

OPERATION TECHNIQUE OF ANTERIOR MENINGOENCEPHALOCELE: Transcranial Combined with Anterior Resection (Case Report)

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Meningoencephaloceles are a frequent condition characterized by protrusion of meningeal and brain tissue through a skull-base defect. Here, we report three cases of anterior meningoencephalocele operated on one-stage procedure, a combined of transcranial and anterior approach, for invagination removal of non-functional extracranial cerebral tissue with watertight closure of the dural defect; written consent was taken. All cases were nasoethmoid, the congenital anomalies between the nasal bones and the nasal cartilage.

Keywords: Menigoencephalocele, Nasoethmoid, Transcranial and Anterior Resection

INTRODUCTION

Encephaloceles is a serious congenital anomaly characterized by protrusion of meningeal and brain tissue along the midline of the cranial vault or through a skull-base defect. The case is far less common compare to spinal dysraphism. It may content meninges (meningocele) or brain tissue and meninges (meningoencephalocele).¹ Encephaloceles occur in 1 : 5.000 to 10.000 births worldwide.² The primary abnormality in the development of an encephalocele is a mesodermal defect resulting in a defect in the calvarium and dura associated with herniation of cerebrospinal fluid (CSF), brain tissues and meninges through defect. According to the location, encephaloceles are classified in: cranial vault, frontoethmoid, basal, occipital and posterior fossae.³ For nasal encephalocele devided into sincipital and basal variety. Sincipital form consists of nasofrontal, nasoethmoid and nasoorbital, whereas basal form consists of: transethmoidal, sphenothmoidal, trans sphenoidal, and sphenoorbita. The commonest site of encephalocele is occipital (75-80%), followed by frontoethmoidal (13-15%), parietal (10-12%) or sphenoidal. Occipital and posterior encephalocele are more common in Western population (European and North America), whereas anterior encephaloceles are higher in Southeast Asia and Southern Asia.^{2,4} It is not easy to manage the congenital case of encephalocele, since there were many perioperative problem and we try to manage each cases carefully. We present here three cases with nasoethmoid meningoencephalocele that

was operated on one-stage procedure which combine the transcranial and anterior resection.

Cases Report

Case 1

A 3 month-old baby boy was complaint by his parents, that there is a lump on his nose since he was born, the lump has gradually increased in size, impulse on crying and coughing was felt over the swelling, the lump's size as big as a pingpong ball (Figure 1).



Figure 1

A) and B) The features of a midline nasoethmoidal encephalocele. Note the interorbital hypertelorism, the inferior canthal dystopia; C) and D) There was bone defect at nasoethmoid bone, with cyst like CSF content

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He was born from P2A1 mother, at term helped by traditional midwife, spontaneous delivery, immediately crying, baby birth weight (BBW) 2800 grams, head circumference 40 cm (with normal size are 39-43 cm), painless swelling, translumination test positive and anterior fontanelle still open but not tense. Associated hypertelorism was noted.

Case 2

A 8 month-old baby girl had a lump on her nose since she was born, the lump getting bigger during time especially when she was crying, until the size as big as a tennis ball (Figure 2). She was born from P4A0 mother, at term and helped by traditional midwife, spontaneous delivery, immediately crying, BBW 4000 grams, head circumference 54 cm (47-52 cm), painless swelling, translumination test positive and anterior fontanelle still open but not tense. Associated hypertelorism was noted.



Figure 2

A) and B) This baby girl demonstrates the features of a nasoethmoidal encephalocele. Note the interorbital hypertelorism, the inferior canthal dystopia. C) and D) There was bone defect at nasoethmoid bone with cyst like CSF content

Case 3

One-year old baby girl has had a lump on her nose since she was born, the lump as big as a bird egg, and not getting bigger during time or when patient crying or coughing (Figure 3). She was born from P3A0 mother, 36 weeks, spontaneously and helped by midwife, immediately crying, BBW 2700 gram. Fontanela anterior was closed, head circumference 42cm (41-46 cm), there was a mass

size 4x3 cm at nasion, soft and tender, translumination negative. Associated hypertelorism was noted.



Figure 3

A) and B) This baby girl demonstrates the features of a nasoethmoidal encephalocele. Note the interorbital hypertelorism. C) and D) There was bone defect at nasoethmoid bone

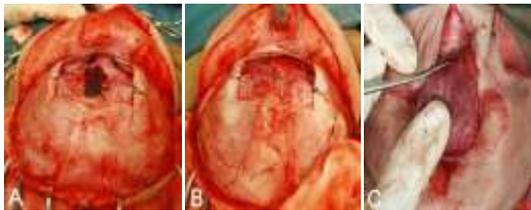
Operation Technique

Inform concern for all patients were signed by their parents. This study approved with ethical clearance obtained from local authority ethical committee medic.

Imaging by computed tomography (CT) scan was carried out for all patients, herniation of meninges and brain tissue of frontal lobe through a defect in the skull base presented as a lump on the nose and suggestive at nasoethmoid (Figure 1, 2, and 3 C and D).

