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Trapped fourth ventricle: A case report and review of literature

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Trapped fourth ventricle (TFV) also known as isolated fourth ventricle (IFV) is a rare clinico-radiologic entity with only a few cases reported in the literatures.

The aim of this article is to present the first case of this condition in our center and highlight the challenges of arriving at clinical diagnosis and treatment in a resource limited setting.

CASE PRESENTATION: An 18 months old girl who had ventriculoperitoneal shunt insertion for post meningitic hydrocephalus 4 months earlier presented with restlessness, ataxia, fever and inability to control her neck of one-week duration. On examination she was restless and had retro-colis with a Glasgow Coma Scale (GCS) score of 11/15 (E4V2M5).

She had an associated facial and abducent nerve palsies with global hypertonia, hyper-reflexia and muscle power of 3/5.

She was initially treated for shunt infection and malfunction. However, shunt series and CSF analysis were within normal limits and CSF culture yielded no growth of microorganisms. A CT scan of the brain which was ordered earlier was delayed for 10 days due to financial constraints. The CT scan revealed a trapped fourth ventricle and slit lateral and third ventricle.

She had emergency fourth ventriculoperitoneal shunt inserted on the left because of the pre-existing supratentorial shunt on the right.

She did well after the surgery and was discharged on the 10th postoperative day. She was doing well 12 months after the surgery.

RELEVANCE AND IMPACT: TFV may occur after insertion of VPS for post-meningitic hydrocephalus. This may present a diagnostic dilemma. Insertion of a second VPS may be an option in a resource limited setting.

1. Introduction

Trapped fourth ventricle (TFV) also known as isolated fourth ventricle (IFV) is a rare clinico-radiologic entity with only a few cases reported in the literatures. Most are detected incidentally without significant neurological deficit [1,2]. It occurs when there is obstruction of both the aqueduct of Sylvius and exit foramina i.e. Magendie and Luschka or basal cisterns of the fourth ventricles [3]. This will lead to progressive accumulation of cerebrospinal fluid (CSF), which initially produces mass effect on the cerebellum behind similar to posterior fossa space occupying lesion and subsequently the brain stem anteriorly. While the exact mechanism by which the fourth ventricle is trapped is not fully known, it occurs more commonly in the setting of post-haemorrhagic and post-infective hydrocephalus and also in patients who have had multiple shunt procedures (youman's Isolated Fourth Ventricle Syndrome) [4]. Careful attention to densities and configurations on computed tomography scan of the brain allows its differentiation from other lesions such as cystic tumors and cysts [3].

The treatment of symptomatic TFV varies depending on the availability of resources. In advance centers endoscopic approaches is the main stay of treatment [5]. In other places insertion of a fourth ventricle shunt which is then connected to the pre-existing supratentorial shunt via a Y-connector is used [5]. Others insert a separate fourth ventriculo-peritoneal shunt (FVPS).[5] The insertion of the FVPS is usually technically challenging.

The aim of this article is to present the first case of this condition in our center and highlight the challenges of arriving at a clinical diagnosis and treatment in a resource limited setting.

This case report has been reported in line with the SCARE 2020 Criteria [6].

2. Case report

An 18 months old girl who presented on self-referral with 3 months history of inability to see and regression of already achieved developmental milestones. She was treated at another hospital for acute bacterial meningitis during 6 months earlier. The patient was not on any medication upon presentation. There is no family history of hydrocephalus or other congenital anomalies.

