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CASE REPORT

CASE REPORT-NASO-ETHMOIDAL ENCEPHALOCELE

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ABSTRACT: A rare case of 5yrs old boy came with chief complaint of swelling at nasal bridge since birth. It was gradually increasing in size, with recurrent attacks of cold. No history of trauma or fever. Radiograph of PNS shows opacification of left nasal cavity with deviated nasal septum towards right. Patient was further evaluated with CT- PNS and MRI- Brain (3-d drive thin sections).Finally diagnosis of naso-ethmoid encephalocele was made. Patient underwent surgery and final histopathological diagnosis was nasoethmoidal encephalocele.

KEYWORDS: Encephalocele, Frontoethmoid.

INTRODUCTION: An encephalocele is a herniation of intracranial contents through a midline skull defect. Also known as cephaloceles**,** these lesions are classified by their contents and location. Encephaloceles usually are located in the occipital (75 percent) or frontal areas (25 percent).

Basal and trans-sphenoidal encephaloceles are rare; they may appear between the ethmoid and sphenoid bones and extend into the upper pharynx. Encephaloceles that extend from the area of the orbit, nose or forehead are termed sincipital encephaloceles.{1}

CASE SUMMARY: 5 yrs old boy came with chief complains of swelling at nasal bridge since birth with recurrent attacks of cold frequently. Patient was referred to radiology department for evaluation. PNS Radiograph shows opacification of left nasal cavity with deviated nasal septum towards right.

NECT (3D) Revealed: Bony defect in cribriform plate and soft tissue density mass in left nasal cavity with intracranial extension, CT values +5 _+7.HU. Nasal septum deviated towards right side. Soft tissue density areas seen in bilateral maxillary, ethmoidal, sphenoidal sinus. Evidence of non pneumatization of frontal sinus.

MRI-BRAIN and PNS revealed mass in left nasal cavity measuring:- 36X21mms in sagittal diameter and 15mms in transverse diameter, with small bony defect in fonticulus nasofrontalis. Lesion extending superiorly through the defect and intracranial communication was noted. No other intracranial abnormality seen.

With above mentioned findings diagnosis of nasoethmoidal encephalocele was made. Patient underwent surgery and specimen was sent for histopathology.

HISTOPATHOLOGY: Report was multiple tissue bits show round to oval cells with scant cytoplasm in a fibrullary background, with areas of glial tissues and fragments of meningothelial cells with few mixed inflammatory cells. Histomorphological features consistent with encephalocele.

DISCUSSION: Nasal encephaloceles can be divided into frontoethmoidal and basal encephaloceles. Both conditions are very rare, but frontoethmoidal encephaloceles show a relatively high incidence (1:5,000) in southeast Asia.{2}

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The probable etiology could be:

1. Genetic component-occurring in families with spina bifida and anencephaly.
2. Teratogens x-ray irradiations, trifan blue, vitamin-a, arsenic etc.

The pathogenesis of encephalocele may be explained by separation of surface ectoderm and neuro-ectoderm in the midline just after closure of neural fold.

Nasalencephalocele can be divided into sincipital and basal variety. Sincipital form consists of naso-frontal, naso-ethmoid and naso-orbital subtypes, whereas basal form consists of trans-ethmoidal, spheno-ethmoidal, spheno-orbital and trans-sphenoidal sub-types.{3}

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.{4}

Computed tomography (CT) scan can be utilized, as it delineates bony defects very well. Magnetic resonance imaging (MRI) which complement the CT findings and is even superior to it by making vessels discernable and better illustrating intracranial communication in majority of cases.

As CT & MRI are very useful in making a diagnosis and to differentiate it from other nasal masses like polyp, mucocele, thus avoids biopsy and helps in proper surgical management.

CONCLUSION: Nasoethmoidal encephalocele is a rare condition, and it is important to differentiate it from other nasal masses like polyps, mucoceles. Imaging plays a very important role in diagnosis of nasoethmoidal encephalocele. CT and MRI are complimentary; CT well delineates the bony defect. MRI- shows intracranial communication and contents of the herniated sac.

