International Journal of Surgery Case Reports 115 (2024) 109037

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Radiological characteristics of the posterior fossa of the fetal skull and presentation of a rare case of antenatal screening for Dandy-Walker malformation using antenatal fetal ultrasound and MRI

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ARTICLE INFO

Keywords:
Dandy Walker
Cystic malformation
Posterior cerebral malformation
Antenatal diagnosis
Case report

ABSTRACT

Introduction and importance: Dandy-Walker malformation is a rare congenital anomaly of the brain that mainly affects the cerebellum region. It is characterised by abnormal dilatation of the fourth ventricle of the brain and partial or total absence of the cerebellar vermis. This malformation may also be accompanied by other anomalies of the brain. Ante-natal diagnosis is becoming increasingly frequent given the performance of medical imaging, in particular ante-natal ultrasound and MRI. The object of this article is to clarify the possible causes of rare cystic malformations of the posterior cerebral fossa, which are very rare congenital malformations.

Case presentation: a 30 year old patient, second gesture, mother of a live child by caesarean section, referred to us at 32 weeks of amenorrhoea at the university hospital centre for management of a cystic malformation of the posterior cerebral malformation detected on 2nd trimester ultrasound and confirmed as a Dandy Walker malformation on 3rd trimester fetal MRI.

Clinical discussion: The Dandy-Walker malformation can be described on prenatal MRI as vermian hypoplasia and can be detected as early as the 1st trimester of pregnancy using ultrasound, This cystic malformation poses a problem of differential diagnosis with other pathologies which also result in a cystic image of the posterior cerebral fossa, in particular Black's pouch cyst, arachnoid cyst and mega magna cistern, which requires careful interpretation of cerebral MRI of the foetus.

Conclusion: Imaging techniques play a fundamental role in diagnosis. Prenatal ultrasound and MRI can reveal a Dandy-Walker malformation as early as the 2nd month of pregnancy. MRI is ideal for differentiating differential diagnoses.

1. Introduction

The formation of the posterior cranial fossa and its contents occurs during the process of ventral induction during embryogenesis. Abnormalities and defects occurring during this development give rise to congenital malformations of the posterior fossa, including the brainstem, cerebellum and skull. Cystic malformations of the posterior cranial fossa encompass a range of disorders, including Dandy-Walker malformation, hypoplasia of the cerebellar vermis, mega cisterna magna and arachnoid cyst. Antenatal diagnosis of Dandy-Walker malformation is generally made by medical imaging, in particular antenatal ultrasound screening, followed by antenatal MRI of the foetus to confirm the diagnosis. Dandy-Walker malformation is generally easily diagnosed thanks to the classic triad characterised by total or partial agenesis of the vermis, cystic dilatation of the fourth ventricle and hypertrophy of the posterior cranial fossa. Hypoplasia of the cerebellar vermis is a general category encompassing congenital malformations with normal size of the posterior cranial fossa, varying degrees of hypoplasia of the vermis and cerebellum, and a prominent retro-cerebellar space in free communication with a normal or enlarged fourth ventricle. Careful analysis of embryonic development is essential for understanding these malformations and for more accurate antenatal radiological diagnosis [1]. This case report has been reported per the SCARE 2020 criteria [2].

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https://doi.org/10.1016/j.ijscr.2023.109037
Received 4 October 2023; Received in revised form 6 November 2023; Accepted 8 November 2023
Available online 20 November 2023

2. Case report

She is a 30-year-old second-time mother of a live-born child by Caesarean section, with no particular pathological history, no genetic disease, no history of congenital neurological disease in the family, no particular medication taken, no known mental or physical illness, no smoker, no alcoholic and never operated on for any disease, followed for her pregnancy since the first trimester and referred to us at 32 weeks' amenorrhoea at the university hospital centre for management of a cystic malformation of the posterior cerebral malformation suggestive a priori of a Dandy Walker malformation, with a second-trimester ultrasound if attached: (Fig. 1) which shows a cystic formation of the posterior cerebral malformation with probable agenesis of the vermis, to be completed by fetal MRI to ensure vermis and to eliminate differential diagnoses.

