wjpmr, 2023, 9(4), 17-20

SJIF Impact Factor: 5.922

WORLD JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.wjpmr.com

Case Study

ISSN 2455-3301

WJPMR

A CASE REPORT OF A DANDY WALKER MALFORMATION IN ONE TWIN

Aicha Bennani*[1&2], Oumaima Sahraoui[1&2], Hanaa Lazhar[1&2], Aziz Slaoui[1&2], Najia Zeraidi[1], Amina Lakhdar[1], Aicha Kharbach[2] and Aziz Baydada[1]

[1]Gynaecology-Obstetrics and Endoscopy Department, Maternity Souissi, University Hospital Center IBN SINA, University Mohammed V, Rabat, Morocco.

[2]Gynaecology-Obstetrics and Endocrinology Department, Maternity Souissi, University Hospital Center IBN SINA, University Mohammed V, Rabat, Morocco.

***Corresponding Author: Aicha Bennani**

Gynaecology-Obstetrics and Endoscopy Department, Maternity Souissi, University Hospital Center IBN SINA, University Mohammed V, Rabat, Morocco.

Article Received on 27/01/2023 **Article Revised on 16/02/2023** **Article Accepted on 08/03/2023**

ABSTRACT

The dandy walker malformation is estimated to be 1 per 10 000 births. It can be isolated or associated with other central nervous system malformations or systemic anomalies. Most cases of DW malformation are diagnosed prenatally. There are neuroimaging diagnostic criteria that make it possible to distinguish the dandy walker malformation from other cystic posterior fossa malformations on ultrasound and MRI. The dandy walker malformations have various aetiologies and diverse outcomes. In our case, a patient with twin pregnancy at 32 weeks of gestation. She was admitted to the emergency room for preterm labour. The ultrasound revealed a dandy walker syndrome and uterine growth restriction in one twin in a monochorionic diamniotic pregnancy.

INTRODUCTION

The dandy walker malformation is developmental anomaly of the cerebellum with a complete or partial agenesis of the cerebellar vermis, cystic dilatation of the fourth ventricle and elevation of the roof of the posterior fossa. It has many etiology and various neonatal outcomes. It can be diagnosed prenatally. We report a case of twin pregnancy with one foetus presenting a dandy walker malformation and intrauterine growth retardation. It was confirmed postnatally by MRI.

CASE REPORT

A 32 years old pregnant patient, primigravida, was admitted to the emergency room for preterm uterine contractions at 32 weeks of gestation.

She had no previous history. The physical exam found a cervix dilated to 2 cm and 80 percent effaced with intact membrane. The patient had only one ultrasound during this pregnancy which revealed a monochorionic diamniotic twin pregnancy, in the first trimester.

We performed an ultrasound which revealed two foetuses with growth discordance. J1's weight was estimated at 2100 grams (80th percentile) and J2's weight was estimated at 1400 (3rd percentile). Twin-to-twin transfusion syndrome was ruled out.

In the axial plane, J2 had an enlargement of the cisterna magna (24 mm) and a communication of the fourth ventricle and the cisterna magna, trapezoid-shaped gap between the cerebellar hemispheres as seen in the picture below.

These antenatal sonographic features suggested the diagnosis of dandy walker malformation. The patient was administered a calcium channel and a first dose of steroids for fetal lung maturation. But, shortly after, the twins were delivered. An MRI performed postnatally confirmed the diagnosis of dandy walker malformation. The twins are still hospitalised in the NICU for pretermaturity and exploration of the dandy walker malformation of the second twin.