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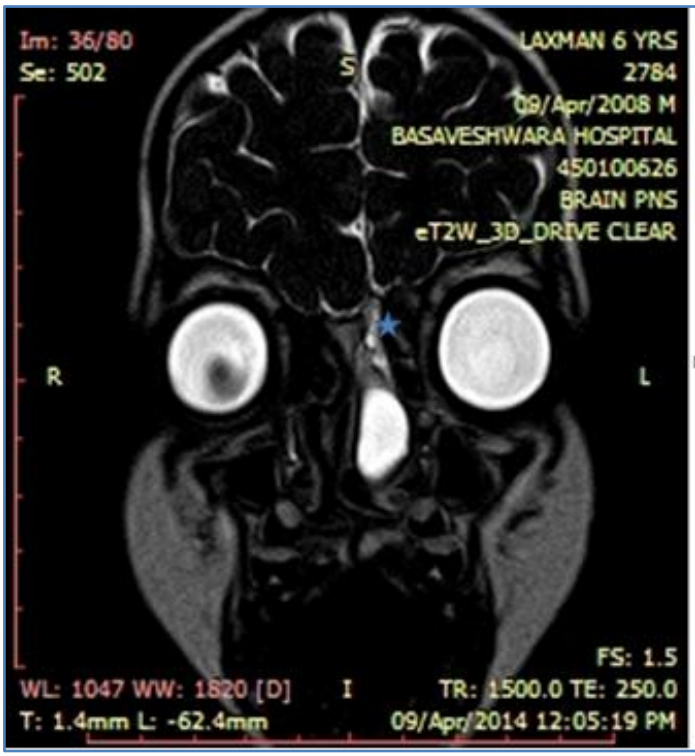
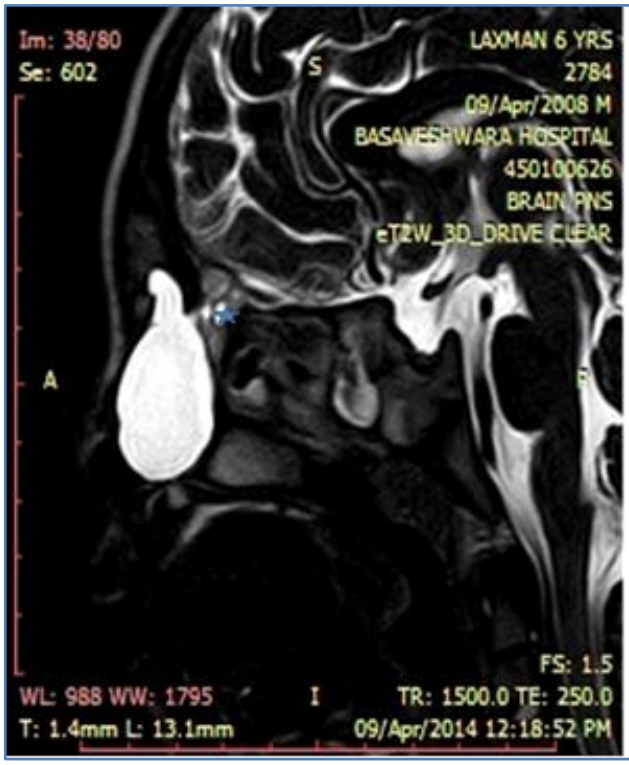


CORONAL T2W 3-D DRIVE SEQ.

Showing t2 hyperintense mass with small hypointense areas occupying whole of left nasal cavity with intracranial communication. .



SAGGITAL T2W-3-D DRIVE SEQ.

Demonstrating communication of the mass intracranially through defect in cribiform plate.