On fetal MRI, sequence 3 D T2 FASTE (Fig. 2), we note the presence of a medial retro cerebellar cystic formation communicating with the fourth ventricle measuring 52 × 34, 6 mm, which appears to exert a scalloping effect on the occipital spine with assent of the cerebellar tent with no dialectation of the upstream ventricular system, no abnormality of the cerebral parenchyma with agenesis of the vermis and the medial septa are in place. This aspect is in favour of a Dandy Walker malformation and the patient maintained the pregnancy until term and then extracted the foetus by caesarean section. Post-operative follow-up was straightforward; the patient resumed intestinal transit on the same day and was discharged 48 h after hospitalisation.

3. Discussion

3.1. Embryogenesis

The development of the nervous system is part of embryonic development and can be broken down into three essential stages: neuronal induction, neurulation and vesicle formation. The different phases of brain development include gastrulation, dorsal induction, ventral induction, neuronal proliferation, differentiation, histogenesis, neuronal migration and axonal myelination. These processes are carefully regulated, both before and after birth, by a complex interaction between genetic and cellular factors, culminating in the complete formation of the nervous system (Fig. 3) [1].

3.2. Radiological imaging appearance

The diagnosis of Dandy-Walker malformation can be made sonographically as early as the first trimester ultrasound between 11 weeks + 6 days and 13 weeks + 6 days of pregnancy, which shows an enlarged