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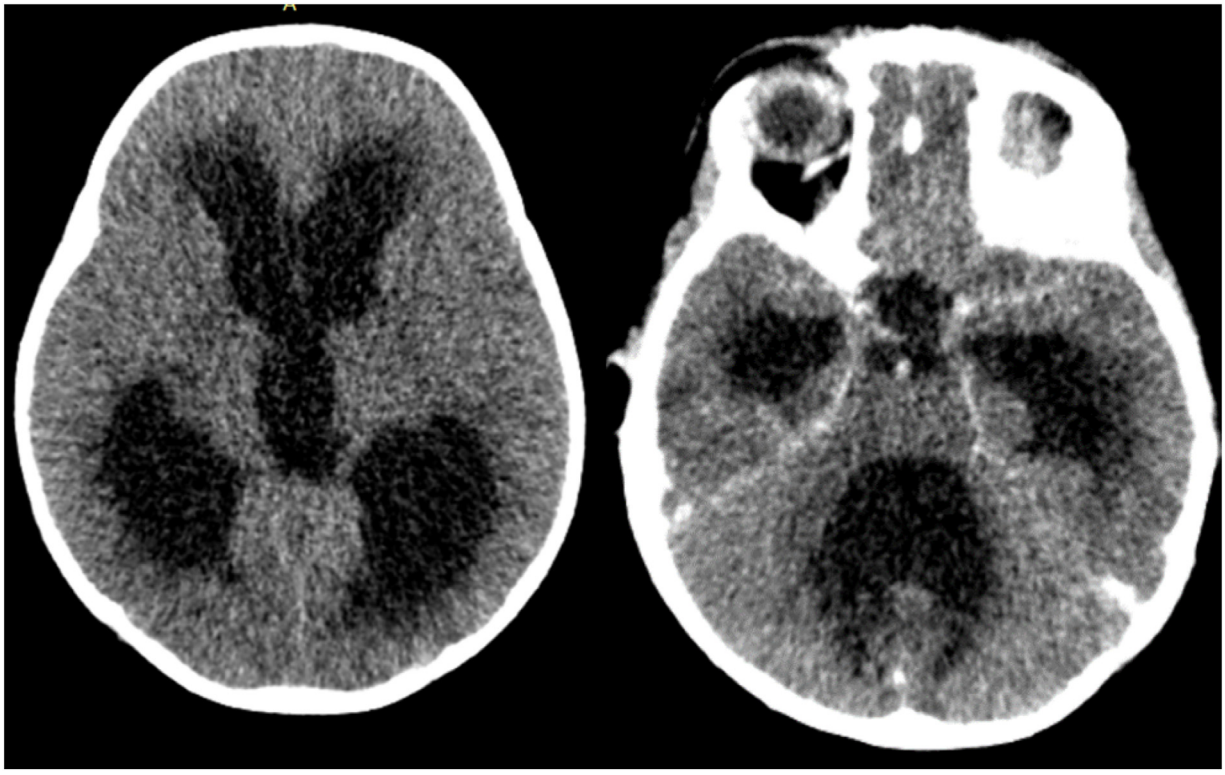

Fig. 1. Initial Preoperative Brain CT Scan showing communicating hydrocephalus.

On examination, her general condition was stable with a Glasgow Coma Scale (GCS) Score of 15/15 and global hyperreflexia and hypertonia. On visual examination she had perception of light in both eyes.

A brain computer tomographic scan showed communicating hydrocephalus; Fig. 1.

Careful examination and appropriate investigations were carried out and an emergency ventriculo-peritoneal shunt (VPS) insertion at the Keen's point was done. The surgery was done by D.S the senior attending neurosurgeon.

Patient symptoms resolved within 4 weeks. However 4 months after the procedure, patient presented with restlessness, ataxia, fever and inability to control her neck of one week duration. On examination she was restless and had retro-colis with a GCS score of 11/15 (E4V2M5).

She had an associated facial and abducent nerve palsies with global hypertonia, hyper-reflexia and muscle power of 3/5. A shunt series done was within normal limit.

