TRANS-SELLAR TRANSSPHENOIDAL NASOPHARYNGEAL ENCEPHALOCELE

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ABSTRACT: Trans-sellar trans-sphenoidal encephaloceles are rare and the trans-sellar variety is the least common. We present a 9-year-old male patient with trans-sellar trans-sphenoidal nasopharyngeal encephalocele which herniated into the nasopharynx through the congenital defect in the floor of sella. Patient presented with complains of nasal obstruction, nystagmus & visual disturbance in left eye. A trans-sellar trans-sphenoidal nasopharyngeal encephalocele was found on computed tomography (CT) and magnetic resonance imaging (MRI). We discuss our clinical findings and the results of preoperative computed tomography and magnetic resonance imaging along with review of this rare condition.

INTRODUCTION: Encephaloceles occur in approximately 1 in 3000 to 5000 live births1.2.3. Basal meningoencephaloceles are rare anomalies, reportedly constituting 1% - 10% of all encephaloceles and originate from a congenital opening in the midline region of the skull base, which permits meninges, neural tissue or both to herniated from the intracranial space4.5.6.7. Basal encephaloceles occur with an estimated incidence of one in every 35,000 live births1. Pollock classifies them as follows: (1) Sphenopharyngeal or trans-sphenoidal, when they protrude into the epipharynx and/or sphenoidal sinus; (2) spheno-orbital, when the protrusion is through the superior orbital fissure into the superior orbit producing unilateral exophthalmus; (3) sphenethmoidal, when the cerebral mass herniates through the sphenoid and ethmoidal bones into the posterior nasal cavity; (4) transethmoidal, when encephalocele extends into the anterior nasal cavity; and (5) sphenomaxillary, when the meningo-encephalocele passes through the superior orbital fissure into the orbit and through the inferior orbital fissure into the pterygopalatine fossa8,9. Transsphenoidalencephaloceles are rare and the transsellar variety is the least common variety.

We discuss our clinical findings and the results of preoperative computed tomography and magnetic resonance imaging of trans-sellar trans-sphenoidal nasopharyngeal encephalocele along with review of this rare condition.

CASE REPORT: A 9-year-old patient with normal built for age presented with complaints of nasal obstruction with anterior nasal discharge, nystagmus, headache and visual disturbances more in left eye since 1 year. Clinical examination showed large nasopharyngeal soft tissue mass lesion causing near total occlusion of nasopharynx. The site of origin of the mass was not able to make out. Visual examination showed horizontal nystagmus in both eyes with decreased vision on left side. He had no history of CSF rhinorrhea, meningitis/seizures. His physical exam was unremarkable and the patient was referred for further work-up.

Radiograph of skull showed defect in the floor of sella [Fig.1]. Axial non-enhanced computer tomography (NECT) showed large cerebrospinal fluid (CSF) density cystic lesion extending from sella, sphenoid sinus into nasopharynx [Fig 2]. Bony defect noted in the sellar floor with widening of sella [Fig.3]. Rest of the brain was normal. Diagnosis of trans-sphenoidalencephalocele was made.
Unenhanced & contrast enhanced magnetic resonance imaging (MRI) of brain showed large CSF intensity lesion extending from base of the brain in middle cranial fossa into the nasopharynx through the sella and sphenoid sinus [Fig 4 & Fig 5]. Part of optic chiasma was seen protruding into the proximal part encephalocele along with sellar widening. Pituitary gland was seen displaced posteriorly against the bone [Fig. 6]. Rest of the brain was normal. Diagnosis of Trans-sellar trans-sphenoidal nasopharyngeal encephalocele was made.

DISCUSSION: Encephalocele is a cystic congenital malformation in which the central nervous system (CNS) structures, in communication with CSF pathways herniate through the defect in the cranium. If it contains only meninges it is termed a meningocele, when it also contains brain tissue it is called a meningoencephalocele. The primary abnormality in the development of an encephalocele is a mesodermal defect resulting in a defect of the bone and dura associated with herniation of CSF pathways, brain tissue and meninges through the defect. One of the earliest and most widely accepted theories of basal encephalocele formation is the “adhesive theory,” published in 1827 by Sir Geoffroy Saint-Hilaire (as cited by Smith et al). This theory attributes the bone defect to a faulty separation of neuroectoderm from the surface ectoderm during neural tube formation, thereby preventing mesodermal tissue, which is to form bone, from interposing between the two germ layers. The root cause is the failure of surface ectoderm to separate from neuroectoderm early in the embryonic development.

Another possibility, as hypothesized by Kaufman et al, is herniation of tissue into the sphenoid sinus as a result of enlargement of congenital sphenoid "pitholes"; it has been estimated that up to 10% of the normal population may possess an aerated middle fossa floor. The hypothesis is that these small dehiscences may enlarge as a result of CSF pressure changes that occur during straining, coughing or normal physiological variations in intracranial pressure, thus allowing herniation of brain matter into sinuses.