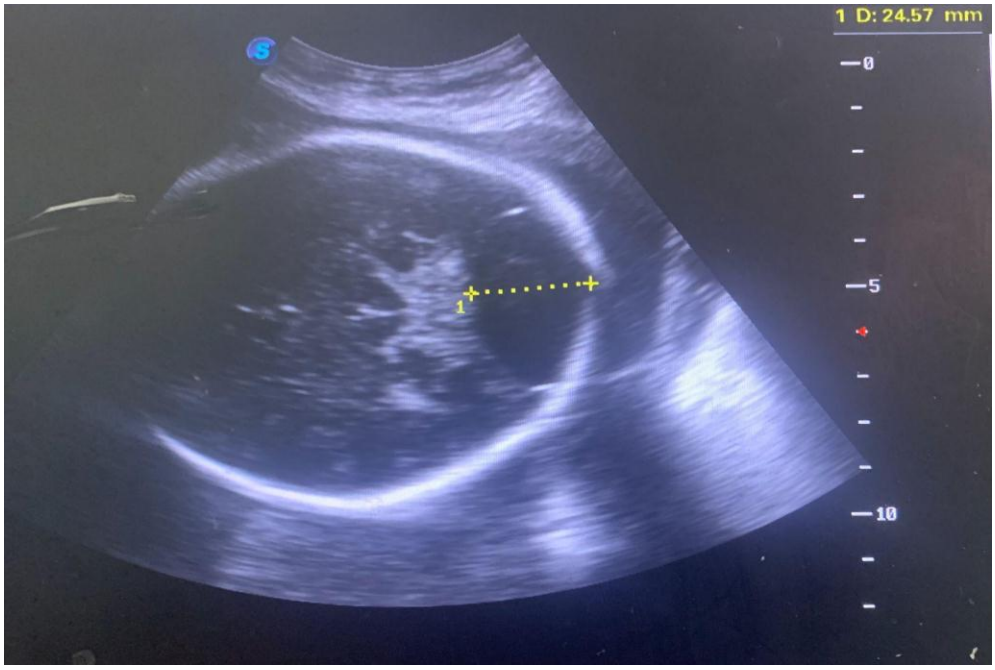

Figure 1: axial plane at the level of the cerebellar plane showing an enlarged cisterna magna 24 mm.

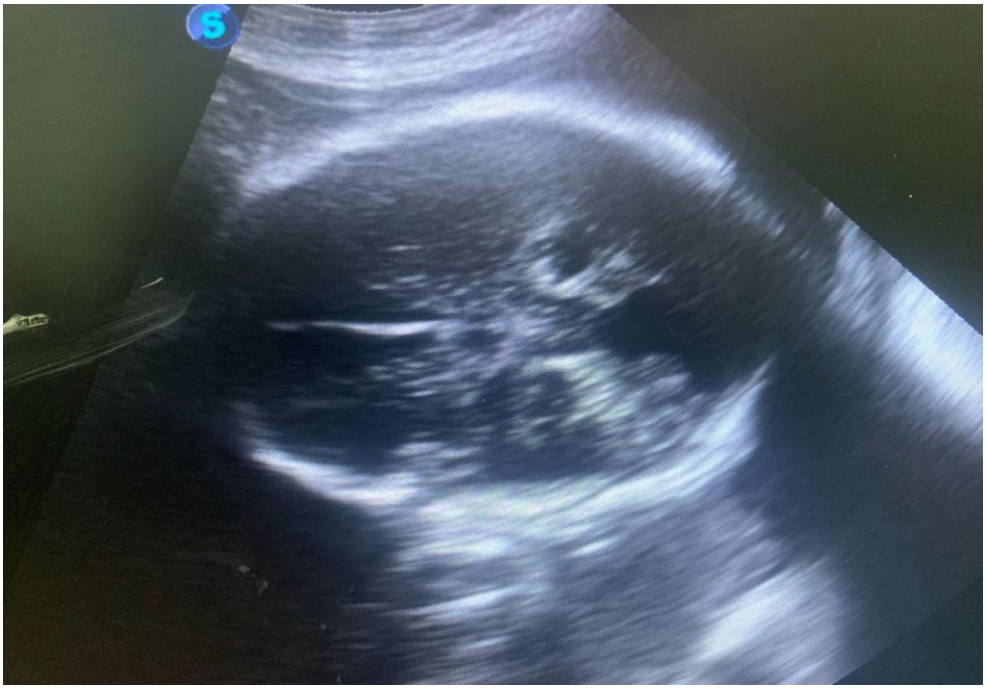

Figure 2: axial plane showing the communication between the fourth ventricle and the cisterna magna on ultrasound.

