Case Lesson *19*

Hydrocephalus and infratentorial hygromas following posterior fossa decompression with dural splitting for Chiari I malformation in pediatric patient: Conservative management.

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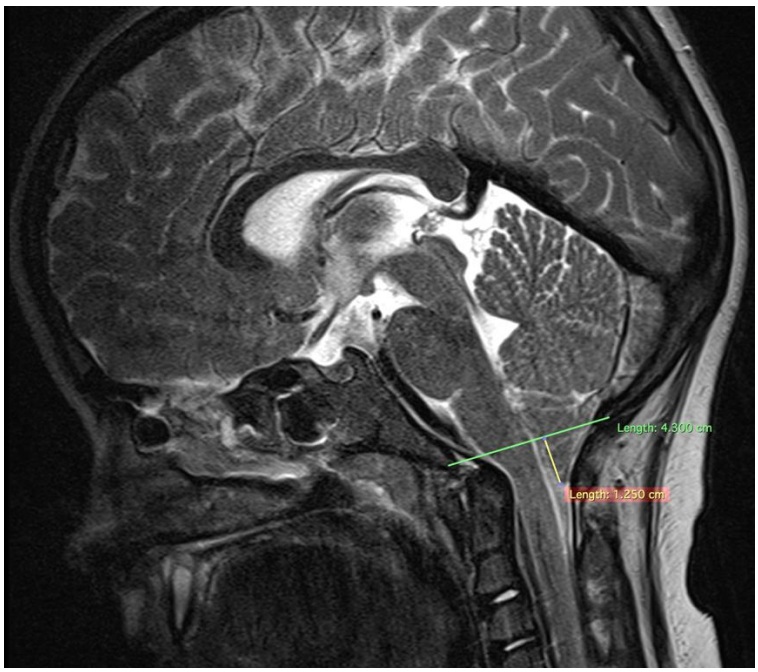
Background

De novo hydrocephalus following posterior fossa decompression is a rare complication with an incidence at about 5% as reported in literature . There is considerable debate regarding the best treatment option. The selection among acetazolamide and dexamethasone treatment, EVD, ETV, VPS or wound exploration for further decompression; has not reached consensus.

Case Lesson

A 9-year-old boy diagnosed with Chiari type 1 malformation in 2020 in a different institute, held under observation with frequent MRI, came to our clinic in 2023 complaining of headache, cervicalgia and difficulty breathing during sleep. In clinical evaluation the patient was of normal development, BMI 26.1 - Class 1 obesity. Fundus oculi examination showed no signs of papilledema. Polysomnography revealed no presence of obstructive sleep apnea. The patient presents with headaches of occipito cervical region that are worsened by Valsalva maneuvers. Neurological examination was normal. NIHSS 0p. GCS 15p. MRI of the brain demostrated a CM1 with pointed tonsills 1.25 cm under the foramen magnum in reference to McRae line (Fig.1a). Evans index was estimated as 0.24 (Fig.1b). There is no evidence of ventriculomegaly or infratentorial hygromas on preoperative imaging and no syrinx is present (Fig 1a,b)



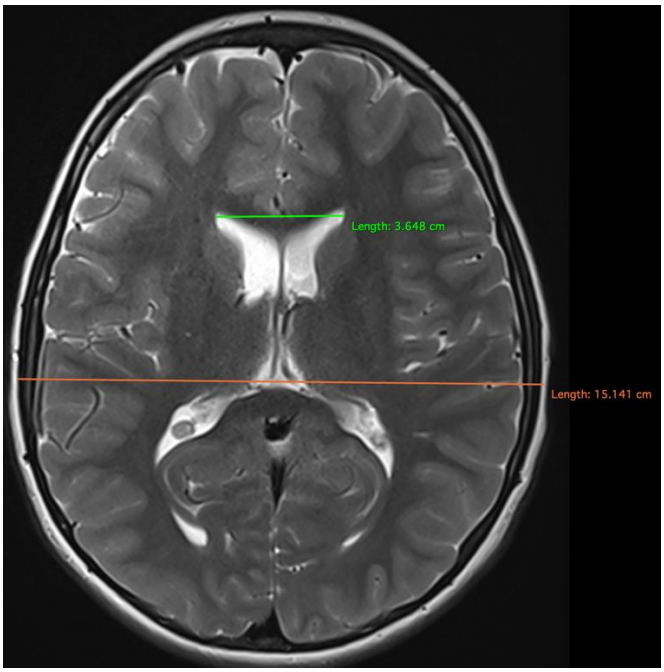


*Fig.1 a)*Sagittal T2-weighted MRI. *b)* axial T2-weighted MRI scans obtained pre-FMD.
(***a***) demonstrating a CM Type I with tonsillar descent 1.25 cm below the McRae line. (***b***) Evans index measured 0.24.

