

Case Report

Occipital Meningocele

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ABSTRACT

Background: Neural tube formation during embryonic life occurs throughout the intrauterine period. Many external and internal factors can cause neural tube defects (NTD). Broad classification of NTD divides the whole group into "Open" or "Closed" NTDs. In open NTDs, primary neurulation is disrupted and may form anencephaly, myelomeningocele (open spina bifida), and craniorachischisis. A Neural lesion covered by skin forms a closed NTD develops, e.g., meningocele and spina bifida occulta.

Case report: Present case 19+2 weeks male fetus, sent for autopsy from Obst & Gynecology depart to Anatomy department of Government Medical College, Chandigarh. The reason for MTP was diagnoses made on USG as occipital Meningocele. The mother was 26 years old with P1A1 obstetrical history. Advised termination of pregnancy. Past, parental, parental history was not suggestive of any causative factor.

Findings: A small meningocele in the occipital region was seen on external examination. After the autopsy, no other associated anomaly was found except an incomplete fissure in the left lung. The meningocele showed herniation of the meninges; no brain tissue was found in the meningocele. On opening the cranial cavity, pachygyria in the cerebral hemispheres were seen.

Conclusion: The present case was classified as Tectocerebellar Dysraphia with occipital encephalocele, as the fetus had pachygyria in the cerebral hemispheres. We could not find any suggestive cause of this defect.

KEY WORDS: Meningocele, Encephalocele, NTD, Occipital.

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INTRODUCTION

When the neural tube's morphogenetic process fails during embryogenesis, severe birth abnormalities of the central nervous system, known as neural tube defects (NTDs), are caused.

NTDs can be broadly categorized as either "Open" or "Closed" NTDs. The primary

neurulation is disrupted in open NTDs, which causes anencephaly, myelomeningocele (open spina bifida), and craniorachischisis. Due to the open neural tube's deformity, neurological function is lost below the lesion level. When lesions are covered by skin, and a closed NTD develops, secondary neurulation is typically disrupted, causing anything from severe

spinal cord tethering to spina bifida occulta. A congenital bone defect in the occipital region of the skull is known as a meningocele, which causes the meninges to herniate. A meningocele is a herniation of the meninges; a meningoencephalocele is a herniation of the meninges and the brain tissue. It may be accompanied by various brain issues and hereditary and chromosomal syndromes and can be classified by the anatomical site of the abnormality into occipital, frontoethmoidal, lumbar, and sacral. The prognosis is poor, and those fetuses that survive almost always have severe neurological defects.

Aims and objectives: In the Department of Anatomy, GMCH-32, Chandigarh, we regularly receive fetuses between 12 weeks to 28 weeks referred from the Department of Obstetrics and Gynaecology, GMCH-32, for autopsies. The aim and objective of this study are to find any associated malformations and causes of the defect.

CASE REPORT

An autopsy was done on a 19+2 weeks male fetus in the Department of Anatomy, Government Medical College and Hospital, Chandigarh. The fetus was sent from the Department of Obstetrics and Gynaecology, Government Medical College and Hospital, Chandigarh, after the medical termination of pregnancy. The Autopsies are performed on fetuses aged 12 - 28 weeks, as per standardized protocol based on the Helsinki Declaration after informed written consent.

Antenatal, obstetrical, and any other relevant history were taken from the mother and father.

The mother was 26 years old with P1A1 obstetrical history. Maternal family history indicated no chromosomal abnormality. No history of hypertension, diabetes, thyroid, epilepsy, tuberculosis, and bronchial asthma. No maternal history of any drug exposure was given. The mother was fully vaccinated for tetanus toxoid and had folic acid intake during the first trimester. Both parents have no history of smoking or drinking alcohol.

- The ultrasound Level 2 indicated a maternal cervix length of 3.90 cm, internal os closed,

and no fluid seen in the cervical canal. The fetal anatomy on ultrasonography indicated a thin linear defect involving the basiocciput through which there was herniation of meninges and neural elements measuring 1.2x1.3 cms. Cavum septum pellucidum was thin. The lateral ventricle measured 8-9mm. The posterior horn of the lateral ventricle showed a pointed configuration. An anechoic lesion measuring 9x4mm was seen interrupting the interhemispheric fissure. There was a widening of the cervical spine.

External Examination: A small meningocele in the occipital region of 2.5x2.5 inches. No other CMF was found externally (Fig no. 2,3 & 5). The fetus was photographed, followed by an autopsy.