DISCUSSION

Dandy walker is posterior fossa malformation and is defined.

- complete or partial agenesis of the vermis;
- enlargement of the posterior fossa with the upward displacement of the tentorium, transverse sinus, and torcular;
- and cystic dilation of the fourth ventricle[1]

The prevalence of DWS has been estimated to be 1 per 10 000 births. It accounts for 14% of cystic posterior fossa malformations and 2–4% of hydrocephalus.[1,2]

The dandy walker malformation typically occurs sporadically with a low risk of recurrence.[3] It can be isolated or associated with other central nervous system malformations or systemic anomalies especially congenital heart disease, cleft or palate lip and neural tube defects.[3] Other systemic anomalies like foetal growth restriction, limb and abdominal wall

abnormalities, diaphragmatic hernia, ambiguous genitalia have been reported.[4]

The dandy walker malformation has been associated with mendelian disorders such as Joubert syndrome, Meckel–Gruber' syndrome and Aicardi's syndrome. Other reports associated the dandy walker malformation with chromosomal aberrations including duplications. The most frequent chromosomal anomalies were trisomy 18 (Edward's syndrome) and trisomy 13 (Patau's Syndrome).[5]

Most genes identified in Aldinger et al.'s discovery cohort were previously associated with known neurodevelopmental disorders including epilepsy and autism.[6]

Liao et al. presented a case report with three foetuses with a de novo adjacent microdeletion/duplication region mapping to chromosome 7p21.3, and suggest that the critical region associated with Dandy Walker malformation may be limited to the 7p21.3 region.[7]

Cerebellar abnormalities have also been linked to non-genetic mechanisms, especially prematurity, twinning, and prenatal cerebellar haemorrhage. It includes DWM in both monozygotic and dizygotic twins, which suggests that factors other than TTTS may be involved.[5] Also environmental agents such as rubella, cytomegalovirus, toxoplasmosis, coumadin, alcohol, and maternal diabetes.[8]

On ultrasound, In the axial plane at the level of the transcerebellar plane, the cisterna magna is enlarged (>10 mm). The cerebellar hemispheres are splayed apart, and the vermis is absent or hypoplastic. The presence of a connection between the fourth ventricle and the cisterna magna after 20 weeks of gestation is indicative of DWM.

Mediane plane, a small vermis that lacks fastigium, fissure suggests partial vermian agenesis. Brainstem-vermis angle >45 degrees strongly suggest DWM.
This helps differentiate DWM from other posterior fossa abnormalities.[1,8]

Most cases of DW malformation are diagnosed prenatally.[5]

A correct diagnosis cannot be made without a good quality MRI including sagittal views of the vermis and T2-weighted images. The following neuroimaging diagnostic criteria usually make it possible to distinguish DWM from other cystic posterior fossa malformations: large median posterior fossa cyst widely communicating with the fourth ventricle, a small, rotated, raised cerebellar vermis, an upwardly displaced tentorium, an enlarged posterior fossa, antero-laterally displaced but apparently normal cerebellar hemispheres, a normal brain stem.[9]

The prognosis following prenatal diagnosis of DWM is variable and it is generally worse when associated with other anomalies. If it is isolated with partial agenesis of the vermis then it is generally compatible with normal life, otherwise it is associated with mental retardation.[10]

CONCLUSION

Dandy walker is a rare malformation of the posterior fossa. It can be diagnosed prenatally by ultrasound and or MRI. Prognosis of DWM depends on the underlying aetiology and whether associated conditions are present.