An initial diagnosis of shunt infection was done and she was placed on broad spectrum antibiotics. However, the cerebrospinal fluid (CSF) analysis performed was within normal limit and CSF culture yielded no growth of any microorganism. A CT scan of the brain which was ordered thereafter was delayed for 10 days due to financial constraints. The CT scan revealed a trapped fourth ventricle with slit lateral and third ventricles. (Fig. 2)

Careful examination and appropriate investigations were carried out and an emergency fourth ventriculoperitoneal shunt was inserted on the left side because of the pre-existing supratentorial shunt on the right side. A Chabra medium pressure shunt manufacture by Surgiwear Limited, India was used.

She did well after the surgery and was discharged on the 10th postoperative day. She was doing well 12 months after the surgery and during the last follow-up visit, the patient's caregiver was satisfied with the current state of the patient.

3. Discussion

The term entrapped fourth ventricle, also called isolated fourth ventricle syndrome, has been used to describe the situation in which the fourth ventricle no longer communicates with the third ventricle, as well as the basal cisterns [7].

It is thought that secondary aqueduct stenosis from adhesions, obstruction of the exit foramina, or infective debris pooling in the basal cisterns may be responsible for this condition.[7] Patients may have the typical symptoms and signs of hydrocephalus or more atypical symptoms such as lower cranial nerve dysfunction. Occasionally, an entrapped fourth ventricle is an incidental finding on imaging [4].

Sometimes, compartmentalization of the ventricular system can be noted after CSF shunting.[8] For instance, after shunt placement in the lateral ventricles there may be persistent and progressive dilatation of the fourth ventricle with associated brain stem and cerebellar dysfunction [8].

The pathogenesis of trapped fourth ventricle in our index case was probably due to collapse of lateral and third ventricles resulting in slit ventricles. This was similar to findings in some literatures [1,3,9]. In contrast, some studies suggested transtentorial downward herniation of dilated posterior ventricle, combined with upward displacement of dilated cerebral aqueduct by enlarging fourth ventricle producing sufficient distortion of the aqueduct leading to an isolated fourth ventricle [10].Other studies reported secondary changes in the aqueduct following a ventricular shunt insertion [8].

In reported cases of TFV the etiology of initial ventricular dilatation included post-meningitic, post-hemorrhagic, dandy walker cyst, cerebellar and brainstem degeneration and enlargement secondary to outlet obstruction by tumor [3]. Our index patient had post-meningitic hydrocephalus.

Clinical presentation of patients with entrapped fourth ventricle is variable and runs a spectrum from being an incidental finding