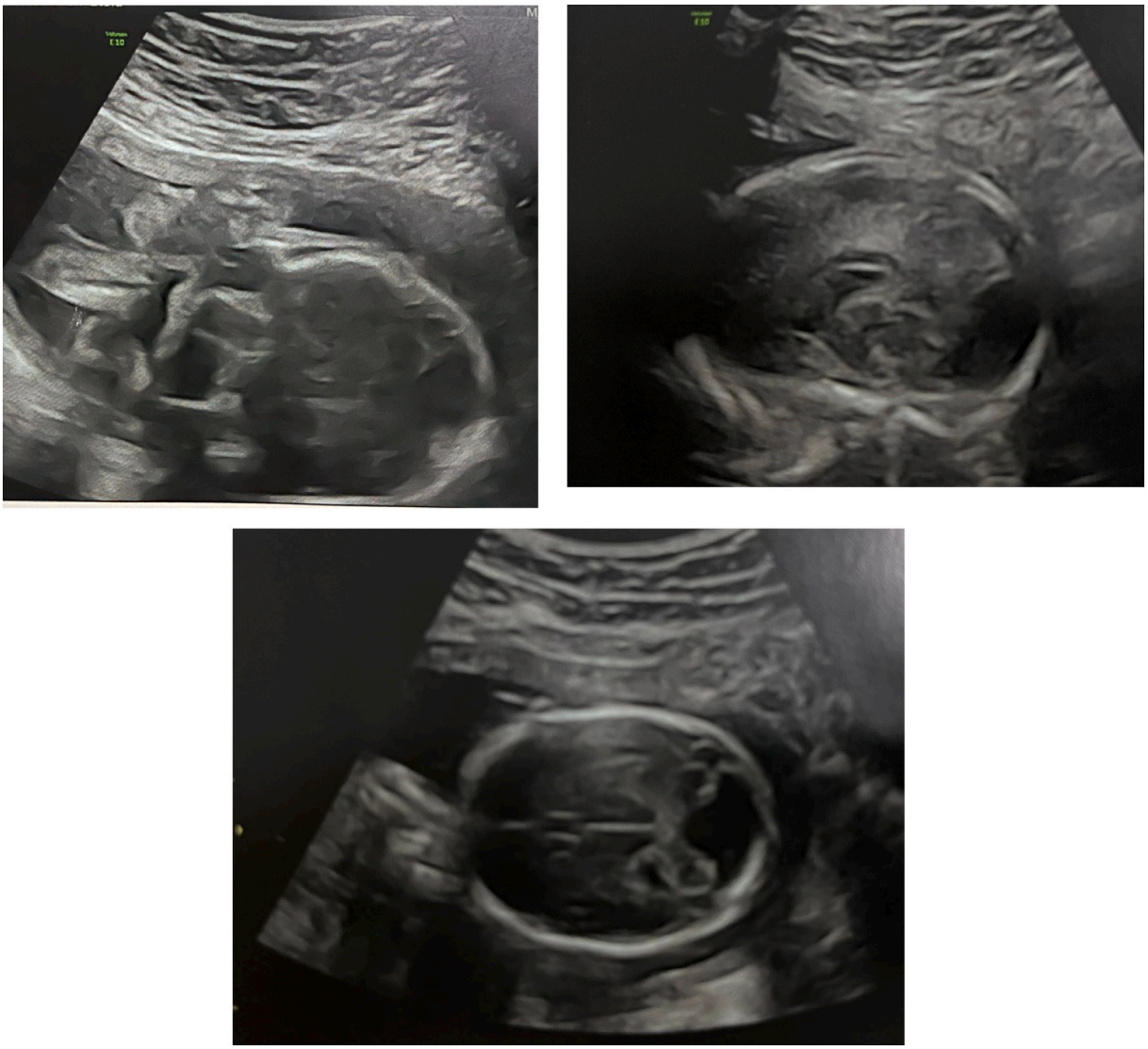

Fig. 1. Second trimester ultrasound shows cystic formation of posterior cerebral malformation.

