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Dandy-walker malformation associated with a large growing occipital meningocele: A rare case report and literature review

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Abstract

Dandy-Walker malformation (DWM) is a rare congenital anomaly of the posterior fossa. It is characterized by hypoplasia or agenesis of the cerebellar vermis, cystic dilatation of the fourth ventricle, and upward tentorial displacement. It is commonly associated with central nervous system (CNS) or systemic abnormalities. Occipital meningocele (OMC) is an extremely rare association with DWM, reported in only around 40 cases in the global literature. To our knowledge, this is the second reported case of a growing OMC associated with DWM in English literature, and the first documented in Saudi Arabia. By sharing our case report, we aim to contribute to the existing literature by adding to the available database for future research, which will help address such cases and establish a standardized management protocol to enhance patient care. We present a 4-month-and-7-day-old male infant diagnosed with DWM who developed progressive head enlargement and a growing occipital mass. The patient underwent surgical resection of the OMC sac with cysto-peritoneal shunt insertion and cranioplasty with titanium mesh to repair the bony defect. The occipital mass was large, firm, oval-shaped, and midline at the occiput, measuring 7 cm × 6 cm. Surgical intervention was performed successfully without perioperative complications. Postoperative recovery was uneventful. OMC in association with DWM is rare, and a progressively enlarging OMC is exceptionally uncommon. Management remains controversial due to the absence of standardized surgical guidelines. Our case underscores the importance of reporting such rare occurrences to expand the existing literature and guide future management strategies. This report contributes to the growing body of knowledge on the rare association of OMC with DWM. Collective case documentation is crucial to establishing an evidence base that supports the development of standardized protocols, ultimately enhancing patient safety, surgical outcomes, and long-term prognosis.

Keywords: Case report, Cephalocele, Cysto-peritoneal shunt, Dandy-Walker malformation, Occipital meningocele.

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1. Introduction

Dandy-Walker malformation (DWM) is a rare congenital anomaly affecting the posterior fossa. It is marked by underdevelopment or absence of the cerebellar vermis, cystic enlargement of the fourth ventricle, and upward tentorial displacement [1]. DWM is commonly associated with other central nervous system (CNS) abnormalities, including aqueductal stenosis, occipital meningocele, and syringomyelia [1, 2] as well as systemic abnormalities such as cardiac defects or urogenital malformations, cleft palate, and capillary hemangiomas [3]. Occipital meningocele (OMC) is a particularly rare anomaly linked with DWM, with roughly 40 cases reported in the global literature [3-5]. Reports of a growing OMC in the context of DWM are highly uncommon, having been reported only once in the English literature [3] in addition to the present case. We present a clinical scenario in which the patient presented with hydrocephalus and occipital meningocele that was small at birth and became progressively larger over time. Surgical management approaches in DWM and OMC are controversial, as there are no established protocols to determine whether to repair OMC alone, shunt alone, or use a combination of both [3]. To the best of our knowledge, this represents the first reported case of this kind in Saudi Arabia.

2. Case Presentation

A 4-month-and-7-day-old male infant, known to have occipital meningocele and Dandy-Walker malformation, was diagnosed by a postnatal brain MRI performed at another hospital [Figure 1, A-F]. He was referred to our tertiary center as a case of active hydrocephalus [Figure 2, A-C] requiring urgent surgical intervention. He was born at 31 weeks of gestation via cesarean section to a primigravida mother. The mother reported a rapid head growth of 11 cm in just one month, progressive enlargement of the meningocele sac, a tense anterior fontanelle, intermittent fever, mild cough, and reduced activity. At another hospital, ventricular tapping was performed with aspiration of approximately 45 ml of CSF to relieve high intracranial pressure. This procedure was complicated by a minor CSF leak that required suturing. Shunt placement was initially planned, but due to limited resources, the surgery was postponed, and the patient was urgently referred to our center. The mother recalled that the occipital mass was the size of a walnut at birth and had enlarged progressively. On admission, physical and neurological examination revealed a large, firm, oval midline occipital mass measuring 7 × 6 cm [Figure 3], covered by intact, hair-bearing skin. His vital signs were stable. He was alert, spontaneously opened his eyes, and moved all extremities symmetrically. Pupils were equal and reactive, and the anterior fontanelle was wide and full. CSF aspiration was performed and sent for analysis and culture. The head circumference measured 45 cm, placing him above the 97th percentile for his corrected age (2 months and 7 days). The patient was unable to support his head and slept only on his sides due to the large occipital swelling. There were no signs of sunset eyes or engorged scalp veins. During hospitalization, the patient developed tachycardia, desaturation (SpO2 87%), increased work of breathing, and respiratory distress. He was stabilized on a high-flow nasal cannula oxygen. A full septic workup was initiated, and he was admitted to the pediatric ICU(PICU) on empirical intravenous ceftriaxone and vancomycin (meningeal dose). The next day, he underwent an external cyst shunt placement for CSF drainage, excision of the occipital meningocele sac, and cranioplasty.