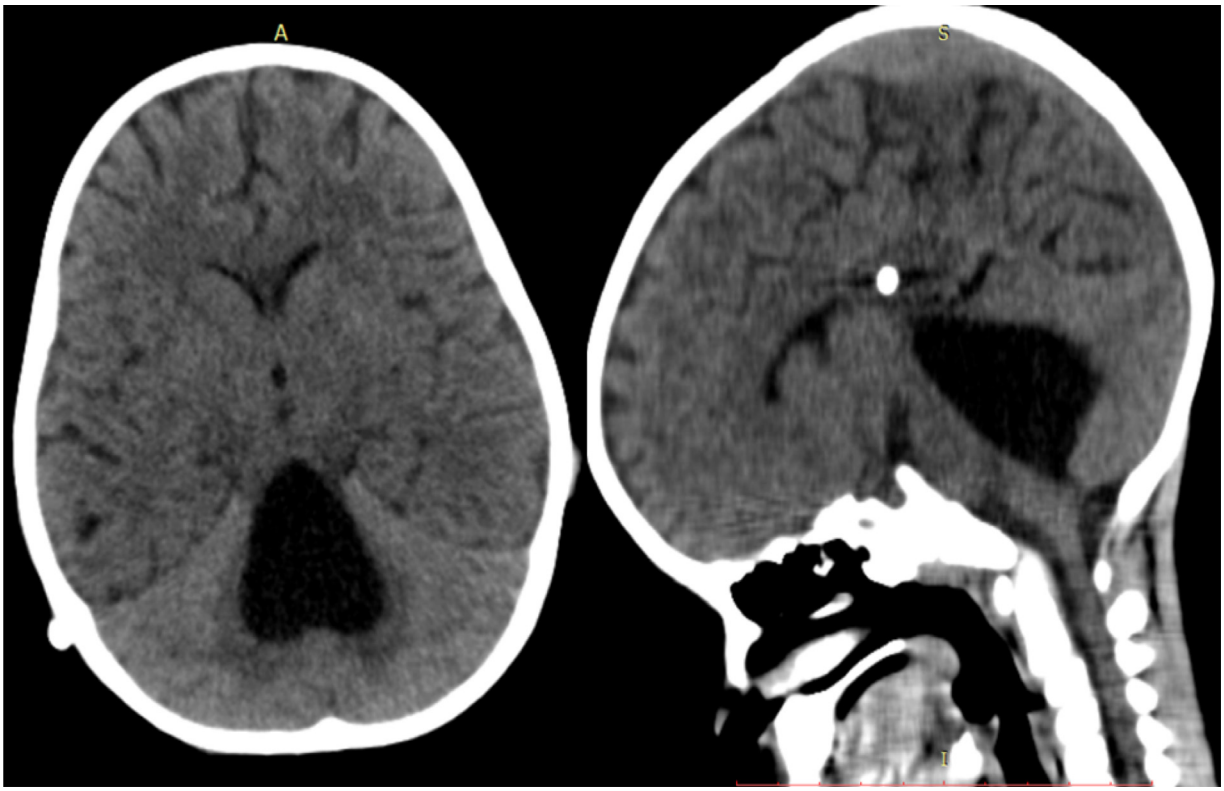

Fig. 2. Brain CT scan showing dilated fourth ventricle and slit lateral and third ventricles.

on brain CT scan to acute posterior fossa syndrome i.e. Ataxia, lethargy, diplopia, nystagmus, mental confusion, dysarthria, and multiple cranial nerve palsies [9]. However our patient presented with retrocolis in addition to ataxia and cranial nerve palsies.

Brain CT or MRI diagnosis of isolated fourth ventricle is mostly accurate. The most striking finding is the presence of a very large fourth ventricle, accompanied by very small or slit-like lateral and third ventricles, similar to our index patient. The fourth ventricle appears rounded or ballooned, the brain stem is displaced ventrally and posterior fossa subarachnoid spaces and cisterna magna are reduced in size or obliterated [9]. A combination of metrizamide ventriculography with CT scanning has also been used to provide an efficient and accurate method to differentiate entrapment from enlargement of the fourth ventricle [9,11].

Treatment options for isolated fourth ventricle include fourth ventriculo-peritoneal shunting which was done in our patient using the chabra medium pressure shunt. Other forms of treatment reported include direct microsurgical approach, veil excision (usually an arachnoid veil) and open aqueduct canalization or outlet fenestration of the fourth ventricle [9]. Endoscopic approaches include aqueductoplasty, aqueductal stenting, and cystoventricular fenestration [5]. Some authors favored a direct microsurgical opening of the outlet of the fourth ventricle, without insertion of a fixed foreign body, as the preferred primary surgical option for entrapment of the fourth ventricle, concluding that foramen magnum decompression is an effective treatment alternative [12]. However, shunting of the fourth ventricle has been considered by most authors as the less invasive and more effective approach [1,9]. Placement of the new fourth ventricular catheter could be through midline or lateral approach which may then be connected to a preexisting lateral ventriculo-peritoneal shunt catheter tubing if feasible [5]. Some authors recommended cannulating the posterior fossa cyst under direct stereotactic technique or ultrasound or endoscopic guidance to improve the safety and effectiveness of shunting, to avoid possible direct trauma to the brain stem and to ensure exact placement [13,14]. Although not done in our patient due to lack of availability of Y connector, it is important to connect the fourth ventricular catheter to the pre-existing ventriculo-peritoneal shunt with a Y connector [5]. The use of Y connector helps prevent the creation of a pressure gradient between the infra and supratentorial compartment, with possible onset of terrible, disabling headaches triggered by the move from reclining to upright standing [5].