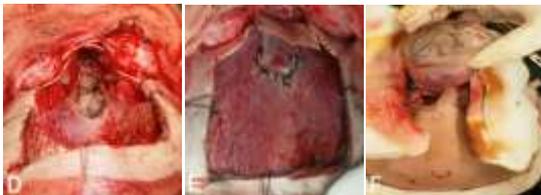
Patient was laid supine with the head above the heart at 30°. The scalp was shaved thoroughly and a marking made for skin incision. The area was then cleaned and draped. For transcranial resection: a bicoronal skin incision was made from the right temporal, from the tragus ipsilateral to the tragus on contralateral side after local infiltration of subcutaneous tissues with pehacain. Rinnies' clips were used to secure the bleeding at the skin edges. Dissection of the scalp was made to the lower limit of the defect or abnormality. The galea layer was incised with monopolar diathermy and periosteum separated from the bony layer by using periosteal

elevator. Bifrontal craniotomy was made using the Smith craniotomes and dura spatulas. The encephalocele was approached via the extradural route without injuring the dural protective layer. The nasal bone defect was identified and the herniated portion of abnormality was opened and neck of the sac was identified, after the occlusion of the sac's pedicle, the whole sac that associated with dysplastic brain tissue was cut. The cut ends of sac closed watertight and supported with galea tissues and 1 ml of fibrinogen product was injected onto the area. the bone defect was reconstructed with autogenous bone graft taken from part of the burr hole bone fragments. The layers were then closed with Dexon 3/0 for subcutaneous and Dafilon 3/0 for skin (5). For anterior resection: a skin incision was made in Y shape manner at the fore head-nasal junction and dysplastic brain tissue in remaining sac was removed, along with closure of skin defect made by the lump (Figure 4). All patients were in a very good condition after the surgery.



Case 1

- A) we exposed the invagination of the duramater,
- B) Duramater close with duroplasty watertight,
- C) Non functional extracranial brain tissue.



Case 2

- D) We cut the invagination of the meningen.
- E) Duramater close with duroplasty watertight.
- F) Non functional extracranial brain tissue



Case 3

- G) We exposed the invagination of the meningen.
- H) Duramater close with duroplasty watertight.
- I) Non functional extracranial brain tissue from anterior.

Figure 4

Operation Technique For all Cases

DISCUSSION

Encephalocele is a severe malformation with the involvement of many facial anatomical areas, treatment of which requires a multidisciplinary neuro-craniofacial team. Fronto ethmoidal encephaloceles present with a facial mass. Nasofrontal encephaloceles appear at the root of the nose above the level of nasal bone. Nasoethmoidal encephaloceles are situated inferior to the nasal bones and naso-orbital encephaloceles cause proptosis and displacement of eye ball. The incidence of sincipital encephalocele compared with occipital encephalocele is substantially greater in the tropical latitudes, particularly in parts of Asia and Africa, and its also more common in parts of Russia. However, the occipital-to-sincipital encephalomeningocele ratio is reversed in North America, Australia, Europe, and parts of South America, varies from 2.5:1 to 15:1. The occipital encephaloceles occur in 1 to 3 per 10.000 live births. The etiology of sincipital encephalocele is unknown but its includes ethnic, genetic, and environmental factors and paternal age.^{2,6}

Anterior encephalocele are associated with hypertelorism, midline craniofacial dysraphism, agenesis of corpus callosum, lipoma and subependymal heterotopias.⁷ In the growing-child patient, it is necessary to reconstruct at an early age to avoid further distortion of the craniofacial anatomy. Many authors have described the various operation techniques in repairing the fronto-nasoethmoidal encephalocele. In the past, operations for meningoencephaloceles were performed in two stage procedures; as the first stage of transcranial resection done, the second stage of anterior resection will perform six months up to 1 year after the fist surgery, waiting until the fibrotic scars occurs. Since the development of advanced neurosurgical technique, an intracranial approach combined with extracranial resection as one stage procedure is one of the option.

Details of the surgical techniques are described above. In authors opinion, the two step procedures has disadvantages which requires a longer period for perform the second surgery and has additional psychological effect to the family and the patient; for not removed the nose-mass skin or lump skin of their children. The advantages of our transcranial combined with anterior resection are: it will provide a wide exposure of the dural defect that make us easier to close the dural in water tight manner, decrease the possibility of CSF leakage, decrease the possibility of infection even and easier to performed duroplasty compare with two stage procedure.

Survival rate is higher, nearly 100% in anterior encephalocele compared to posterior encephalocele (55%), where vital structure of brain parenchyma might have herniated to the skull defect. Indication of surgical repair of the lesion

(anterior encephalocele) is usually cosmetic. The principal is to return the cerebral components in to the cranial cavity along with amputation of dysplastic tissue, closure of bony defect and reconstruct the skin. The presence of microcephaly with a large occipital encephalocele containing significant brain tissue also a predictor of poor neurological outcome.⁶⁻⁸

CONCLUSION

The difficulties of management of anterior meningoencephalocele can be reduced with a good perioperative management and right decision to performed surgery that give more beneficial. We believe this combine technique, one stage procedure, has many advantages then the conventional two stage procedure. Thus rarity of occurrence of nasoethmoidal meningoencephalocele, highlights the importance of research in the field diagnosis and surgical management of these cases.

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