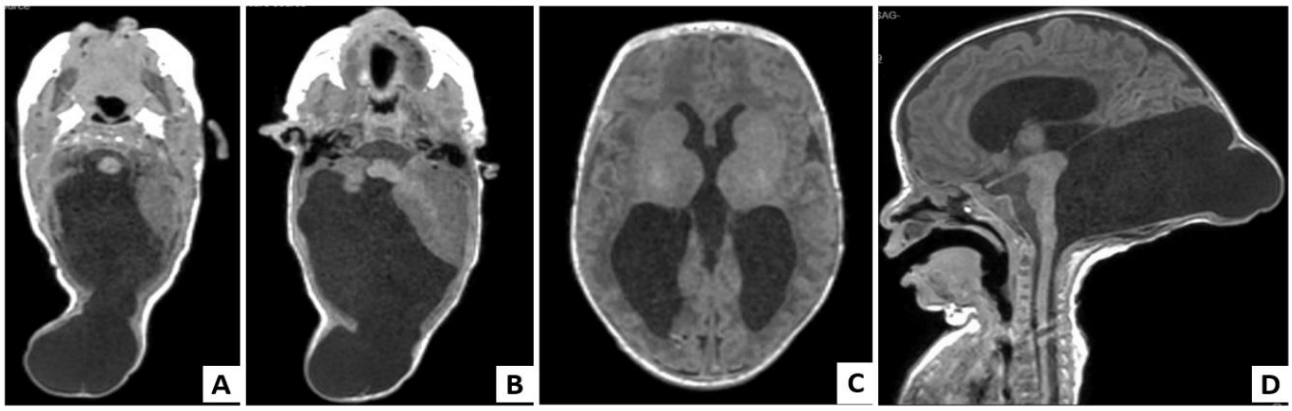

Figure 1.
Pre-operative non-enhanced MRI brain, T1W images. A, B) Axial views showing an occipital meningocele sac herniated through the occipital calvarial defect. Enlarged posterior fossa with a cyst measuring 7.3 cm × 6.3 cm connected to the fourth ventricle, compressing and displacing the hypoplastic right cerebellar hemisphere and the vermis anteriorly and the left cerebellar hemisphere laterally. C) Axial view showing racing car sign of corpus callosum dysgenesis. D) Cystic dilation of the fourth ventricle with communication to a large cystic dilated posterior fossa with accompanying elevation of the tentorium cerebelli.

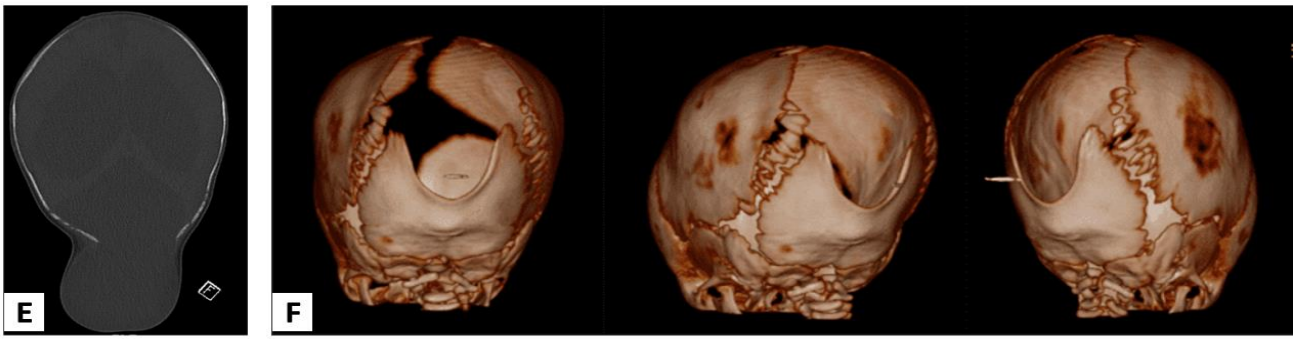

Figure 1.
E) CT scan axial view bone window showing the occipital bony defect. F) 3D reconstruction of the skull demonstrating different views of the occipital bony defect.