Considering a symptomatic CM1 with suboccipital headaches that are worsened by Valsalva maneuvers and a tonsillar descent greater than 1cm, surgical intervention was proposed.
The patient underwent an elective foramen magnum decompression with C1 laminectomy and dural splitting. Arachnoid membrane was not opened (Fig.2). Tonsils were observed through the arachnoid undulating with cardiac pulsations. No CSF leak and no subarachnoid bleeding was apparent during the procedure. Postoperative course was uneventful, headaches were improved and the patient was discharged by day 3 postsurgery.

Fig.2 Dural splitting is shown with no craniectomy of occipital bone. Black arrow pointing at arachnoid membrane, shining, no scars visible. The tonsils can be seen under the arachnoid with cardiorespiratory free up and down movement of both tonsils (on video).

Four weeks after surgery the patient complains of severe headache during the last 5 days, nausea and vomiting. The patient was readmitted to our institution. The character of presenting symptoms at readmission was different from pre FMD, headache was constant and located in frontal and orbital region. Neurological examination findings were normal. The ophtalmology results were normal with no signs of papilledema. A brain MRI was obtained revealing de novo hydrocephalus, bilateral infratentorial extraaxial fluid collections, pseudomeningocele and cerebellar ptosis. (Fig.3a,b).

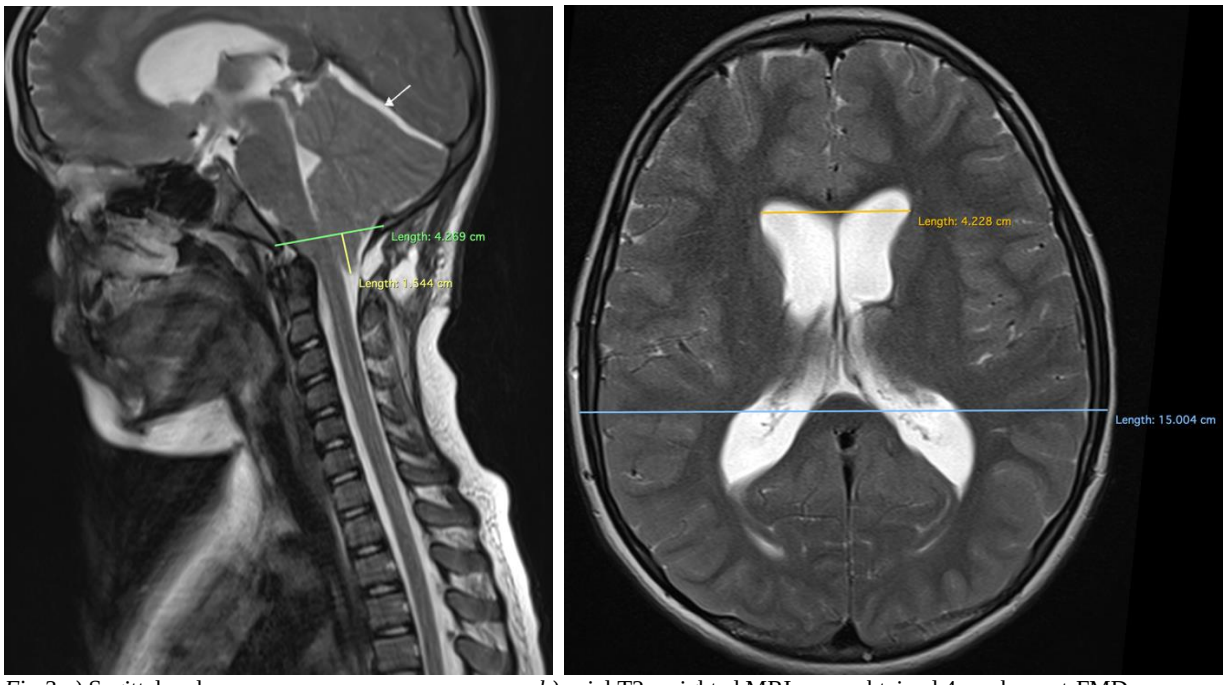


Fig.3 a) Sagittal and *b)* axial T2-weighted MRI scans obtained 4 weeks post-FMD.
(**a**) *(white arrow)* revealing infratentorial hygromas, pseudomeningocele and a cerebellar ptosis. (**b**) showing the enlargement of lateral ventricles and an Evans index estimated at 0.28. Of note, no suboccipital craniectomy was performed to alter the opisthion.

Pre operative and 4 weeks post-FMD scans, were compared. Cerebellar descent was evaluated in reference to McRae line. Of note, no suboccipital craniectomy was performed to alter the opisthion (Fig.2). With regard to this line, cerebellar ptosis was measured: 1.25 cm pre FMD and 1.544 cm 4-weeks post FMD, resulting in 0.29 cm ptosis that we believe is due to infratentorial hygromas pushing the cerebellum downward. The Evans index was used as a radiological measurement before and after decompression to evaluate ventricles enlargement.
A 0.28 index at readmission compared to 0.24 pre operatively was noticed. An ETV procedure was first considered but in the presence of an odontoid compression in the MRI, we believed that it would not provide a natural regulation of the CSF dynamics. Taking into account these radiological findings and clinical evaluation, the patient was started on high dosage of dexamethasone for 2 weeks and acetazolamide with gradual weaning after a month. Patient was discharged the next day. Resolution of ventriculomegaly, PMC and infratentorial hygromas was noted in the MRI obtained 1 month after readmission (Fig.4). A restoration of a previously obstructed cisterna magna is shown (Fig.4a). Evans index 0.24 (Fig.4b). At 1 year follow-up the patient is doing well and has not experienced headaches or vomiting. He is no longer taking acetazolamide. A repeated MRI at this time showed a reduction of the tonsils descent.

