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DANDY-WALKER SYNDROME:antenatal diagnosis: About a case and review of the literature.

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***Abstract**: Dandy-Walker syndrome (DWS) is a rare cerebral malformation that can be isolated or associated with other malformations. The diagnosis can be made antenatally or postnatally, and the prognosis is related to the importance of the underlying hydrocephalus and to the frequent malformative associations. Through an observation and review of the literature, we will try to establish the contribution of obstetrical ultrasound and fetal MRI in the antenatal diagnosis of Dandy-Walker syndrome, and the role of the derivation methods in the symptomatic management.*

Keywords— Hydrocephalus, Dandy-Walker Variant, Dandy-Walker Malformation, Vermis Agenesis

1. INTRODUCTION

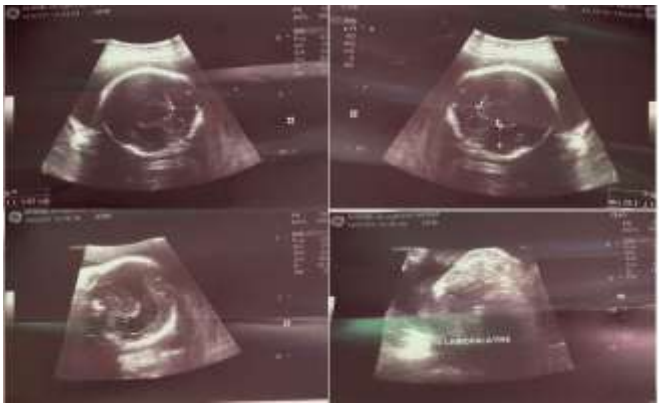
Dandy-Walker syndrome (DWS) is a rare cerebral malformation that can be isolated or associated with other malformations. The diagnosis can be made antenatally or postnatally, and the prognosis is related to the importance of the underlying hydrocephalus and to the frequent malformative associations. We propose, through this observation, as well as a review of the literature, the contribution of obstetrical ultrasound and fetal MRI in the antenatal diagnosis of Dandy-Walker syndrome, and the role of the derivation methods in the symptomatic management.

2. CASE REPORT :

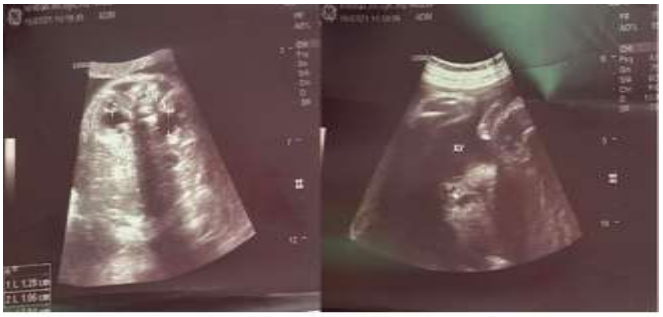
this is a 26 year old patient, with no particular pathological antecedents, G2P1, not followed up, referred to our prenatal diagnostic unit at 36 SA for hydrocephalus with posterior cerebral fossa anomalies. Obstetrical ultrasound: objective of a progressive eutrophic pregnancy with quadri-ventricular hydrocephalus with lateral ventricle (LV) at 18mm, with posterior cerebral fossa (PCF) anomalies such as vermian agenesia, anteroposterior enlargement with PCF cyst, cerebellar tent overexcavation, cleft lip and palate with bilateral pyelectasia and micropenis.
MRI was not done antenatally.

The surveillance was regular with a good evolution until 39 SA + 2 days when the patient gave birth to a male newborn, Apgar 10/10 with a birth weight of 3600g, presenting a cleft lip and palate without any other particular signs. The ETF : objectify a moderate quadri-ventricular hydrocephalus The decision of the pediatric surgeons was a clinical monitoring without immediate derivation.

