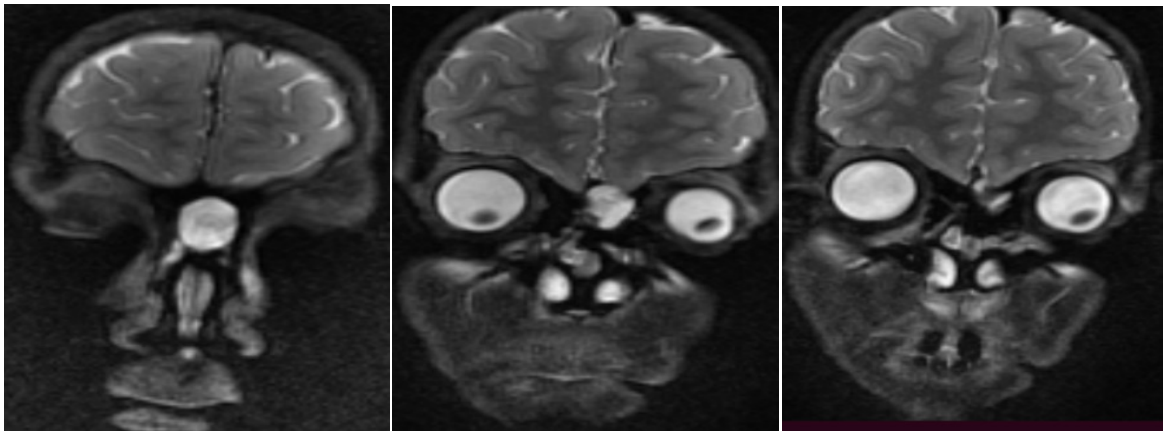
(a) (b) (c)

Figure 3:- Coronal CT bone window images(a,b,c) showing defect in the cribriform plate on left side.



(a) (b) (c)

Figure 4:- (a,b,c)Axial MRI images(T2,T1 and Diffusion weighted images respectively) showing well circumscribed, encapsulated, fluid signal intensity lesion with diffusion restriction on DWI seen in fronto-nasal region in the midline.



(a) (b) (c)

Figure 5:- Sequential(anterior to posterior) Coronal T2 weighted MRI images showing well defined T2 hyperintense lesion in frontonasal region causing mass effect on left basifrontal region with minimal intracranial extension along the floor of left anterior cranial fossa through a focal defect in cribriform plate.

Discussion:-

The case of frontonasal epidermoid that we have reported here is categorised under broad term Congenital midface anomalies (CMFA) or more specifically congenital midline nasal masses. These lesions are rare and the various differential diagnosis for congenital midline nasal masses include glioma, encephalocele, dermoid cyst, epidermoid cyst, hemangioma, lymphangioma and dacryocystocele[4].The understanding of normal embryological development of the frontonasal area is very important for correct diagnosis and characterisation of the lesions. Between the nasal bone and the inferior frontal bone, fonticulus nasofrontalis is formed which fuses and form frontonasal suture. Between the posterior aspect of the frontal bone, nasal bone and anterior aspect of the nasal cartilage, prenasal space is formed which extend from foramen cecum to osteocartilagenous junction. Now a dural diverticulum comes through the foramen cecum traverses the prenasal space, reaches upto the subcutaneous area and then involutes leading to obliteration of prenasal space[5]. Abnormality in this developmental process lead to various lesions of nasofrontal region. No or incomplete involution of dural diverticulm lead to formation of dermal sinus tract. When dural diverticulum become adherant to the ectoderm, during involution process skin elements are pulled into the prenasal space, it leads to formation of the dermoid and epidermoid cyst. If sequestrated neurogenic tissue is present in the partially involuted dural diverticulum, it leads to formation of nasal glioma with intracranial connection .When brain tissue herniates through the skull defect like patent fonticulus nasofrontalis and foramen caecum it leads to formation of frontonasal and nasoethmoidal encephaloceles respectively[4]. Accurate diagnosis and characterisation of these lesions can only be done by imaging modalities like CT and MRI which ultimately

helps in deciding the treatment plan. Dermoid and epidermoid cysts both contain ectodermal elements but dermoid cysts also contain skin appendages. Epidermoids are usually paramidline and occur near to columella whereas dermoids are usually midline and occur near glabella. [6,7]. Dermoid and epidermoid cysts are associated with a sinus tract opening, dimple, or tuft of hair on skin in 84%[8]. In a study of 40 adults with nasal dermoid sinus cyst, Vaghela and Bradley found intracranial extension in 11 (27.5%), mostly men[9]. In maximum cases intracranial extension is limited to a dural attachment in the anterior cranial fossa and not a true intracerebral involvement[10].On imaging epidermoid cysts usually have fluid attenuation on CT and fluid signal intensity on MRI whereas dermoid cysts also contain fatty component. MRI helps in differentiating epidermoid from arachanoid cysts as epidermoid cysts show diffusion restriction on DWI.

Conclusion:-

Midline frontonasal masses in children should be carefully evaluated in the light of clinical findings along with multimodality imaging approach including CT and MRI so that accurate diagnosis can be confidently given because these lesions can have possible intracranial extension that has to be established before any surgical intervention.

References:-

1. Pratt LW. Midline cysts of the nasal dorsum: embryologic origin and treatment. Laryngoscope1965;75:968-980.
2. Hughes GB, Sharpino G, Hunt W, Tucker HM. Management of the congenital midline nasal mass: a review. Head Neck Surg 1980;2:222-233.
3.Zapata S, Kearns DB. Nasal dermoids. Curr Opin Otolaryngol Head Neck Surg 2006;14(6):406-11.
4. Lowe LH, Booth TN, Joglar JM, Rollins NK. Midface anomalies in children. Radiographics. 2000;20(4):907-22.
5. Rodriguez DP, Orscheln ES, Koch BL. Masses of the Nose, Nasal Cavity, and Nasopharynx in Children. Radiographics. 2017;37(6):1704-1730.
6. Sadler TW. Langman's medical embryology. 5th ed. Baltimore, Md: Williams & Wilkins, 1985.
7. Castillo M. Congenital abnormalities of the nose: CT and MR findings. AJR Am J Roentgenol 1994; 162:1211-1217.
8. Barkovich AJ, Vandermarch P, Edwards MSB, Cogen PH. Congenital nasal masses: CT and MR imaging features in 16 cases. AJNR Am J Neuroradiol 1991; 12:105–116.
9. Vaghela HM, Bradley PJ. Nasal dermoid sinus cysts in adults. J Laryngol Otol 2004;118(12):955-62.
10. Chu EA, Ishii LE. Adult nasal dermoid sinus cyst. Ear Nose Throat J. 2010;89(8):E12-5.