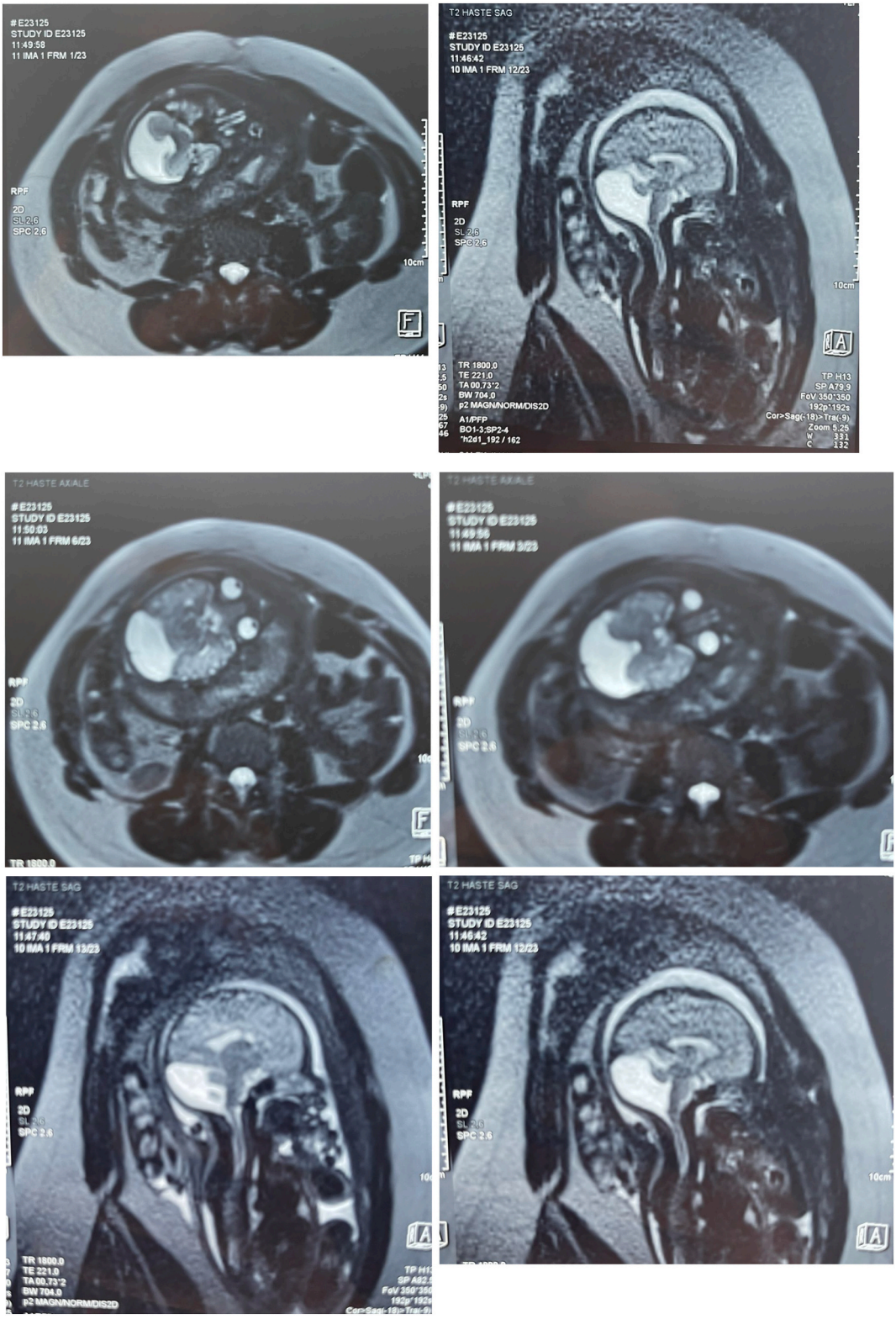

Fig. 2. Fetal MRI sequence 3 D T2 FASTE showing the presence of a medial retro cerebellar cystic formation in favour of Dandy Walker malformation.

posterior cranial fossa and the absence of the cerebellar vermis. According to a study carried out in 200 by Ecker JL on 50 foetuses with Dandy Walker malformation, 85 % of foetuses with DW had other anomalies identifiable by ultrasound, the most common being ventriculomegaly in 32 % and cardiac malformations in 38 %. Karyotypes were abnormal in 46 % of cases [3].

The Dandy-Walker malformation can be described on prenatal MRI by vermis hypoplasia, which can be associated with rotation. This variant was called the Dandy Walker variant: DWV. This shows hypoplasia of the vermis, which is rotated, but the posterior cranial fossa has a normal volume with existing hypoplasia of the vermis [4].

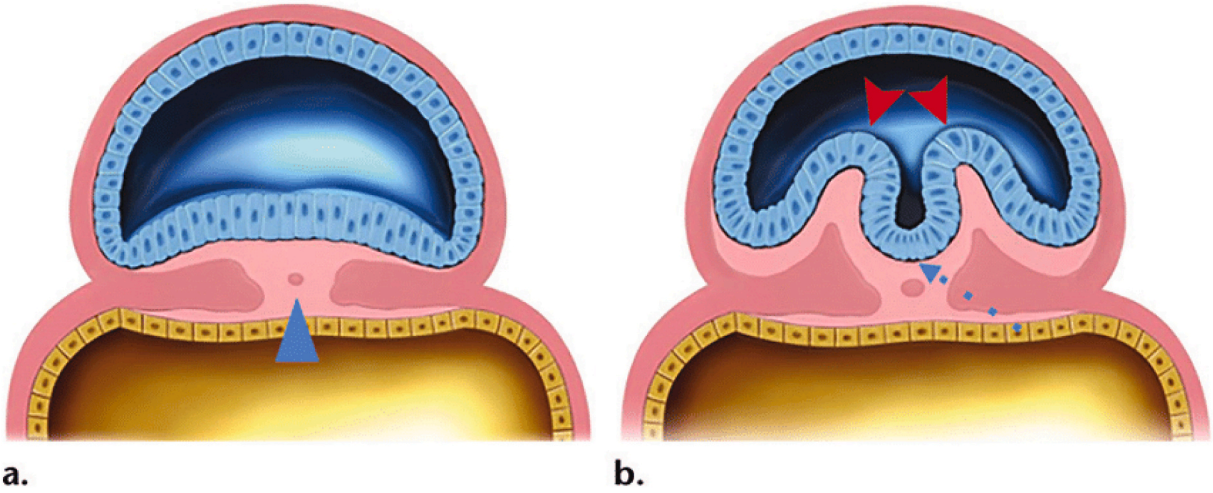

Fig. 3. Stages in the embryogenesis of the nervous system.
a. Neuronal induction: During the 3rd week of development, the neuroectoderm and later the neural crest emerge from the ectoderm.
b. Neurulation: On day 18, the neural plate deepens into a neural groove and finally forms the neural tube. The neural plate also forms the neural crest, from which ganglion cells, cells of the adrenal medulla, certain types of CNS glial cells and Schwann cells arise. The brain develops from the anterior part of the neural tube and the spinal cord, from the posterior part of the neural tube.

3.3. Differential diagnoses

3.3.1. Arachnoid cyst

Intracranial arachnoid cysts are accumulations of cerebrospinal fluid (CSF) between the dura mater and the brain substance. Unlike normal arachnoid membranes, arachnoid cysts contain a thick layer of collagen and hyperplastic arachnoid cells in the cyst wall, but fewer trabecular processes in the cyst. Location in the posterior cerebral fossa is a differential diagnosis with Dandy Walker malformation (Fig. 4) [5].

