Coexistent Chiari Malformation and Idiopathic Intracranial Hypertension: Which Should Be Treated First? - Case Report and Review

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Abstract
Previous reports have described concomitant presentation of papilledema and Chiari malformation type 1 (CM1). The pathophysiology of each and their relationship remain poorly understood. As such, different management approaches regarding treating the CM1 or the pseudotumor initially have been described, with varying degrees of success. We report the case of an 11 year old girl with 6 mm tonsillar herniation, a cervicothoracic syrinx, and papilledema who underwent foramen magnum decompression with duraplasty (FMDD). Intracranial pressure (ICP) monitoring revealed elevated ICP before surgery, which resolved postoperatively. The patient had complete resolution of all symptoms and her papilledema and syrinx also resolved.

ABBREVIATIONS
CM: Chiari Malformation; CM1: Chiari Malformation Type I; IIH: Idiopathic Intracranial Hypertension; ICP: Intracranial Pressure; FMDD: Foramen Magnum Decompression and Duraplasty; MRI: Magnetic Resonance Imaging; CT: Computed Tomography; LP: Lumbar Puncture

INTRODUCTION
Previous reports have presented cases of concomitant Chiari malformation type 1 (CM1) and Idiopathic Intracranial Hypertension (IIH) [1-19]. These are two common but incompletely understood diagnoses with similar clinical presentations; [1-4,6] a pathophysiologic relationship between the two related to craniocephalic disproportion has been suggested [3]. In this setting, knowing which to treat first can be difficult. Some suggest treating IIH first, [1,10,13] while others have described treating IIH after failed foramen magnum decompression with duraplasty (FMDD) [3,6,7,20]. We present a patient with Chiari I malformation, syringomyelia and bilateral papilledema successfully treated with FMDD alone, raising provocative questions about common etiologies and the possibility that there are more undiagnosed cases of coexistent IIH and CM1 that may respond well to FMDD rather than medical treatment.

CASE PRESENTATION
This 11-year-old girl was diagnosed with migraine headaches at age 5, which resolved after 6 months of prophylactic headache medication. Frontal headaches returned 6 years later and became associated with intermittent blurry vision and photophobia but not emesis; they were not exacerbated by coughing, sneezing, or physical exertion. She was born at term with no other medical or family history.

Physical examination showed a healthy and neurologically intact 38 kg female except for the presence of bilateral papilledema, which was not present at age five. MRI of the brain (Figure 1) and spine (Figure 2) showed 6 mm of tonsillar displacement at the foramen magnum, syringomyelia between C5 and T9 (4.5 mm in maximal diameter), and no evidence of tethered spinal cord or low-lying conus. CT and MR venography of the head and neck were negative.

Continuous intracranial pressure (ICP) monitoring (Camino Intracranial Pressure Monitor, Integra Life Sciences Corp) for 24 hours prior to FMDD showed fluctuating but consistently elevated ICP between 25 and 40 mm Hg, when relaxed. Foramen magnum decompression, C1 laminectomy, dural opening, shrinking of the cerebellar tonsils, and expansile duraplasty with autologous pericranium was performed under a separate anesthetic. No
veil of tissue, as described by Tubbs [21] was appreciated as the cerebellar tonsils were separated.

The Camino monitor was left in place after Chiari decompression, confirming ICP normalization to the 5-15 mm Hg range. Her postoperative course was uneventful. She had complete resolution of her symptoms and papilledema on dilated fundoscopic examination at her 6 week office visit. Her postoperative MRIs are shown in Figure 3 and Figure 4. At 9 months’ follow up, papilledema remained absent and the syrinx was nearly resolved.

DISCUSSION

The relationship between IIH and CM has been established but is poorly understood, with some clinical overlap between the two. There is a predilection for females of child bearing age in both disorders [22,23]. Obesity is strongly associated with IIH, but this has not been described for CM1. Presenting symptoms of both CM1 and IIH include headaches, paresthesias, visual disturbances, visual loss, shoulder/arm pain, diplopia, motor weakness, and numbness, and these symptoms may be exacerbated by Valsalva maneuver in both cases [3]. Bilateral papilledema is the hallmark of IIH but has also been reported in CM1 [4,11,12,15-17,19,24].

Radiographs are not absolutely diagnostic for either entity. Although CM1 is defined by herniation of the cerebellar tonsils below the foramen magnum of 5 mm or more, [12] not all patients with this finding are symptomatic or require surgical intervention [25,26]. Conversely, there are patients not meeting radiographic criteria for CM1 who are symptomatic. Aiken and Barkovic have reported a mean tonsillar location in control patients between 0.7 to 1 mm above the foramen magnum with a standard deviation of 1.9 mm; based on this, a small percentage of control patients would be expected to have a tonsillar location of 2.8 to 3.1 mm below the foramen magnum. The correlation between the degree of tonsillar herniation and presence and severity of symptoms is imperfect.

For IIH, radiographic evaluation is less helpful. Absence of a mass lesion or other cause of increased ICP is required for the diagnosis of IIH as part of the modified Dandy criteria [23,27]. However, findings of optic nerve sheath enlargement and tortuosity, empty sella syndrome, reversal of the optic nerve head, transverse sinus stenosis, and flattening of the posterior sclera have been reported [8,28]. Tonsillar herniation has been reported to have a higher incidence in patients with IIH, ranging from 1.3% to 5.9%, compared to an incidence of 0.77% in the general population without CM1; [1-3,9] one author reported an eight-fold increase in CM1 in IIH patients [3].

