

Case Report

Prenatal Differential Diagnosis and Prospective Management of Hydranencephaly

Diagnosis Diferensial Prenatal dan Manajemen Prospektif dari Hidranensefali

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Abstract

Objective: To report a rare case of hydranencephaly that was diagnosed during prenatal period. We also provided further review of differential diagnosis and management performed in Ende District General Hospital, based on appropriate literatures and guidelines available.

Methods: Case Report.

Case: A 27-year old primigravida women was diagnosed with term pregnancy (37 weeks of gestation) and intra-uterine singleton live fetus with hydranencephaly via ultrasonography. In this patient, emergency caesarean section (CS) was performed to prevent complication of cephalo-pelvic disproportion (CPD), involving teamwork between obstetrics and perinatology. A male neonate was born weighing 3000 grams, head circumference of 32 cm, and APGAR score of 2/4/7/8 suggestive of asphyxia and respiratory distress. The newborn was immediately transferred to Neonatal Intensive Care Unit (NICU) for further resuscitative management and observation. The newborn remains in stable condition after resuscitative management was given. Post-natal transcranial sonography (TCS) of the newborn was performed by a pediatrician, and the diagnosis of hydranencephaly was confirmed. The newborn was later referred to facilities with neurosurgery department for further evaluation and intervention.

Conclusion: Early prenatal recognition of hydranencephaly and exclusion of similar differential diagnosis, which includes: hydrocephalus, holoprosencephaly, porencephaly and schizencephaly, are fundamental in formulating proper multidisciplinary management with pediatric and neurosurgery department, which may consequently improve the newborn's life expectancy.

Keywords: differential diagnosis, hydranencephaly, management.

Abstrak

Tujuan: Melaporkan suatu kasus langka hidranensefali yang kami diagnosa dalam periode prenatal; dan memberikan ulasan lanjut mengenai diagnosis diferensial dan manajemen yang kami kerjakan di Rumah Sakit Umum Ende, berdasarkan literatur dan pedoman ilmiah yang tersedia.

Metode: Laporan Kasus.

Kasus: Seorang perempuan primigravida berusia 27 tahun di diagnosa dengan kehamilan aterm (37 minggu gestasi), janin tunggal hidup intrauterin dengan hidranensefali via ultrasonografi. Pada pasien ini dilakukan Sectio Caesarea (SC) cito untuk mencegah komplikasi dari disproporsi kepala-panggul dengan kerjasama tim kebidanan dan perinatology. Lahir bayi laki-laki dengan berat badan 3000 gram, lingkaran kepala 32 cm, dan skor APGAR 2/4/7/8 dengan kesan asfiksia dan distres pernapasan. Bayi segera dipindahkan ke Neonatal Intensive Care Unit (NICU) untuk penanganan resusitatif lanjut dan observasi. Bayi tetap bertahan dalam kondisi stabil setelah diberikan manajemen resusitatif. Pemeriksaan sonografi transkranial pasca-natal dilakukan oleh dokter anak, dan diagnosis hidranensefali terkonfirmasi. Bayi kemudian di rujuk ke fasilitas dengan ketersediaan departemen bedah saraf untuk evaluasi dan intervensi lanjut.

Kesimpulan: Deteksi dini prenatal hidranensefali dan eksklusi diagnosis diferensial serupa, yang mencakup: hidrosefalus, holoprosensefali, porencefali, dan skizensefali, adalah dasar dari formulasi manajemen multidisiplin yang baik antara departemen pediatrik maupun bedah saraf, sehingga dapat meningkatkan kualitas dan ekspektasi hidup bayi baru lahir.

Kata kunci: diagnosis diferensial, hidranensefali, manajemen.

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Received: July, 2021 Accepted: May, 2022 Published: July, 2022

INTRODUCTION

Hydranencephaly (HE) is a condition most commonly associated with cerebral cortex absence in bilateral hemispheres or unilateral hemisphere (Hemi-Hydranencephaly / HHE) in rare occasions. Cerebral cortex tissue is replaced with cerebrospinal fluid, necrotic glial, and/or ependymal debris filled sac covered in membranous leptomeninges.^{1,2} It is one of the most rare severe forms of bilateral cerebral cortical anomaly which only affects approximately 2.1 to 20 out of 100,000 births or pregnancies; and shares close resemblance with other conditions, such as: hydrocephalus, holoprosencephaly, porencephaly and schizencephaly.^{2,3} A specific incidence especially in Indonesia is still difficult to determine with very limited data, and currently no case has been reported. Despite recent medical advancements, doubts still persist on etiopathogenetic, onset, and diagnostic aspects of HE due to its similarity with conditions mentioned above.¹ HE commonly has a poor prognosis; since most affected individuals died in-utero or have a life expectancy of 1 year at most with a burden of developmental delay, drug-resistant seizures, spastic diplegia, severe growth failure and respiratory infections. However, some survivors with proper maintenance of brainstem functions (temperature, blood pressure, and cardiovascular function) have been reported to survive at the age of 20, 22, and 32 years old respectively.⁴