3.3.2. Blake's pouch cyst

In 1900, Joseph Blake described a transient posterior evagination of the choroidal web of the fourth ventricle in the 130-day-old normal human embryo. He was the first to recognise and fully elucidate the true nature of Magendie's foramen as an opening, which develops within a saccular expansion of the embryonic fourth ventricular cavity. The persistence of this temporary fourth ventricle in the postnatal period and its significance as a distinct entity or as part of the Dandy-Walker continuum has been one of the most controversial topics in the neurosurgical and neuroradiological literature over the years [6].

On MRI, Blake's pouch malformations are characterised by an increased tegmentovermian angle (Fig. 5) [4].

3.3.3. Megacisterna magna

The cisterna magna is the sub-arachnoid space between the medulla oblongata and the lower surface of the cerebellum and was first

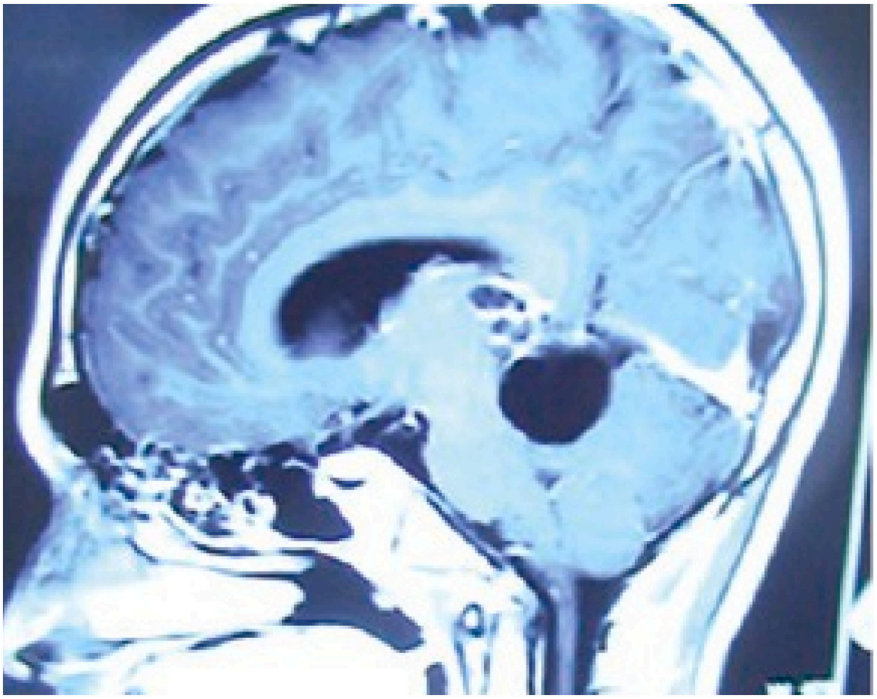

Fig. 4. Arachnoid cyst of the posterior fossa intra-fourth medial ventricle.

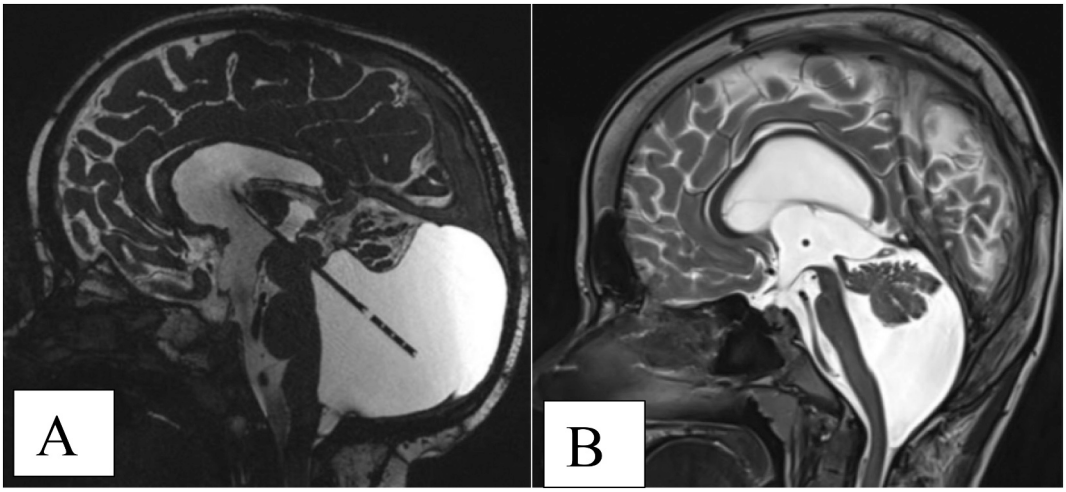

Fig. 5. Difference between Dandy Walker malformation (A) and Black pouch cyst (B) on MRI after birth.