The newborn died at D+3 of life in a context of neonatal respiratory distress.



ultrasound at 36 weeks of amenorrhea showing hydrocephalus quadri-ventricular hydrocephalus with lateral ventricle (LV) at 18mm, with posterior cerebral fossa (PCF) anomalies such as vermian agenesia, anteroposterior enlargement with PCF cyst



ultrasound at 36 weeks of amenorrhea showing bilateral pyelectasis and micropenis

3. DISCUSSION

Dandy-Walker syndrome (DWS) or Dandy-Walker malformation (DWM) is defined by the association of cystic dilatation of the fourth ventricle, complete or partial agenesis of the vermis and hypoplasia of the cerebellar hemispheres responsible for distension of the posterior cerebral fossa with ascension of the cerebellar tent with or without hydrocephalus.
The first autopsy description was reported in 1887 by Sutton, (1) and it was not until 1914 that Dandy and Blackfan (2)

studied an association between hydrocephalus and cystic dilatation of the fourth ventricle. The malformation was further characterized by Dandy (3) in 1921 and by Taggart and Walker (4) in 1942 as being related to congenital atresia of the foramen of the fourth ventricle. But it was Benda (5) in a series of autopsies in 1954 who first used the "Dandy-Walker syndrome" to describe this malformation where he proposed a new theory of etiopathogeny, He proposed a new theory on the etiopathogeny, assuming that the failure of normal regressive changes in the posterior medullary velum and the absence of the cerebellar vermis lead to the formation of cysts from the distal extremity of the fourth ventricle that separates the two cerebellar hemispheres.

MDW is a well-known malformation that has been associated with central nervous system abnormalities, genetic abnormalities, environmental factors, teratogens and congenital infections. (6) It accounts for 14% of posterior cerebral fossa cystic formations and affects 1/30,000 births, and is responsible for 40% of deaths in the first year of life. Psychomotor retardation is common in affected children, and correlates with the degree of vermis involvement and associated malformations or genetic abnormalities. (7)

Associated malformations are frequent with cardiac anomalies (38%), facial dysmorphia and cleft palate (26%), dysraphia, poly- and syndactyly (28%), genitourinary malformations (28%) and digestive malformations. (7) Chromosomal abnormalities are also frequent, reaching 50% in some series, and can be detected on a standard karyotype (triploidy, trisomies 13 and 18...) 834, but also in molecular biology, MLPA, CGH-array (deletion on chromosome 3, 6...). It is also present in many genetic or sporadic polymalformative syndromes. The modes of inheritance may be X-linked or autosomal recessive, as indicated in rare familial cases. If SDW is associated with a single disorder hereditary genetic disorder, the risk of recurrence may be high for subsequent pregnancies. However, this risk is generally estimated at 1% to 10% in sporadic cases. (1-3)

Accurate antenatal diagnosis is essential not only for the management of the current pregnancy, but also for prenatal genetic counseling in future pregnancies.

Dandy-Walker malformation (DWM) has been described by Spennato et al (8) and Klein et al (9) as requiring six radiological criteria necessary for diagnosis:

1. large posterior cerebral fossa cyst communicating widely with the fourth ventricle.
2. absence of part of the inferior vermis.
3. Hypoplasia, anterior rotation and upward displacement of the remaining vermis.
4. Absence or flattening of the angle of the cerebellar fastigium.
5. Large posterior fossa with torcular elevation.
6. anterolateral displacement of the cerebellar hemispheres. (8,9)

Dandy-Walker variant (DWV) is a currently abandoned term, and was first introduced by Harwood-Nash and Fitz to describe less severe presentations of SDW that did not conform to the classic definition, but a clear clinical separation has not been defined. DWV consists of a defect in the inferior cerebellar vermis and a communication between a large cistern of normal size and the fourth ventricle.

Dandy-Walker Complex (DWC) is another term coined to describe a continuum of posterior fossa anomalies classified as mild, moderate, and severe.