Based on these reports, recognizing hydranencephaly from its differential diagnosis is fundamental in formulating proper multidisciplinary management with pediatric and neurosurgery department, which may consequently improve the newborn's life expectancy. In this paper, we reported a case of at term fetus with prenatal and postnatal sonographic findings consistent with hydranencephaly in our obstetrics and gynecology department.

CASE

A 27-year old primigravida women on her 37th week of gestation was referred by a private midwife practice for the first time to our labor and delivery unit in Ende District General Hospital due to prolonged active phase labor with cephalo-pelvic disproportion suspicion. The patient had experienced signs of labor such as constant

progressive uterine contractions with bloody show for the past two days, and fluid leakage symptom was denied; throughout the pregnancy, the patient felt normal fetal movements. The patient rarely had prenatal check-up visits; only visited once in each first and second trimesters respectively with no third trimester prenatal visits. The patient also denied having history of previous ultrasound examinations throughout her pregnancy. The patient had previously done laboratory blood hemoglobin (Hb) examination during her sixth week of gestation to which her Hb was 12 g/dL; previous urine dipstick, Hepatitis B Surface Antigen (HbsAg), anti-syphilis (VDRL) and anti-HIV showed no significant abnormalities; She recalled having tetanus toxoid injection once during her sixth week prenatal check-up visit. The patient denied having previously tested for TORCH (Toxoplasma, Rubella, Cytomegalovirus and Herpes Simplex). The patient admitted that she skipped routine folic acid and ferrous sulfate medication due to rare prenatal check up visits.

Physical examination and vital signs were within normal limit. The patient's height and weight measured 147 cm and weight 46 kg respectively, which is considered to be normal. The clinical obstetric examination revealed oblique lie fetus with hardly distinguishable fetal presentation and it still floats beyond upper pelvic outlet. Vaginal examination revealed her cervix was dilated at 4 cm with intact amniotic membrane, with confirmed head presentation still floating higher than hodge 1 pelvic plane suggestive of cephalo-pelvic disproportion, and occipital fontanel facing the left side of the patient.

2D-Transabdominal ultrasonography was performed, revealed a singleton intrauterine pregnancy in oblique lie position, with fetal heart rate of 134 beats per minute (M-Mode) and fetal movement was unremarkable. Fetal biometry was equivalent to 37 weeks of gestation (Hadlock Standard). Intracranial sonography showed fluid filled cranial cavity with absent of most bilateral cerebral cortex and falx cerebri disruption; thalami and midbrain structures were still present; cerebellum and other posterior fossa structures were still present **[Figure 1]**. Amniotic fluid index was measured 150 mm still within normal range with no polyhydramnios present. Overall sonography results was suggestive of hydranencephaly.



Figure 1. 2D Transabdominal Ultrasound of fetal head (Axial View) at 37 weeks' gestation: (A) transventricular plane showing absent of bilateral cerebral cortex with no intact cortical rim and falx cerebri disruption, (B) transthalamic plane showing thalami and midbrain structures were still present, (C) transcerebellar plane showing cerebellum and other posterior fossa structures were still present.

Given the severity of the clinical picture, we convinced the patient to undergo emergency caesarean section (CS) since the patient's gestational age is at term and to prevent further obstetric complications that may arise from cephalo-pelvic disproportion. The patient's pre-operative blood work revealed O positive blood group with blood count results within normal range; urinalysis, anti-HIV and HbsAg infection markers were confirmed uneventful. The Pediatric-Perinatology department was informed of the about the situation and parents were given proper counseling regarding the child's poor prognosis and potential post-natal management. The newborn was a 3000 grams male, with normal physical appearance [Figure 2]. The head of the newborn was of normal size (head circumference = 32 cm), however the head was particularly transilluminated. Placenta and umbilical cord appearances were normal. Post-operative course of the patient (mother) was uneventful.

