INTRODUCTION

Hydranencephaly is a rare, isolated abnormality occurring in less than 1 in 10,000 births worldwide. It is the most severe form of bilateral cerebral cortical destruction (1, 16).

Hydranencephaly integrates a group of brain malformations that vary in time of onset, pathogenesis and cerebral organization of any remnant persists, resulting in a cerebral parenchymal cavity does not communicate with the subarachnoid space, a common effect of markedly reduced mass cerebral spinal fluid is replaced with (1). This defect usually occurs in the uterus by a destructive process in midgestation, after formation of the telencephalon. (1.5)

Its etiology has been attributed to multiple causes that lead to ischemic injury, viral infections are the main involved, such as herpes viruses, paroviruses, cytomegalovirus, and others (1.11); Another possible toxic etiology are processes (alcohol) (10) or such genetic Fowler syndrome (12).

Generally facial features are normal, which differs with other malformations of the central nervous system. The focal motor deficit depends on the location and extent of injury, being contralateral.

Pathophysiologically is considered that these etiological agents originate previous impaired cerebral circulation and necrotic destruction of brain mass and subsequent cavitation. Necrosis of germinal center plates and infiltration of inflammatory cells nervous system, especially macrophages located. (13)

Experimentally it was found that the prevalence of hydranencephaly dependent agent and the maternal immune status. (11, 12)

The pathological findings are related to survival, when the anatomic relationships and histologically preserved structure and connections are preserved, survival is greater. Thalamic atrophy and hypoplasia of the cerebral peduncles with absence of the medullary pyramids have lower survival. (13)
Hydranencephaly not presented after hydrocephalus, while it may be initiated as multicystic encephalopathy, and progress hydrancephaly since in this there is also a severe vascular injury background. (3, 5)

Ultrasound plays a fundamental role in prenatal diagnosis and now with improved neuroimaging, MRI and cerebral magnetic resonance angiography, more cases of hydranencephaly described, differentiating severe hydrocephalus and other brain malformations that present with cavities in the brain. The long-term survival of these patients is lower than in the general population, and many of the fetuses are deaths or die within days or weeks after birth. (2, 3.16)

In the postnatal period there is a progressive growth in head circumference, with significant neurodevelopmental disorders. (6.8)

MATERIALS AND METHODS

Obstetrics Gynecology Hospital Isidro Ayora (HGOIA). Patient aged 27, born and lives in Quito, with no personal or family history of relevance. Segundigesta, interpregnancy period of 4 years, first feat with normal prenatal, natal and postnatal period. Current Gesta presents with 30 weeks gestation, with two ultrasound scans at 12 and 20 weeks with normal result.

Saote ultrasound equipment brand, with convex transducer, CA Using harmonic 3.5 MHz, Color Doppler, Power Doppler, spectral Doppler, with image enhancement using low frequency, high densities, adequate number of foci according to needs.

GE Voluson 730 ProV brand, with 3.5 MHz convex transducer Using 3D.
Philips 1.5T Magnetic Resonance Imaging: Magnetic Resonance Imaging (MRI) Philips 1.5 Tesla (T). Axial, coronal and sagittal sections were made. T1 and T2 ultrafast sequences, radial MR cholangiopancreatography sequence were used. Body coil and placed further on the pelvis. MRI study the patient had a six-hour fasting.

RESULTS

Obstetric ultrasound Doppler.

Ultrasound examination shows live singleton fetus, cephalic, anterior dorsum, fetal movements present.

Morphologically level head in the supratentorial region occupying almost entirely by cerebrospinal fluid, identifying part of the cerebral mantle along the inner table in the bilateral parieto occipital region.

Falx is identified.

The thalamus and posterior fossa structures are normal morphology and echotexture. Doppler studies in cerebral vascular structures that correspond to the subsequent movement is observed. In bilateral cervical internal carotid artery is identified.

According to the values obtained in the fetal biometry pregnancy corresponds to 30.4 weeks gestation.

3D images corroborate the normal fetal morphology.

The analysis demonstrates IUGR biometric indexes.
Obstetric MRI showed better anatomical detail with morphological findings described in ultrasound. Fetal - No other abnormalities were identified maternally.

DISCUSSION:

Hydranencephaly is one of the pathologies of the central nervous system more complicated and poor prognosis. It results from a variety of congenital or destructive lesions that produce a defect in brain development, since the end of the third trimester of pregnancy to 2 years of age abnormalities. (4, 7, 14)

The exact etiology of hydranencephaly not known, although it is hypothesized that it is due to occlusions supraclinoid artful bilateral internal carotid with intact posterior circulation. (4, 14)

Most cases have originated in utero and postnatal few are described appearance (3).
The differential diagnosis has to be performed with a high call hydrocephalus, better prognosis, in which a small cortical mantle is identified in the carotid territory and others with more severe neurological involvement such as alobular holoprosencephaly, severe bilateral schizencephaly parted lips and cystic encephalomalacia. (1.3)

It is important to further investigate the pathophysiology, diagnosis, prevention, treatment, prognosis and survival, to provide a better quality of life for these young

CONCLUSIONS

Ultrasound remains the technique of choice for the study of fetal morphology although MRI can help in cases of poor ultrasound visualization (obesity, oligohydramnios) with suspected malformation pathology.

Ultrasound remains a method of initial study of fetal malformations.

The doppler study in these patients is essential for the diagnosis of this pathology.

MRI not only confirms the findings of ultrasonography but allows others to discover previously undetected by ultrasound and also dismiss breast pathology. Allows good visualization of the fetal anatomy, especially CNS, thorax and abdomen.

The diagnosis of hydranencephaly, given its rarity and its implications, has to be done with extreme care, and have to take appropriate precautions in anticipation of an eventual failure of prenatal diagnosis.

THERE IS NO CONFLICT OF INTEREST.

Bibliography