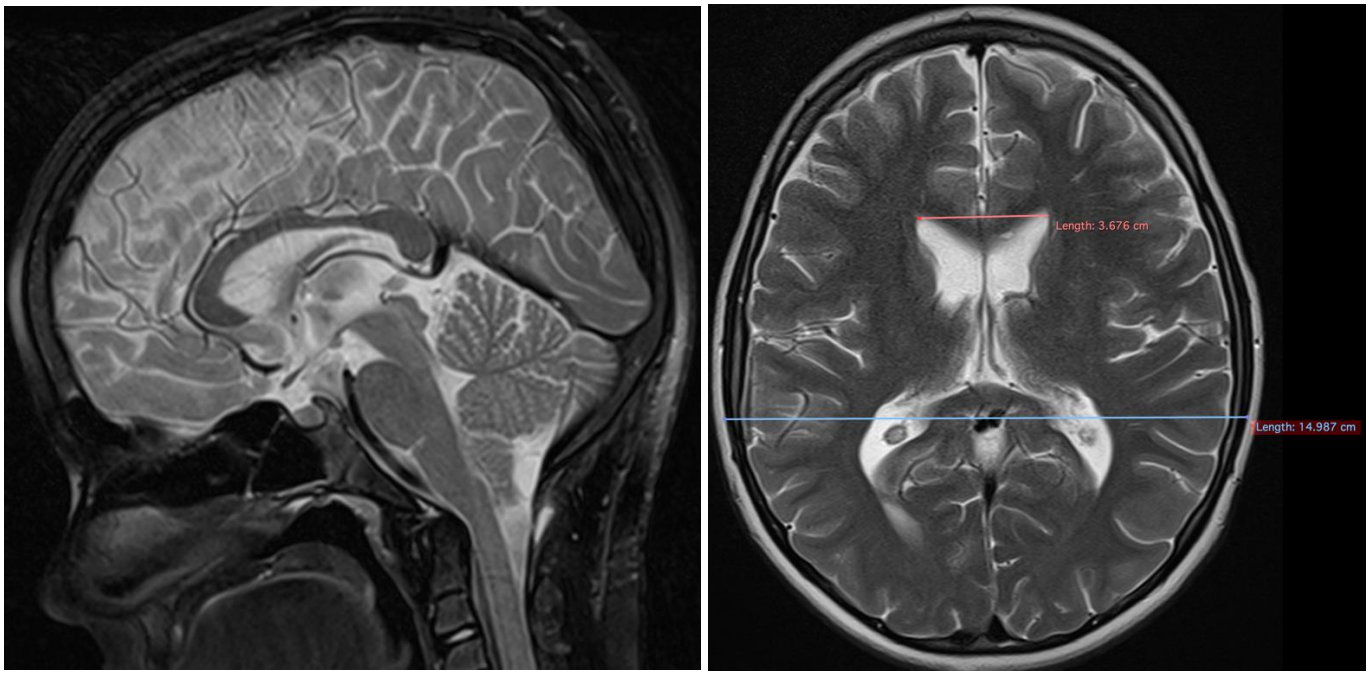


Fig.4 a) Sagittal *b)* axial T2-weighted MRI scans 1 month after readmission showing a restored cisterna magna and resolution of ventriculomegaly, PMC and infratentorial hygromas. Evans index 0.24.

Discussion

De novo hydrocephalus following posterior fossa decompression is a rare complication with an incidence varying between 5-7% as reported in literature [2]. Delayed postoperative subdural hygromas and de novo hydrocephalus following decompression for CM1 generate considerable debate regarding the best treatment option. Current treatment modalities include acetazolamide and dexamethason, EVD , ETV, VPS or wound re-exploration for further decompression. Our patient underwent a foramen magnum decompression with C1 laminectomy and dural splitting with arachnoid sparing. Congress of Neurological Surgeons in their systematic review and evidence-based guidelines for CM1 treatment in 2023 recommend that either PFDD or PFD can be utilised as first-line procedure in the CM1 patients to improve preoperative symptoms [4]. Dural splitting decompression is found to have satisfactory radiological outcomes in cisterna magna restoration and also offers similar clinical results as other techniques but with less complications [5]. Osborne-Grinter et al. in 2021 published a large meta-analysis describing a higher risk of CSF complications and reoperations in the arachnoid dissection group compared to arachnoid preservation[8]. Despite our meticulous technique, the patient subsequently developed de novo hydrocephalus, bilateral infratentorial extraaxial fluid collections and pseudomeningocele. Possible theories are described in literature to explain the pathophysiological mechanism behinde this phenomenon and are also mentioned by Bartoli et al in their multicentered study [2]. In our case we believe to be a post surgery arachnoid