Additional uncertainty regarding the management of concurrent CM1 and IIH stems from an incomplete understanding of the pathophysiology of each disorder and any relation between the two; it is unclear if CM causes IIH or if raised ICP from IIH causes tonsillar displacement. One argument is that tonsillar herniation causes alterations in CSF flow dynamics at
the foramen magnum. This could occur by a partial ball-valve mechanism, [29] with reduced dampening effects to pressure changes and altered CSF-brain compliance [22]. Abnormal torcular position and compression of draining veins, [16,30] as well as arachnoid scarring from rubbing of the tonsils against bone have also been hypothesized as causative factors [31,32]. Ultimately there is CSF obstruction at the foramen magnum [33] with creation of a CSF pressure gradient between the cranial and cervical compartments [29,34]. The demonstration of 4-8 mL CSF flow across the foramen magnum during periods of Valsalva, with 0.6 mL CSF flow caudally with transmission of resting cardiac impulse, [35] combined with a ball-valve mechanism allowing ingress of CSF into the cranial compartment without allowing sufficient egress into the cervical compartment would lead to increased ICP and IIH.

Acquired inferior tonsillar displacement has been reported, usually following lumbar CSF diversion [9,14,33,36,37]. Tonsillar herniation has also been reported in association with both supra- and infratentorial mass lesions and hydrocephalus, with resolution of the tonsillar herniation after primary treatment of the cyst, tumor or hydrocephalus [38-40].

Bejjani hypothesizes that relative craniocephalic disproportion could account for both; a small posterior fossa could give the tonsillar herniation associated with CM1 and compound the effects of brain engorgement from venous stasis or altered CSF absorption [3]. This idea of craniocephalic disproportion, as well as the association of CM1 with achondroplasia as well as osteochondrodysplastic and craniofacial disorders (e.g., Apert, Crouzon), is consistent with Milhorat’s theory of CM1 being a disorder of mesenchymal origin [9,12,22]. At the same time, the notion of impaired CSF absorption due to constriction of cerebral venous drainage and/or a dynamic effect both exerted on and by the superior sagittal sinus leading to increased brain turgor has also been postulated as a cause of both slit-ventricle syndrome and so-called normal volume hydrocephalus [30].

Given the current level of understanding regarding the pathophysiology of both disorders, and its chicken-and-egg nature, it is unclear if the primary treatment should be acetazolamide (for IIH) or FMDD (for CM1). To our knowledge, there is only one case of successful primary acetazolamide treatment for concomitant CM1 and IIH leading to resolution of tonsillar herniation [17]. Other reports in which the IIH was treated first have yielded satisfactory results with regards to the IIH, but without change in the CM; [1,10,13] one author concluded that the two entities were coincidental [31]. Additional reports have described development or exacerbation of CM1 following treatment for IIH, requiring subsequent treatment with FMDD [1,4,5,16].

Other cases of concomitant CM1 and IIH describe primary CM1 treatment with resolution of papilledema, besides this case report [11,16,18,19]. In two of these, ICPs were elevated preoperatively and normal postoperatively [11,18]. In another, FMDD effected resolution of papilledema, but the patient developed optic nerve atrophy and residual visual field deficit [19].

Fagan described a “Chiari Pseudotumor Cerebri Syndrome” in which patients developed recurrent Chiari-like symptoms, elevated opening pressure on LP, and transient relief with high volume CSF drainage following FMDD, ultimately proceeding to shunting. They postulated that FMDD altered CSF compliance at the foramen magnum, providing temporary relief, but that the underlying IIH remained until CSF diversion, [6] a theory also espoused by Bejjani. In his series of 6 patients of FMDD for CM1, however, none had papilledema; as the dura became fibrotic following surgery, the compliance was altered further, and all six proceeded to CSF diversion. He postulated that they had IIH without papilledema preoperatively [3]. Additional treatment failures with FMDD have been described, requiring subsequent pharmacologic treatment in one case [7] and ventriculoperitoneal shunting after elevated ICPs 2 years postoperatively were found in another [20].

We present a case of coexistent papilledema, CM1 and syrinx successfully treated with FMDD, raising the possibility that FMDD should be considered more frequently as the treatment of choice for concomitant papilledema and CM1, particularly when syrinx is present. Unfortunately, in our review, only in one case was spinal cord syringes present [10].

In the absence of a syrinx, the evaluation and clinical judgment are critical. Workup and treatment of hydrocephalus is the first priority, but if negative, the presence of papilledema, together with the data likely accrued to this point in the workup (i.e., referable signs and/or symptoms, tonsillar herniation, papilledema, and absence of hydrocephalus) leads to several options, including ICP monitoring, acetazolamide trial, and FMDD, and raising the fundamental question raised by this case. Additional findings such as spinal cord syringes, scoliosis, or bulbar signs and symptoms may favor FMDD over the others [10]. Proceeding with FMDD, as in this case, would address the CM1 and the papilledema (or IIH) can be followed postoperatively. The management would revert to that as for IIH without tonsillar herniation, including LP, acetazolamide, optic nerve sheath fenestration, and shunting, although FMDD may decrease ICP through several mechanisms, as discussed earlier, [3,6] and obviate the need for CSF diversion. Although it has been argued
that FMDD in the setting of elevated ICP leads to an increased risk of CSF leak, [10] we believe this risk to be small.

CONCLUSION

In this case, we proceeded directly with FMDD; this was largely driven by the presence of the cervicothoracic syrinx. It would also be reasonable to proceed with FMDD in the presence of neurological deficits, especially bulbar signs. As the preceding discussion illustrates, the management of concomitant Chiari I malformation and papilledema can be complex. This case and review of the literature are presented in the hope that it may guide management of similarly challenging cases.

REFERENCES