REFERENCE

1. Society for Maternal-Foetal Medicine (SMFM); Monteagudo A. Dandy-Walker Malformation. Am J Obstet Gynecol, 2020 Dec; 223(6): B38-B41. doi: 10.1016/j.ajog.2020.08.184. Epub 2020 Nov 7. PMID: 33168220.
2. Kollias, S.S. (2003). Dandy–Walker syndrome.
3. Lerman-Sagie, T et al. 2018. Chapter 1 - Fetal cerebellar disorders. In M. Manto & T. A.G.M. Huisman (Eds.), The Cerebellum: Disorders and Treatment (pp.3-23). Elsevier. https://doi.org/10.1016/B978-0-444-64189-2.00001-9.
4. Harper T, Fordham LA, Wolfe HM. The fetal dandy walker complex: associated anomalies, perinatal outcome and postnatal imaging. Fetal Diagn Ther, 2007; 22(4): 277-81. doi: 10.1159/000100790. PMID: 17369695.
5. Santoro M, Coi A, Barišić I, Garne E, Addor MC, Bergman JEH, Bianchi F, Boban L, Braz P, Cavero-Carbonell C, Gatt M, Haeusler M, Kinsner-Ovaskainen A, Klungsøyr K, Kurinczuk JJ, Lelong N, Luyt K, Materna-Kiryluk A, Mokoroa O, Mullaney C, Nelen V, Neville AJ, O'Mahony MT, Perthus I, Randrianaivo H, Rankin J, Rissmann A, Rouget F, Schaub B, Tucker D, Wellesley D, Yevtushok L, Pierini A. Epidemiology of Dandy-Walker Malformation in Europe: A EUROCAT Population-Based Registry Study. Neuroepidemiology, 2019; 53(3-4): 169-179. doi: 10.1159/000501238. Epub 2019 Jul 12. PMID: 31302658.
6. Aldinger KA, Timms AE, Thomson Z, Mirzaa GM, Bennett JT, Rosenberg AB, Roco CM, Hirano M, Abidi F, Haldipur P, Cheng CV, Collins S, Park K, Zeiger J, Overmann LM, Alkuraya FS, Biesecker LG, Braddock SR, Cathey S, Cho MT, Chung BHY, Everman DB, Zarate YA, Jones JR, Schwartz CE, Goldstein A, Hopkin RJ, Krantz ID, Ladda RL, Leppig KA, McGillivray BC, Sell S, Wusik K, Gleeson JG, Nickerson DA, Bamshad MJ, Gerrelli D, Lisgo SN, Seelig G, Ishak GE, Barkovich AJ, Curry CJ, Glass IA, Millen KJ, Doherty D, Dobyns WB. Redefining the Etiologic Landscape of Cerebellar Malformations. Am J Hum Genet, 2019 Sep 5; 105(3): 606-615. doi:

10.1016/j.ajhg.2019.07.019. Epub 2019 Aug 29. PMID: 31474318.

7. Liao C, Fu F, Li R, Yang X, Xu Q, Li DZ. Prenatal diagnosis and molecular characterization of a novel locus for Dandy-Walker malformation on chromosome 7p21.3. Eur J Med Genet, 2012 Aug-Sep; 55(8-9): 472-5. doi: 10.1016/j.ejmg.2012.04.008. Epub 2012 May 19. PMID: 22617776.
8. Volpe P, Contro E, De Musso F, et al. Brainstem-vermis and brainstem tentorium angles allow accurate categorization of fetal upward rotation of cerebellar vermis. Ultrasound Obstet Gynecol, 2012; 39: 632–5.
9. Bosemani T, Orman G, Boltshauser E, Tekes A, Huisman TA, Poretti A. Congenital abnormalities of the posterior fossa. Radiographics, 2015 Jan-Feb; 35(1): 200-20. doi: 10.1148/rg.351140038. PMID: 25590398.
10. Klein O, Pierre-Kahn A, Boddaert N, Parisot D, Brunelle F. Dandy-Walker malformation: prenatal diagnosis and prognosis. Childs Nerv Syst, 2003 Aug; 19(7-8): 484-9. doi: 10.1007/s00381-003-0782-5. Epub 2003 Jul 16. PMID: 12879343.