The newborn was immediately transferred to the neonatal intensive care unit (NICU) due to moderate asphyxia and respiratory distress, with APGAR score of 2/4/7/8. At birth, the newborn was cyanotic, weakly crying when stimulated, with slow heart rate and respiratory below 100 bpm and 60 bpm respectively; however the condition improved after proper neonatal management was given. Continuous positive airway pressure (CPAP) was installed with positive expiratory end pressure (PEEP) of 7, oxygen flow rate (FLOW) of 6 L/min, and fraction inspired oxygen (FiO2) of 30%. The newborn was advised for oral fasting and oral-gastric tube (OGT) was installed to minimize risk of aspiration; fluid administration of intravenous 10% dextrose solution (D10%) was given at a starting rate of approximately 7 cc/hours (60 cc/kg/24 hours with increments of 10 cc/kg/24 hours). Antibiotic injections of ampicillin 150 mg/12 hours (50 mg/kg/12 hours)

and gentamicin 15 mg/24 hours (5 mg/kg/24 hours) for at least 7 days were given since the newborn showcased potential signs of neonatal sepsis. The newborn's postpartum blood work were within normal range, further laboratory analysis of TORCH (toxoplasma, rubella, cytomegalovirus, and herpes simplex) infections were advised but the patient refused for checkup. Transcranial sonography (TCS) of the newborn was performed by a pediatrician one day after delivery. TCS revealed fluid filled cranial cavity with absent of most bilateral cerebral cortex and falx cerebri disruption; however, midbrain structures, cerebellum and other posterior fossa structures were less visible this time [Figure 2]. TCS therefore confirmed the prenatal diagnosis of hydranencephaly. The newborn was observed for seven days before getting referred to facilities with neurosurgery department to further diagnostic evaluation the case and perform surgical intervention if indicated.



Figure 2. (A) General Appearance of The Newborn, (B) (C) 2D Transcranial Sonography (TCS) of The Newborn's Head (Mid-Coronal View) and (Mid-Sagittal View) respectively revealed absent of bilateral cerebral cortex with falx cerebri disruption; while midbrain structures, cerebellum and other posterior fossa structures are less visible.

DISCUSSION

Hydranencephaly (HE) is a condition most commonly associated with cerebral cortex absence in bilateral hemispheres or unilateral hemisphere (Hemi-Hydranencephaly/HHE) in rare occasions. It is one of the most rare severe forms of bilateral cerebral cortical anomaly which only affects approximately 2.1 out of 100,000 births or pregnancies. The etiopathogenesis of HE remains unknown; however, there are some hypothesized theory, which includes: vascular accident/circulation developmental theory, early organogenesis disruption theory and encephaloclastic destruction theory. Many reports suggested that vascular accident has been associated with: intrauterine infections (toxoplasmosis, enterovirus, adenovirus, parvovirus, cytomegalovirus, herpes simplex, Epstein Barr and respiratory syncytial virus), toxic exposures (smoking, cocaine abuse, estrogen, sodium valproate), and other risk factors (young maternal age, mono-chorionic twin pregnancy, factor XIII deficiency and prior intra-cerebral hemorrhage).^{2,3}

Prenatal Differential Diagnosis

Prenatal diagnosis of hydranencephaly (HE) can be determined early during prenatal period by ultrasonography (US). Maternal clinical manifestations during pregnancy are unreliable in determining diagnosis, since mother typically feels normal fetal movements just similar to this case; unless there is a decrease or absent in fetal movements with suspicion of intra-uterine fetal demise, which promotes further ultrasonography examination. Fetus with HE mostly died before birth, however those who survive do not initially show evident clinical or neurological signs; in this case, the newborn has a normal head circumference at birth, without any evident disturbance in archaic reflexes, extremity movements, and sucking-swallowing reflexes. Subtle signs such as feeble crying, feeding difficulty, hypotonia or wide anterior fontanel may be seen at birth and more rapidly pronounced after a few days; consequently progress to severe hypotonia, irritability, and seizure; these signs were not found in this case throughout a week of observation.³

Ultrasonography (US) can detect abnormal features of HE for as early as 21st – 23rd weeks of gestation with impressions of absent cerebral

hemispheres without any intact cortical rim, which is replaced by homogenous echogenic materials (cerebrospinal fluid, necrotic glial tissues and/or ependymal debris) filling the supratentorial space with preserved brain stem (midbrain), thalamus, basal ganglia, choroid plexus and cerebellum in most cases. Cerebral cortex is fully absent in most cases; but there may be partial preservation of frontal, occipital, or both lobes. Our case presented prenatal ultrasonography with impressions of bilateral cerebral cortex absences without any intact cortical rim; while thalami, midbrain structures, cerebellum and other posterior fossa structures were still present. Falx cerebri is still commonly present; however in some reported cases including this case, falx cerebri is shown partially or completely disrupted with unclear anterior or posterior margins.³ Diagnosis of HE should be narrowed down from many other similar differential diagnosis in order to plan proper post-natal management; which includes **[Figure 3]**³:

Hydrocephalus is defined as a progressive enlargement of ventricular system due to inadequate passage of cerebrospinal fluid (CSF) from its production site within cerebral ventricles to its site of absorption into the systemic circulation, resulting in abnormally accelerated head growth.⁸ It is still considered as a common problem in fetus and newborns; which accounts for approximately 2,7 out of 1,000 births globally, in which the incidence of congenital hydrocephalus was approximately 1,2 out of 1,000 births.⁵ classified based on its etiology as congenital/developmental (intrinsic causes) and acquired (extrinsic causes); traditionally it was also classified based on its pathogenesis (obstructive or non-obstructive) and genetics (syndromic or non-syndromic). The pathogenesis of hydrocephalus is the consequence of imbalance in intracranial CSF inflow and outflow, classified into: (1) CSF flow obstruction (non-communicating), (2) impaired CSF absorption (communicating, due to subarachnoid villi inflammation or venous sinus pressure elevation), and (3) CSF overproduction (communicating, due to functional choroid plexus papilloma). Prenatal diagnosis of congenital hydrocephalus is done by ultrasonography (US) with persistent enlargement of posterior horn lateral ventricle width (atrial width) measured 10-15 mm (mild ventriculomegaly), > 15 mm (severe ventriculomegaly); however these findings must also be accompanied by increase in post-

natal head size and tense major fontanelle (due to increase in intracranial pressure). Unlike hydranencephaly, hydrocephalus has increase in head size due to increase in intracranial pressure with intact cortical rim and intracranial structures including the ventricle is still distinguishable.⁶

Holoprosencephaly (HPE) is described as incomplete cleavage of prosencephalon; hence it does not clearly divide in diencephalon, two halves of telencephalon and lateral ventricles; resulting in complete or partial union of forebrain with only a single ventricle instead of two. It is the most common forebrain developmental malformation, affecting 1 out of 250 prenatal fetus and only 1 out of 6000 live births.⁷ HPE is classified into three main types and one variant: alobar, semi-lobar, lobar, and inter-hemispheric average fusion variant. Prenatal diagnosis with ultrasound reveals presence of midline malformation and brainstem anomalies, which are preserved in holoprosencephaly. In alobar HPE, ultrasonography reveals a single primitive ventricle structure with fused thalamus in the midline and complete absence of: inter-hemispheric fissure, corpus callosum, septum pellucidum, neuropophysis, third ventricle, olfactory bulb, and tract. Inter-hemispheric fissure is still partially preserved posteriorly in semi-lobar HPE and in anterior-posteriorly in lobar HPE. Unlike in hydranencephaly, HPE especially in alobar HPE is also associated with facial abnormalities such as cyclopia (one middle eye), proboscis (middle anterior appendage structure), etmocephaly (extremely close ocular distance with proboscis), cebocephaly (close ocular distance with single nostril), anophthalmia/micropthalmia (very small or no eyeballs), cleft lip and absence of nose.⁸

Porencephaly/pseudo-porencephaly is a very rare condition affecting only approximately 5.2 out of 100,000 live births; described as the presence of cystic cavities that usually communicate with ventricular system, subarachnoid space

or both within brain parenchyma.^{2,9} pseudo-porencephaly (encephaloclastic / porencephalic cysts) is different from schizencephaly / true porencephaly in the lining of the cavity, which usually contains white matter instead of gray matter; and is not associated with neuronal migration abnormality. Fetal ischemic stroke and middle cerebral artery occlusion was postulated as the main cause of porencephalic cyst. Prenatal diagnosis with ultrasound reveals cystic cavity(s) within the brain that usually interconnects with ventricular system and located in middle cerebral artery territory accompanied by ischemic infarcts. This condition is hardly differentiated with hydranencephaly in severe cases, usually parieto-occipital cortex is still preserved in porencephaly. Further evaluation with post-natal MRI can be used to precisely differentiate this condition with other similar abnormalities.⁹