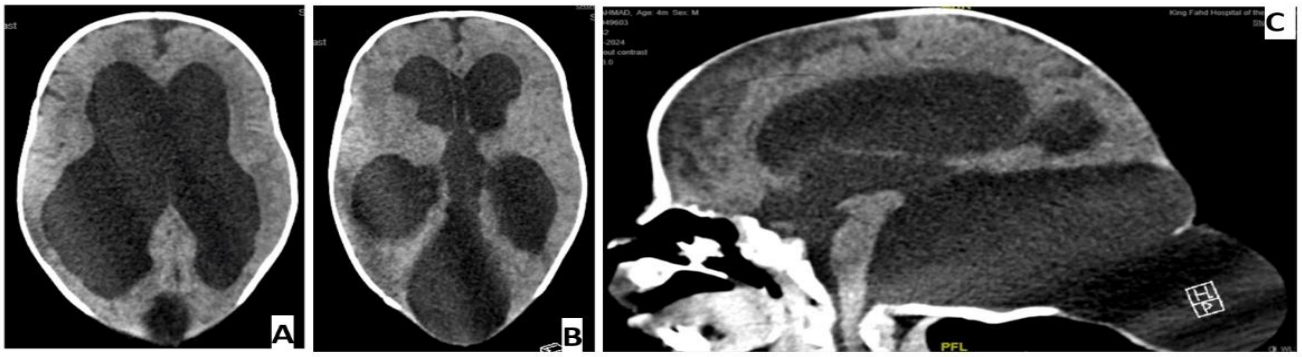

Figure 2.
Pre-operative non-enhanced CT brain scans upon admission. A, B) Axial views showing global dilation of the ventricular system with colpocephaly. C) Sagittal view showing global dilation of the ventricular system, enlargement of the posterior fossa, upward tentorial displacement, and a large meningocele protruding through an occipital bony defect.

The decision to perform temporary external shunting was made to control intracranial pressure while ruling out infection before inserting a permanent cysto-peritoneal shunt (CP-S). Intraoperatively, after induction of general anesthesia, the patient was positioned prone on a pediatric U-shaped Mayfield horseshoe headrest [Figure 4, A, B]. The skin was dissected off the meningocele sac [Figure 4, C, D]. The dural sac was opened, releasing approximately 50 ml of CSF [Figure 4, E]. Redundant dura and excess skin [Figure 4, F] were excised, followed by a water-tight dural repair. A titanium mesh cranioplasty with miniplate screw fixation was performed to cover the large bony defect, and an external cyst shunt was inserted before skin closure [Figure 4, G, H].

Figure 3.
Pre-operative physical examination: demonstrating a large, firm, oval-shaped midline mass at the occiput measuring 7 cm × 6 cm.

The skin overlying the mas is covered with hair and is intact.