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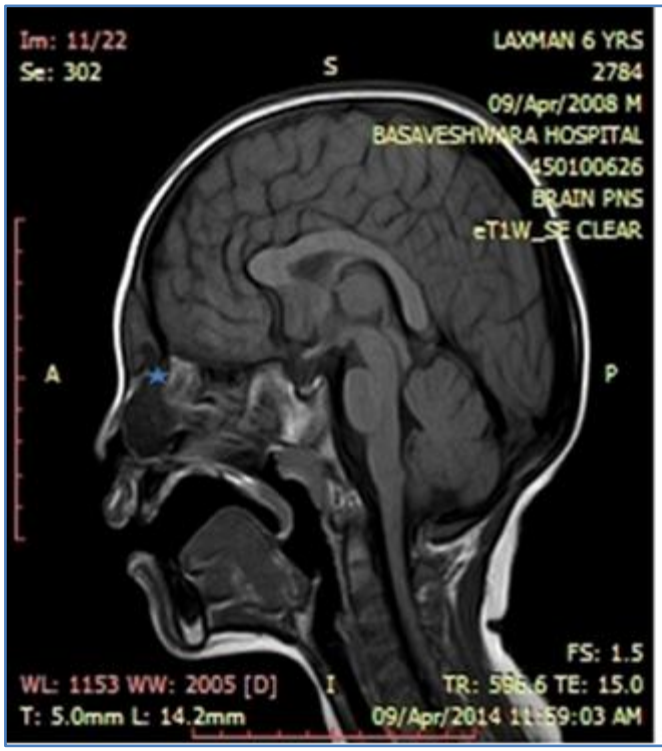
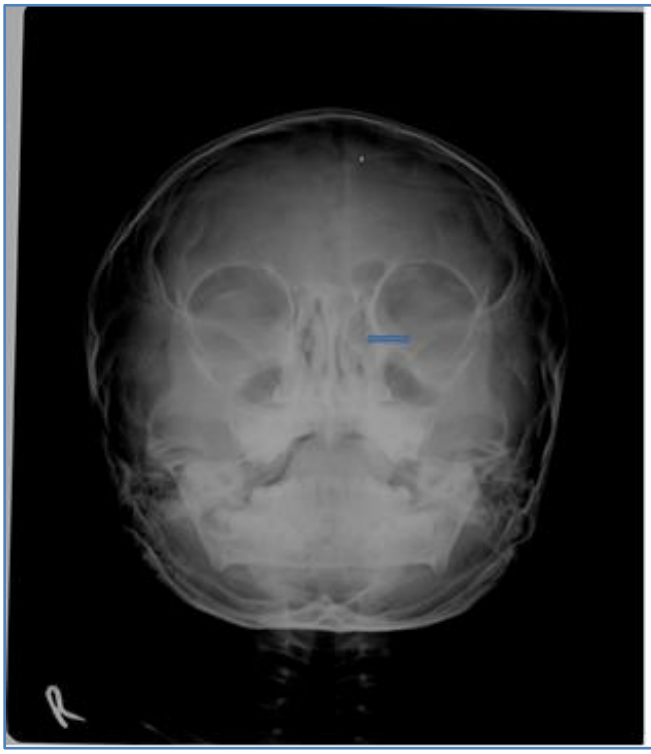


T1 SAGGITAL IMAGE

SHOWING THE INTRACRANIAL COMMUNICATIN .WITH SOFT TISSUE MASS OCCUPYING LEFT NASAL CAVITY.



X RAY PNS

Shows the deviated nasal septum towards right . With soft tissue opacity in left nasal cavity.

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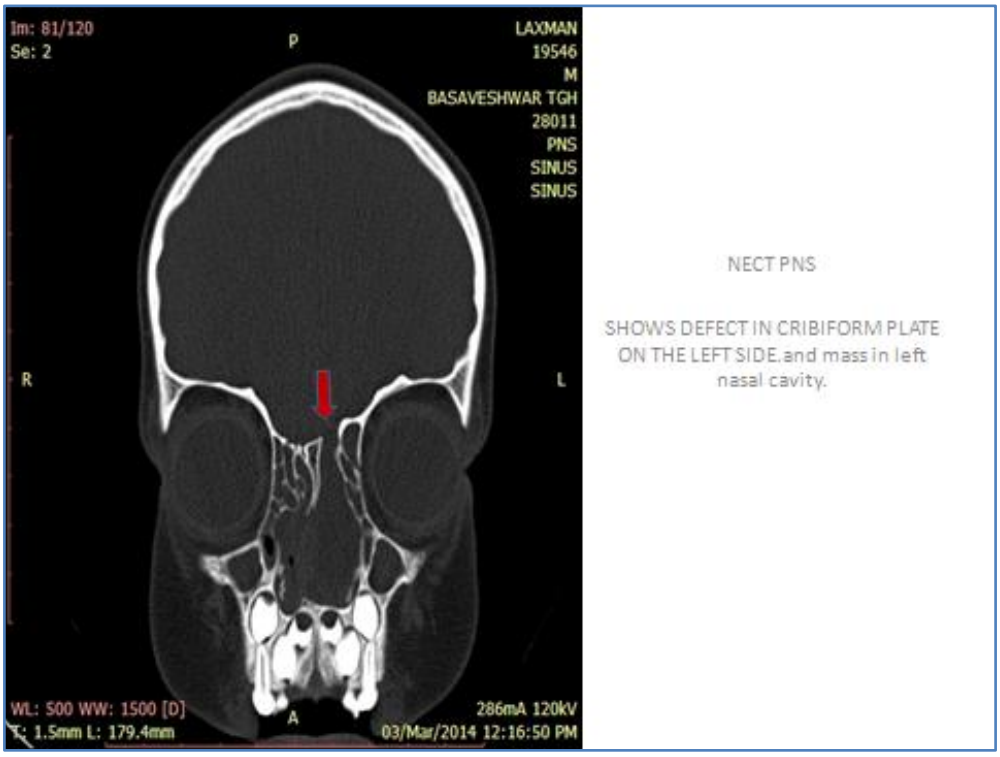

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