Schizencephaly / true porencephaly is a condition characterized by full thickness, gray matter-lined clefts of cerebral mantle; which is often bilateral and symmetric, providing communication between lateral ventricles and external subarachnoid space. It is a very rare condition, which only affects 1 out of 100,000 live births. A genetic/developmental anomaly (familial chromosomal abnormality) is believed to be the cause of schizencephaly.^{2,9} Schizencephaly is further classified into type 1 (fused clefts in cerebral mantle) and type 2 (majority of cases, in which clefts are separated in cerebral mantle with associated ventriculomegaly). Prenatal diagnosis with ultrasound reveals clefts in the area of sylvian fissures, connecting enlarged lateral ventricles medially and subarachnoid space laterally; bilateral frontal horns may be fused with absent septum pellucidum. It is often difficult to differentiate with lobar holoprosencephaly and large porencephalic cysts or intracranial cysts in sylvian fissure area. Further evaluation with post-natal MRI is required to precisely differentiate this condition with other similar abnormalities.⁹

	Hydranencephaly	Hydrocephalus	Holoprosencephaly	Porencephaly	Schizencephaly
Head Circumference	Normal or slightly smaller	Larger	Normal	Normal	Normal
Midline Malformations	Absent	Absent	Present	Absent	Absent
Brainstem Anomalies	Absent	Absent	Present	Absent	Absent
Intact Cortical Rim	Absent	Present	Present	Present	Present
Ventricular System Abnormality	Lateral ventricle structures are irregular and indistinguishable Third ventricle remain intact and not-dilated	Lateral ventricle structures are regular with persistent enlargement Third Ventricle enlargement only present in obstructive forms	Overall abnormal ventricular system, structures are irregular and indistinguishable	Lateral ventricles and subarachnoid space enlargement Frontal horns not fused	Lateral ventricles and subarachnoid space dilatation Frontal horns fused in some cases.
Angiographic Investigation	Bilateral internal carotid artery occlusion (Not Always)	Normal	Normal	Involvement of middle cerebral artery resulting in localized areas of cortical destruction	Normal
Other Fetal Malformations	Absent	Absent	Present	Absent	Absent

Figure 3. Differential Diagnosis of Hydranencephaly

Prospective Management

Post-natal diagnostic evaluation of HE with magnetic resonance imaging (MRI) or computed tomography (CT) is considered as gold standard, which allows precise differentiation of HE with severe congenital hydrocephalus, holoprosencephaly, porencephaly and schizencephaly. Other diagnostic evaluation includes electroencephalogram (EEG), brainstem auditory evoked response test, digital subtraction angiography (DSA) and brain magnetic resonance angiography (MRA). Since our facility (type C hospital) has limited diagnostic resources and technology, only transcranial sonography (TCS) was performed by a pediatrician. Coronal and sagittal plane TCS confirmed the diagnosis of HE by revealing fluid filled cranial cavity with absent most of bilateral cerebral cortex and falx cerebri disruption; however progression was shown, since midbrain structures, cerebellum and other posterior fossa structures were less visible this time. Nevertheless these findings were less reliable in excluding differential diagnosis of HE and the newborn was referred to facilities with appropriate diagnostic technology for further evaluation.^{3,10}

Post-natal management ranges from supportive non-surgical interventions (initial neonatal resuscitation, physiokinestherapy, drugs for epileptic seizure and nutritional interventions) and neurosurgical interventions. Surgical interventions must be considered in

cases with evidence of intracranial hypertension, which include: ventriculo-peritoneal shunting (VP-Shunting), choroid plexus coagulation with endoscopic third ventriculostomy (ECPC), and choroid plexectomy (CP). VP-shunts are commonly associated with complications such as: secondary infection and malposition, which therefore requires regular replacement and re-positioning. ECPC was reported with better success rate of 50-80% when compared to VP shunting, therefore ECPC is considered as a standard treatment for hydranencephaly. Anatomic characteristics that provides advantage for ECPC over VP-Shunting are lack of septum pellucidum (hemispheric separation) and brain parenchyma in hydranencephaly, which allows easy access to bilateral choroid plexus. Arachnoid collapse is a complication commonly associated with ECPC, which occurs when the opened dura liberates intracranial pressure, consequently lead to difficulty in coagulating the choroid plexuses. A few neurosurgeons have reported that repeated complex surgeries such as choroid plexectomy (CP) may be useful.^{3,10,11}

In this case, the newborn patient was observed in NICU for a week with unremarkable progression in clinical status. The newborn was later referred to facilities with neurosurgery department to further evaluation and surgical intervention if indicated. Prognosis determination and proper post-natal management, which will consequently improve the newborn’s life expectancy, are achievable through precise diagnosis of HE and exclusion of differential diagnosis.^{3,10,11}

CONCLUSION

Early prenatal recognition of hydranencephaly and exclusion of similar differential diagnosis, which includes: hydrocephalus, holoprosencephaly, porencephaly and schizencephaly, are fundamental in formulating proper multidisciplinary management with pediatric and neurosurgery department, which may consequently improve the newborn's life expectancy.

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