It is worthy to note that the placement of a new shunt system in the fourth ventricle in our case significantly relieved the symptoms of raised intracranial pressure without the development of any complications related to the fourth ventricular shunting. Post-operative course was uneventful and complications were not noticed in the 12 months follow-up period. The patient caregiver was satisfied with the significant improvement made by the patient which include her abilities to see and walk again.

4. Conclusion

Trapped fourth ventricle may occur after insertion of VPS for post-meningitic hydrocephalus. This may present a diagnostic dilemma. Insertion of a second VPS may be an option in a resource limited setting.

Declaration of Competing Interest

No conflict of interest.

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Ethical approval

Ethical approval was obtained from the hospital ethical review committee.

Consent

Written informed consent was obtained for the case report.

Author contribution

The Authors contributed equally in all aspect of the case report

Registration of research studies

Not applicable.

Guarantor

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Provenance and peer review

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References

[1] H.E. James, Spectrum of syndrome of the isolated fourth ventricle in post hemorrhagic hydrocephalus of the premature infant, Pediatr. Neurosurg. 16 (1990-91) 305–308.
[2] C. Dollo, A. Kanner, V. Siomin, Outlet fenestration for isolated fourth ventricle with and without an internal shunt, Child's NervSyst 17 (2001) 483–486.
[3] R.A. Zimmerman, L.T. Bilanuik, E. Gallo, Computed tomography of trapped fourth ventricle, Am. J. Roentgenol. 130 (1978) 503–506.
[4] NicoleC. Keong, Marek Czosnyka, Zofia Czosnyka, John D. Pickard, Clinical evaluation of adult hydrocephalus, in: H. Winn (Ed.), Youmans Neurological Surgery, 6th edition, Saunders, 2011.
[5] Beng Ti Ang, Paul Steinbok, D. Douglas Cochrane, Etiological differences between the isolated lateral ventricle and the isolated fourth ventricle, Childs NervSyst 22 (2006) 1080–1085.
[6] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230.
[7] Ali Hikmat Azeez, Trapped fourth ventricle: case study and review of literature, Iraqi PG Medical Journal 13 (3) (2014).
[8] Alia Hdeib, Alan R. Cohen, Hydrocephalus in children and adults, in: Richard Ellenbogen, Saleem Abdulrauf, Laligam Sekhar (Eds.), Principles of Neurological Surgery, 3rd edition, Saunders, 2012.
[9] Pietro Spennato, Giuseppe Cinalu, Giuseppe Carannante, Claudio Ruggiero, Maria Laura Del Basso de Caro, Multiloculated hydrocephalus, in: G. Cinalli, W.J. Maixner, C. Sainte-Rose (Eds.), Pediatric Hydrocephalus, Springer-Verlag, Milan, 2005.
[10] A.J. Raimondi, Pediatric Neuroradiology, Saunders, Philadelphia, 1972.
[11] Mauricio Collada JR., Joseph Kott, David G. Kline, Documentation of fourth ventricle entrapment by Metrizamide ventriculography and CT scanning: report of two cases, Neurosurgery J. 55 (1981) 838–840.
[12] U. Suhas, Xiao Bo, Liat Ben Sira, C. Shlomi, Unusual subacutediencephalic edema associated with a trapped fourth ventricle: resolution following foramen magnum decompression, Childs NervSyst 25 (2009) 1517–1520.
[13] H.G. Eder, K.A. Leber, W. Gruber, Complications after shunting isolated fourth ventricle, Child's NervSyst 13 (1997) 13–16.
[14] J.L. Montes, D.B. Clarke, J.P. Farmer, Stereotactic transtentorial hiatus ventriculoperitoneal shunting for the sequestered fourth ventricle, J. Neurosurg. 80 (1994) 759–761.