Fig. 1: Image showing the cerebral hemispheres and the cerebellum.



Fig. 2: Lateral view of the fetus showing occipital meningocele.

Internal examination: No other associated anomaly except an incomplete fissure in the left lung was found (Fig. no.4).

The meningocele showed herniation of the meninges; there was no brain tissue found in the meningocele (Fig no. 1).



Fig. 3: Posterior view of the fetus showing occipital meningocele



Fig. 4: Image showing internal findings.



Fig. 5: Meningocele with brain tissue.

DISCUSSION

A congenital abnormality of the dura and cranium with extracranial herniation of any intracranial structure is called a cephalocele. Cranial Meningoceles, Cranial Meningoencephaloceles, Cranial Glioceles, and Atretic Cephaloceles are the four subtypes of cephaloceles.

Cephaloceles have irregular skin cover, size, and shape. They could be head-sized masses, small nubbins, broad-based or pedunculated growths. Some are bilobed. They might range from mixed solid-cystic to cystic in consistency.

Cephaloceles are a result of errors with the fusing of the neural tubes. Anencephaly and encephalocele are conditions that are specifically brought on by changes in the rostral neural tube's closure. The deficient caudal fusion of the neural tube results in myelomeningocele. Encephalocele and myelomeningocele usually appear around the 26th day of life, whereas anencephaly typically appears around the 24th day of life.

Although these structural anomalies can affect any part of the skull, they most frequently affect the occipital region (70%–90% of cases). Encephaloceles occur in 0.84 out of every 10,000 live births. There has been a reported rise in incidence among females and babies weighing under 2500 grams.

The multifaceted etiology of NTD involves both genetic and environmental factors. Maternal exposure to environmental risks, such as occupational pesticide exposure; comorbid conditions, such as obesity and diabetes; infections, such as cytomegalovirus and toxoplasma; nutritional deficiencies, such as folic acid deficiency; drug history, and other factors are among the risk factors that have been implicated. It has been demonstrated that folic acid intake by pregnant women can significantly reduce the chance of neural tube abnormalities (up to 70%). Genetic and environmental factors are both involved in the multifactorial etiology of NTD. Maternal exposure to environmental risks, such as occupational pesticide exposure; comorbid conditions, such as obesity and diabetes; infections, such as cytomegalovirus and

toxoplasma; nutritional deficiencies, such as folic acid deficiency; drug history, and other factors are among the risk factors that have been implicated. It has been demonstrated that folic acid intake by pregnant women can significantly reduce the chance of neural tube abnormalities (up to 70%). Two rare subsets of occipital cephalocele are:

An occipital cephalocele called the posterior fossa ventriculocele is almost entirely made up of a cyst that connects to the fourth ventricle. The occipital cephalocele can be high or low. The cyst may develop from the fourth ventricle's lateral recess or the midline rhombic roof. The ventriculocele may be accompanied by hydrocephalus and callosal dysgenesis.

Inverse cerebellum with occipital cephalocele is a deformity in which a complicated occipital encephalocele comprises hypoplastic cerebellar hemispheres connected to the brain stem by a dorsal extension of the midbrain tectum. It is also known as tectocerebellar dysraphia with occipital encephalocele. There is no vermis. The olives and the pons are (partially) covered by the ventral and lateral extensions of the cerebellar hemispheres. The upper surface of the brain stem is exposed to the fourth ventricle and aqueduct. The midbrain's tectum is widened, forked, and devoid of the corpora quadrigemina. The aqueduct and tectum expand posteriorly through the foramen magnum and ostium of the encephalocele. Each hypoplastic cerebellar hemisphere has a sizable ventricular structure and appears to have developed independently from one fork of the cleft stem. The corpus callosum is inadequate. Gyrus aberrations are common, mainly focal polymicrogyria.

CONCLUSION

The present case of meningocele resembled more Tectocerebellar Dysraphia with occipital encephalocele, as the fetus has pachygyria in the cerebral hemispheres.

No associated congenital malformations could be found in the central nervous system or any other system.

We could not find any suggestive cause of this defect.

Author Contributions

Parneet Sandhu: Autopsy and Practical work, Interpretation & summary. **Anshu Sharma:** Concept & design of the study, Interpretation, drafting for publication. **Kanchan Kapoor:** Facilitation of work

Conflicts of Interests: None

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