described by Liliequist in 1952. Megacisterna magna is characterised by an enlarged cisterna magna associated with a normal 4th ventricle, normal cerebellar hemispheres and a normal vermis.

Mega-cisterna magna is defined as a large cistern >10 mm in diameter, with no change in the cerebellar vermis.

It is a relatively common disorder, accounting for almost half of all cystic malformations of the posterior fossa. It is generally asymptomatic and discovered by chance. A relationship between megacisterna magna and psychiatric disorders in children is currently being investigated [7,8]. Imaging can distinguish megacisterna magna from Blake's pouch cyst by the absence of hydrocephalus (Fig. 6) [9].

Patient consent

The patient has given written informed consent for the publication of this case report and accompanying images. A copy of the written consent is available for review by the editor of this journal upon request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

I'm Ayoub Amghar and I declare on my honor that the ethical approval has been exempted by my establishment.

Funding

None.

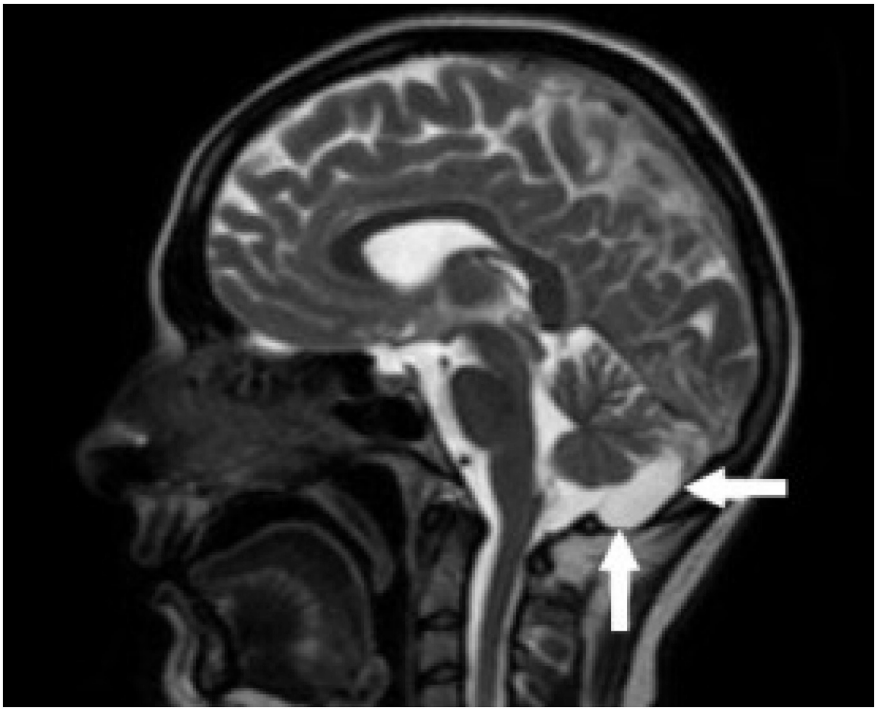

Fig. 6. Sagittal plane T2 sequence MRI shows a megacisterna magna (arrows) with the same intensity as the cerebrospinal fluid extending from the interhemispheric area towards the posterior part, which is more prominent [8].

Author contribution

AYOUB AMGHAR: Corresponding author writing the paper.
EL ABBASSI IMANE: writing the paper.
MOHAMED JALAL: study concept.
AMINE LAMRISSI: study concept.
ASMAA ASSAL: study concept.
SAID BOUHYA: correction of the paper.

Guarantor

Ayoub Amghar.

Research registration number

researchregistry2464.

Conflict of interest statement

None.

Data availability

None.

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