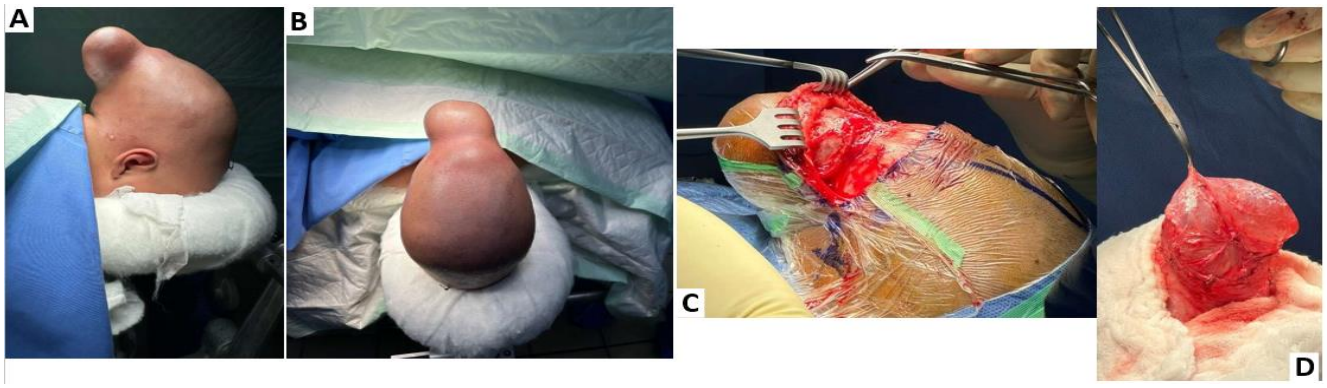

Figure 4.
Intraoperative images. A, B) Showing intra-op positioning with the head resting on the pediatric U-shaped Mayfield horseshoe. The OMC sac is showing an intact overlying skin. C) Showing a skin incision around the OMC sac, dissecting the skin and subcutaneous tissue off the OMC sac carefully, and cutting off the adhesions to the sac without violating the sac layer. D) exposing the fluid-filled dural sac.

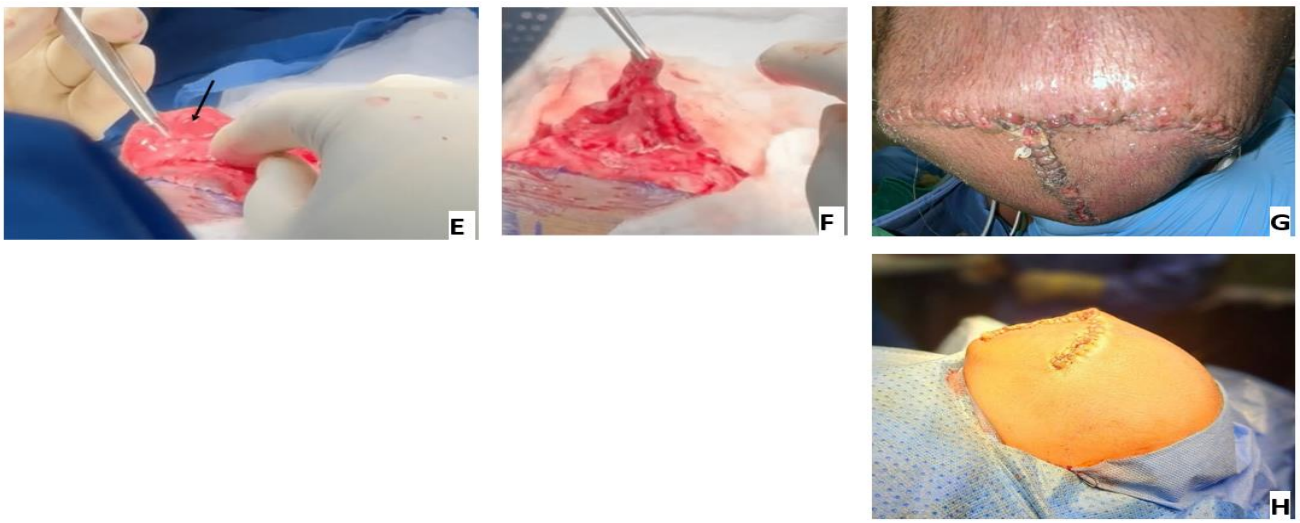

Figure 4.
Intra and postoperative images. E) Showing a gush of CSF (black arrow) once the dural sac was opened. F) Showing the redundant dural sac. G, H) The occipital region of the head postoperatively after repairing the defect.