Encephaloceles are classified as anterior (frontal, sincipital and basal) and posterior (infratorcular and supratorcular). Posterior encephaloceles are most common (75%) and basal ones most infrequent (1.5%). Encephaloceles occur more commonly in females than in males. Currently, most encephaloceles are diagnosed antenatally and present at birth. Postnatally, infants may present with CSF rhinorrhea and recurrent meningitis. They are often associated with midline craniofacial dysraphism. Some, particularly sphenoidal encephaloceles are often clinically occult and usually become apparent at the end of the first decade of life.

Basal encephaloceles are rare with an estimated prevalence of one in 35,000 births anomalies and are classified into five types: (1) sphenoethmoidal, transthyroidal, sphenooptoral, sphenomaxillary and trans-sphenoidal. (2) Sphenopharyngeal or trans-sphenoidal, when they protrude into the epipharynx and/or sphenoid sinus; (3) spheno-orbital, when the protrusion is through the superior orbital fissure into the superior orbit producing unilateral exophthalmus; (4) sphenoehtmoidal, when the cerebral mass herniates through the sphenoid and ethmoidal bones into the posterior nasal cavity; (5) transethmoidal, when the encephalocele extends into the anterior nasal cavity; and (6) sphenomaxillary, when themeningo-encephalocele passes through the superior orbital fissure into the orbit and through the inferior orbital fissure into the pterygopalatine fossa.

The trans-sphenoidal variant represents approximately 5% of basal lesions. It is divided into intrasphenoidal, extending into the sphenoid sinus, and true trans-sphenoidal, traversing the floor of
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the sinus and protruding into the nasal cavity or nasopharynx\textsuperscript{17}. Trans-sphenoidalencephaloceles are rare congenital anomalies that may be immediately apparent in infants with multiple cranial midline defects\textsuperscript{18}. In adults they can be of a spontaneous, traumatic or congenital origin\textsuperscript{19}. Associated findings in trans-sphenoidalencephaloceles include agenesis of the corpus callosum, CSF rhinorrhea, an ephypharyngeal soft tissue mass, visual defect, an endocrinological disturbance, and various optic and midface abnormalities\textsuperscript{17}, such as an abnormal development of the optic nerve, the so-called morning glory syndrome\textsuperscript{20}.

The majority of trans-sphenoidalmeningoencephaloceles are diagnosed during the first year of life due to manifestations such as respiratory distress caused by epipharyngeal obstruction, feeding difficulties, cranial midline defects with cleft lip or cleft palate, hypertelorism, optic malformations with anophthalmia, retinal abnormalities, optic nerve hypoplasia, unexplained bouts of recurrent meningitis or endocrine abnormalities\textsuperscript{9,21,22,23,24}. However, if there are no considerable difficulties and no distinctive facial anomalies during childhood, the diagnosis of the disease may be delayed up to adulthood, when distinctive symptoms such as rhinorrhea, visual defect or endocrine dysfunction occur.

It must be noted that not all cases fall into the above categories; Leblanc et al. have reported three cases in which meningoencephaloceles were found in the pterygopalatine fossa protruding through a defect at the base of the greater sphenoid wing near the foramen rotundum and the pterygoid process without the involvement of the orbit. These encephaloceles were not readily noticeable on CT scans of the middle fossa and were difficult to diagnose.

Several imaging features aid in the preoperative characterization of intrasphenoidalencephaloceles and subsequent management. Advanced imaging studies are necessary to confirm the diagnosis of trans-sphenoidalencephalocele and to define any neural or vascular elements that may be included in the herniation. CT scan and MRI are the most useful modalities for diagnosing encephalocele. In the present case, CT scan including reconstruction shows bone defects in the skull base and a well-circumscribed CSF density mass lesion in the extracranial area communicating with the intracranial space. MRI with gadolinium enhancement evaluated the content of the encephalocele and eliminated other intracranial anomalies. MR angiography may be needed to evaluate intracranial vasculature before surgical repair is performed.

REFERENCES:


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Fig. 1: Lateral skull radiograph showing bony defect in the floor of the sella.

Fig. 2: Axial NECT showing CSF density cystic lesion in sphenoid sinus.

Fig. 3: Reconstructed CT showing defect in the floor of sella & widening of sella.

Fig. 4: T1W sagittal MR imaging of brain showing CSF intensity cystic lesion herniating from base of brain into nasopharynx - Encephalocele.
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Fig. 5: T2W sagittal MR imaging of brain showing CSF intensity cystic lesion herniating from base of brain into nasopharynx - Encephalocele.

Fig. 6: CEMRI showing posteriorly displaced pituitary gland against bone.