The Dandy-Walker Complex (DWC) is another term coined to describe a continuum of posterior fossa anomalies ranging from mild (mega large cistern only) to moderate (mild vermian hypoplasia, enlarged fourth ventricle) to severe (agenesis of the vermis, cystic dilatation of the posterior fossa and fourth ventricle). (10)

Obstetrical ultrasound can confirm the diagnosis by looking for the following signs - In the posterior cerebral fossa:

In cavo-thalamo-cerebellar section, an enlargement of the PCF by the cystic dilatation of the V4, anechoic,triangular with external base. The hypoechogenic cerebellar hemispheres are laterally and anteriorly compressed, and are more or less hypotrophic, symmetrically or not. The vermis, echogenic, is hypotrophic or even absent (25%).

In sagittal section, the tent of the cerebellum is elevated, the vermis presents a more or less important amputation of its inferior part. The inferior part of the vermis may extend towards the torcular in a linear "comet tail" image corresponding embryologically to the upper wall of Blake's pouch. The brain stem is normal.

At the supratentorial level, abnormalities are found in 70% of cases: hydrocephalus (60%), agenesis of the corpus callosum (15%), migration abnormalities, interhemispheric lipomas and cysts, cephalocele, holoprosencephaly. (7)

Fetal MRI is essential for the differential diagnosis of various cystic malformations of the PCF including arachnoid cyst, Blake's pouch cyst, cystic dilatations of the V4 resulting from other types of vermian and/or cerebellar anomalies grouped under the label of "DW variant"; Thus, any pressurized median fluid collection developed under the vermis, such as neuroepithelial cysts, some arachnoid cysts, and some collections described as "mega-large cisterns", may mimic MDW by overlying the vermis, opening Magendie's orifice, and even overlying the cerebellar tent. Diagnosis requires medial sagittal sections of the posterior fossa, allowing fine analysis of the vermis.

Cystic malformations of the posterior cerebral fossa constitute a broad spectrum whose differential diagnosis is essential in prenatal screening. These include DWV, mega-large cistern, posterior fossa arachnoid cyst, Blake's pouch cyst, fourth ventriculocele, and congenital vermian hypoplasias(10-11).

The incidence of WDS is 1 in 25,000 to 30,000 newborns (12) with a predominance in the first year of life with hydrocephalus (often before 3 months of age). (12-13) However, the diagnosis can be delayed until

However, diagnosis can be delayed until adolescence or even adulthood in less severe cases.

Numerous associated malformations are described in the literature. The incidence of associated central nervous

system (CNS) abnormalities is variable (up to 68%), with systemic abnormalities present in approximately one quarter of patients. (14) The most common CNS abnormality is agenesis of the corpus callosum. The other most common anomalies are capillary hemangiomas and cardiac malformations. (15)

Currently, CSF drainage via shunts is the first-line treatment for SDW. However, significant controversy exists regarding which procedure gives the best results. Options include supratentorial compartment shunting, cerebellar cyst shunting, and double shunting.

Regardless of the treatment modality, the primary goal is to maximize functional survival of patients while limiting morbidity and mortality.

Diagnosis, mainly obstetric ultrasound and fetal MRI, in combination with cytogenetic characterization of SDW may allow a better prognosis for affected newborns in the future.For treated hydrocephalus, prognosis depends mainly on the presence of associated conditions. Seizures, hearing or visual disturbances, and other systemic or CNS abnormalities are predictive of poor prognosis. (16) In patients without other abnormalities, some studies have reported intelligence quotients (IQ) of 80 or higher in 50% of survivors. (9,16,17) Studies have concluded that the quality of vermilion lobulation may be a determining prognostic factor for functional development.

4. CONCLISION

Dandy-Walker syndrome is a relatively rare cause of hydrocephalus and may be accompanied by multiple congenital CNS and other systemic abnormalities.

The severity of Dandy-Walker syndromes is variable and they do not present any pathognomonic clinical syndrome. The different surgical methods are symptomatic and allow to manage symptomatic hydrocephalus and to improve the functional prognosis of the children, which depends essentially on the associated anomalies.

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