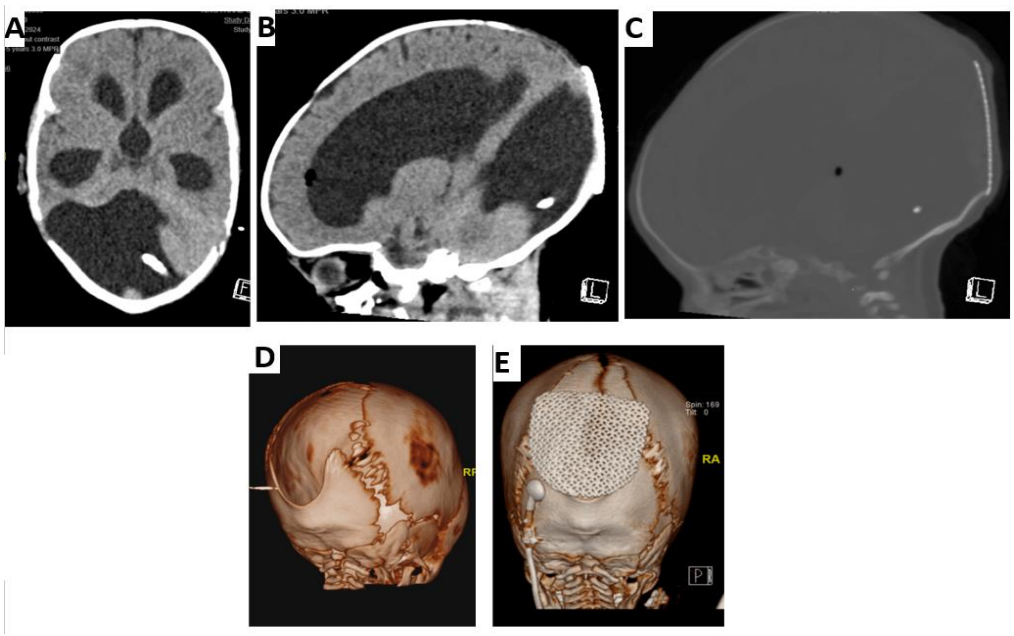

Figure 5.
Postoperative non-enhanced CT brain scans. A) Axial view showing the proximal end of the external, temporary shunt placed at the cystic enlarged posterior fossa. B) Sagittal view showing the tip of the shunt at the cystic enlarged posterior fossa and the repaired occipital bony defect by titanium mesh. C) Sagittal view bone window showing repair of the occipital bony defect. D, E) 3D reconstruction showing titanium mesh covering occipital bony defect.

Postoperatively, the external drain was initially kept closed to assess the need for permanent shunting. The patient remained intubated and was returned to the PICU. Serial CSF cultures from the external drain were negative. Postoperative head CT [Figure 5, A-E] was performed. On postoperative day 2, the patient became bradycardic and desaturated (SpO2 60%). The drain was opened, draining 40 mL of CSF, and maintained at a level of 5 cmH2O; after this, the patient stabilized. Blood and urine cultures were negative. After one week, the external drain was removed, and a permanent CP-S was inserted without complications. He was shifted back to the PICU in a stable condition. By postoperative day 1, he was alert, with spontaneous eye opening, equal limb movements, reactive pupils, and a soft, non-pulging anterior fontanelle. On day 2, he was extubated and weaned to room air. By day 3, he was transferred to the regular pediatric ward. A follow-up CT scan was performed [Figure 6, A, B], and the patient was discharged home in stable condition.