tearing at the foramen magnum level, probably from a severe coughing, that through a slit valve mechanism is allowing CSF to accumulate in the subdural space and altering the CSF dynamics [2]. A conservative treatment with high-dose dexamethasone for 2 weeks and acetazolamide was initiated. Acetazolamide was gradually reduced after a month. When seen in his clinical follow-ups the symptoms had resolved and the MRI scans had returned to normal.

To the best of our knowledge, the largest published series of pediatric post CM1 decompression hydrocephalus and literature review is described by Jallo et al. 2018 presenting a series of their 5 pediatric patients who underwent CM1 decompression with placement of a dural graft complicated by posterior fossa hygromas and hydrocephalus that were successfully managed nonoperatively. In their review of literature they also include the nonoperative management described by Filis et al 2009 in a female 2 years of age with hydrocephalus and supra & infra tentorial hygromas after PFD for CM1 that became asymptomatic at one month. In addition, Constantini et al 2019 present another pediatric patient successfully treated with acetazolamide. Among these 7 patients aforeementioned, only two of them had not undergone arachnoid opening during surgery; they all experienced headaches, nausea and vomiting at hydrocephalus presentation; none of them had loss of consciousness, symptoms of herniation or signs of papilledema. The mean time to presentation of hydrocephalus was estimated at about 15 days. We add to the literature one more pediatric case that was managed nonoperatively with excellent long term outcome without the need of further CSF diversion.

Conclusion

Through this case report we share our successful experience with non operative treatment of subdural hygromas of posterior fossa, pseudomeningocele and acute hydrocephalus following FMD in pediatric patient. We believe that in selected cases, with no alteration of consciousness, acetazolamide and dexamethasone can be an effective treatment.

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