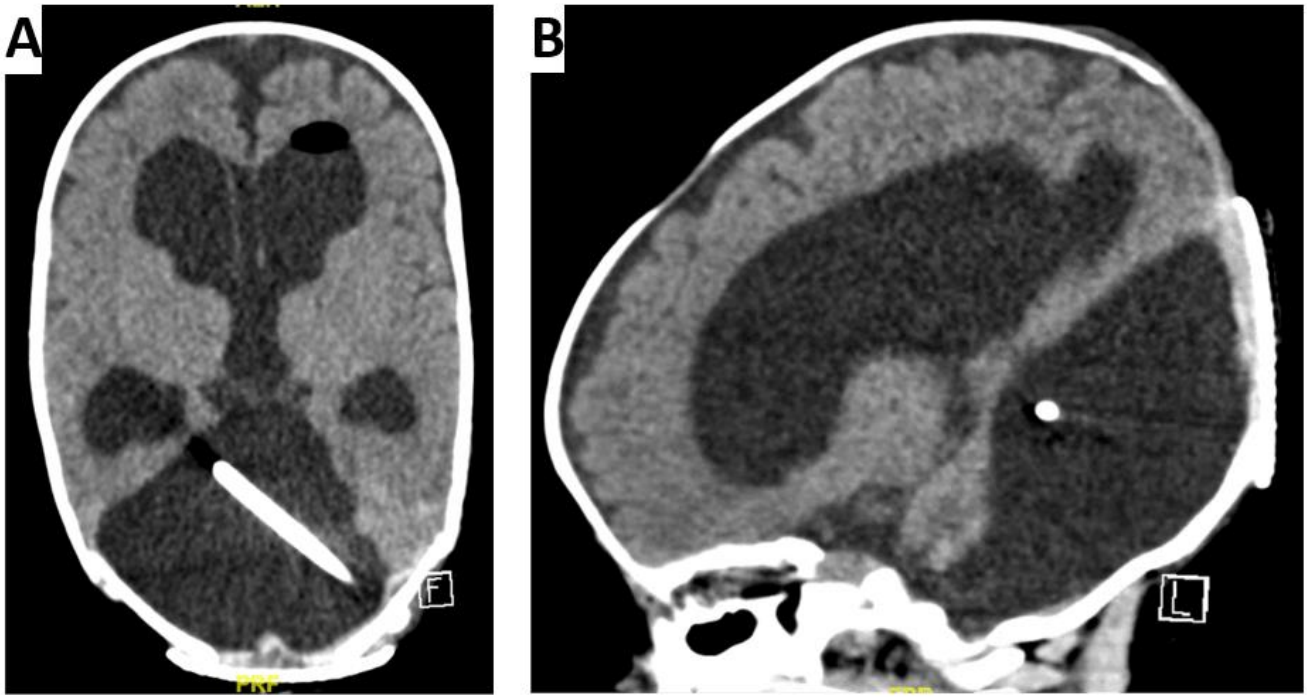

Figure 6.
Postoperative non-enhanced CT brain scans upon discharge. A) Axial view showing the proximal end of the permanent CP-S placed at the cystic posterior fossa with interval regression of the ventricular size. B) Sagittal view showing postoperative changes and the tip of the CP-S at the cystic enlarged posterior fossa and the repaired occipital bony defect by titanium mesh.

3. Discussion

Dandy-Walker malformation (DWM) is a rare congenital anomaly of the posterior fossa. It is frequently associated with central nervous system (CNS) abnormalities, including heterotopia, aqueductal stenosis, occipital meningocele, agenesis of corpus callosum, neural tube defects, hydrocephalus, epilepsy, polymicrogyria, and syringomyelia [1, 2]. Systematic abnormalities may also coexist, such as cardiac anomalies or urogenital defects, cleft palate, capillary hemangiomas, and, in dome cases, PHACES syndrome [3]. The reported incidence of DWM is approximately 1 in 30,000 live births [5] with a slight female preponderance [6]. Occipital meningocele (OMC) is an exceedingly rare malformation found in association with DWM Mankotia, et al. [4] and Al-Obaidi, et al. [5]. Matsumoto, et al. [3] documented that the coexistence of DWM and OMC has been reported in only about 40 cases worldwide. This number may underestimate the true incidence, as the widespread availability of antenatal ultrasound often leads to early detection of congenital anomalies and subsequent termination of the pregnancy [4]. Furthermore, because most cases of DWM with OMC undergo surgical correction in infancy, the natural history of unprogressive OMC is likely underreported Talamonti, et al. [7]. Long, et al. [8] conducted a population-based study in 2006 using the British NorCAS survey, which records all significant abnormalities in fetuses, stillbirths, and live-born children. Over the course of 18 years, involving approximately three million births, the authors identified no cases of combined DWM and OMC. Similarly, Di Nora, et al. [9] in a 2022 retrospective study evaluating the clinical and neuroimaging features of DWM and its variant in pediatric patients in the period from 2005 to 2021, reported no cases of OMC associated with DWM. In contrast, Bindal, et al. [10] identified eight patients with associated OMC in a 1991 review of their institutional series of 50 DWM cases. They also collected an additional eleven cases from the global literature, concluding that OMC was present in 16% of DWM cases. More recently, Talamonti, et al. [7] proposed a classification of OMC based on size, dividing cases into small (<5cm), large (5-9 cm), and giant (>9 cm). The mortality risk is significantly higher in patients with large and giant OMC associated with DWM compared to those with smaller OMC [3]. OMC is thought to develop during the sixth or seventh week of gestation [4, 5]. Prenatal diagnosis of DWM is typically made using ultrasound after the 18th week of gestation, as the cerebellar vermis is not fully developed until this stage; attempts at diagnosis before this point are unreliable [1]. In the neonatal period, brain MRI serves as the gold standard for confirming DWM, allowing detailed evaluation of the posterior fossa as well as other associated

intracranial abnormalities [11]. MRI also plays a critical role in assessing the OMC by differentiating the contents of the sac. [11]. By definition, a cranial meningocele refers to the herniation of the meninges through a cranial bony defect, whereas encephalocele involves the herniation of both meninges and neural tissue. The broader term "cephalocele" encompasses both entities [4, 5]. In most cases, the overlying skin remains intact [7]. Importantly, in the presence of an open fontanelle and an OMC sac communicating through a bony defect, the raised intracranial pressure caused by hydrocephalus may be partially relieved through this communication [4]. Furthermore, the OMC sac may function as a one-way valve due to the presence of incomplete septations, permitting CSF to escape from the cranial cavity into the OMC sac without reflex, leading to its progressive enlargement [7]. Surgical repair of OMC alters CSF flow dynamics and reduces the available surface area for CSF resorption [4]. The optimal surgical strategy for OMC in the setting of DWM remains controversial [11] as no standardized protocols exist to guide the choice between OMC repair alone, shunting alone, or a combined approach [3]. Early surgical intervention is considered essential for both minimizing the risk of rupture [4] and reducing the likelihood of neurological deficits [5]. Based on a review of 32 published cases, the most favorable outcomes were achieved when OMC repair and shunt placement were performed simultaneously as the initial operation [3] [2,12]. Repair of OMC alone has generally been associated with poor outcomes [3]. In cases of small OMC, shunting alone may be considered as the first step, with subsequent repair depending on the clinical course [3, 4, 7]. Conversely, for large or giant OMCs, shunting alone yields poor outcomes, and a combined surgical strategy is strongly recommended to achieve better results [4]. The choice of shunt type in DWM with OMC remains unsettled. Options include ventriculoperitoneal shunt (VP-S), cysto-peritoneal shunt (CP-S), or, in some cases, a combination of both, with no consensus yet established regarding superiority [3, 4]. The most critical requirement for a single shunt placement is the patency of the cerebral aqueduct [12]. Careful selection of the shunt type is essential. According to the literature, CP-S has been more commonly employed as the initial surgical intervention, as it maintains the downstream flow of CSF. Some authors advocate for the use of a VP-S as the first-line procedure, as it provides early decompression of the supratentorial compartment and may lower the risk of complications. Others, however, advocate for CP-S because VP-S can lead to an isolated fourth ventricle and can develop acquired stenosis of the cerebral aqueduct [3]. Various cranioplasty techniques have been described for repairing skull defects in pediatric patients, including the use of hydroxyapatite bone cement, titanium mesh, demineralized bone matrix, methyl methacrylate, autologous calvarial bone grafts, and rib grafts [13]. In children, autologous cranioplasty is generally preferred over allografts or synthetic materials because of its lower infection and rejection rates, improved blood perfusion and bony fusion, and its ability to expand with the child's growth [14, 15]. For dural closure in OMC-associated skull defects, Alojan, et al. [15] described a modified cable suturing technique designed to minimize the risk of postoperative CSF leak and prevent cerebellar tissue herniation. This is achieved by dissecting the dura from the bony edges and subcutaneous tissue, then applying tension by suturing it repeatedly in oblique planes, thereby eliminating dural dead space without opening the dura. The prognosis of DWM largely depends on the severity of the malformation and the extent of associated anomalies [16] with overall morbidity reported to be between 26% and 50% [4]. Fetal ultrasound measurement of the lateral ventricle diameter has a significant prognostic value; diameters of 11-15 mm are associated with a 21% risk of developmental delay, whereas measurements >15 mm increase this risk to over 50% [1]. A recent retrospective study evaluating postoperative neurodevelopmental outcomes in patients with occipital encephaloceles from 2012 to 2022 based on different variables [17] stated that the presence of hydrocephalus, sac size >5cm, and the presence of neuronal tissue within the sac, based on histopathological assessment, are all correlated with developmental delay and poor neurological outcomes.

4. Conclusion

Occipital meningocele (OMC) in association with Dandy–Walker malformation (DWM) represents an infrequent and diagnostically challenging entity. The occurrence of a progressively growing OMC within this setting is even more exceptional, and its management remains a subject of considerable debate. To date, there are no globally established guidelines to guide surgical decision-making, whether to pursue OMC repair alone, CSF diversion procedures, or a combined approach. By reporting this case, we aim to enhance the collective understanding of the clinical course, diagnostic complexities, and therapeutic challenges inherent to OMC with DWM. Such a contribution is essential for developing standardized management protocols, which can improve surgical outcomes, patient safety, and long-term prognosis. In this regard, our report not only documents a rare occurrence but also emphasizes the importance of continuous data sharing and collaborative research in shaping the future of care for these complex patients.

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