A child with unilateral abducens nerve palsy and neurovascular compression in Chiari malformation type 1 resolved with posterior fossa decompression: illustrative case

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BACKGROUND Unilateral cranial nerve (CN) VI, or abducens nerve, palsy is rare in children and has not been reported in association with Chiari malformation type 1 (CM1) in the absence of other classic CM1 symptoms.

OBSERVATIONS A 3-year-old male presented with acute incomitant esotropia consistent with a unilateral, left CN VI palsy and no additional neurological symptoms. Imaging demonstrated CM1 without hydrocephalus or papilledema, as well as an anterior inferior cerebellar artery (AICA) vessel loop in the immediate vicinity of the left abducens nerve. Given the high risk of a skull base approach for direct microvascular decompression of the abducens nerve and the absence of other classic Chiari symptoms, the patient was initially observed. However, as his palsy progressed, he underwent posterior fossa decompression with duraplasty (PFDD), with the aim of restoring global cerebrospinal fluid dynamics and decreasing possible AICA compression of the left abducens nerve. Postoperatively, his symptoms completely resolved.

LESSONS In this first reported case of CM1 presenting as a unilateral abducens palsy in a young child, possibly caused by neurovascular compression, the patient’s symptoms resolved after indirect surgical decompression via PFDD.

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KEYWORDS Chiari malformation type 1; cranial nerve VI palsy; abducens nerve; pediatric neurosurgery

In cranial nerve (CN) VI, or abducens nerve, palsy, lateral rectus muscle paresis limits abduction of the affected eye and causes esotropia due to unopposed activity of the medial rectus.1 In adults, this can be associated with damage or compression due to posterior fossa brain tumors, stroke, aneurysms, and trauma.2 In children, unilateral CN VI palsies are rare, but bilateral palsies can be related to tumors, elevated intracranial pressure, congenital malformations, postinfectious or postvaccination inflammatory disease, post–lumbar puncture, multiple sclerosis, or genetic syndromes such as Miller Fisher syndrome and Duane retraction syndrome.7–13 In both children and adults, “nonlocalizing” abducens nerve palsies have also been described, often associated with increased or decreased intracranial pressure.14

However, Chiari malformation type 1 (CM1) has never been reported to present with isolated abducens nerve palsy in the absence of other symptoms. CM1 is a disorder involving a compromised cisterna magna in which free craniospinal cerebrospinal fluid (CSF) flow is limited due to herniation of the cerebellar tonsils. This is often compounded by a congenitally small occipital bone as well as intradural anomalies.15 Symptomatic CM1 usually presents with headache, neck pain, ataxia, and vertigo.16,17 Some patients also experience cognitive and affective symptoms, or upper-extremity symptoms if syringomyelia

ABBREVIATIONS AICA = anterior inferior cerebellar artery; CISS = constructive interference in steady state; CN = cranial nerve; CM1 = Chiari malformation type 1; CSF = cerebrospinal fluid; CT = computed tomography; ICP = intracranial pressure; MRI = magnetic resonance imaging; PFDD = posterior fossa decompression with duraplasty; POD = postoperative day; VA = vertebral artery.

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is present. Asymptomatic CM1 is also well reported.\textsuperscript{18,19} The few cases of CN palsies and divergence palsies reported in association with CM1 have included additional classic signs and symptoms such as headaches, dizziness, and papilledema.\textsuperscript{20–22} We present the unique case of a toddler with acute, isolated, unilateral CN VI palsy and no other symptoms, whose imaging revealed a vessel loop near the abducens nerve along with tonsillar ectopia. Following posterior fossa decompression with duraplasty (PFDD), the patient exhibited complete resolution both clinically and radiographically.

Illustrative Case
Presentation and Workup
A previously healthy 3-year-old male presented with sudden-onset, spontaneous eye-crossing episodes for 1 day, which had been triggered by looking at distant objects. Outpatient pediatric ophthalmology examination demonstrated left incomitant esotropia (25 prism diopters esotropia at distance and 15 prism diopter esotropia at near, worse in left gaze). No optic nerve swelling was seen. This was concerning for a left CN VI palsy, and the patient was sent to the emergency department.

On physical examination, the patient was alert and oriented, with pupils equal, round, and reactive to light. Significant incomitant esotropia worse in left gaze was seen (Fig. 1). The left eye was unable to abduct past midline on leftward gaze, graded as an abduction tropia worse in left gaze was seen (Fig. 1). The left eye was unable to abduct past midline on leftward gaze, graded as an abduction deficit of \( \frac{1}{4} \) (abduction one-quarter of the distance toward the lateral canthus). The patient demonstrated a right gaze preference, fixing and following using his right eye and keeping the right half of his face in the midline with a slight leftward head turn. The eyes remained straight and conjugate in the rightward gaze. Other extraocular movements of the left eye appeared full, and his right eye moved in all directions. There was no ptosis.

Bedside fundoscopic examination revealed no definite papilledema. RetCam photographs (Fig. 2) revealed a scalloped border to the left optic nerve but no significant elevation of overlying tissue, as well as sharp vessels, with no definitive swelling of either optic nerve. Optical coherence tomography was considered to rule out increased intracranial pressure (ICP) in the absence of definite papilledema, but this was forgone given the patient’s age.

The remainder of his face was symmetrical, his hearing was intact, and there was no torticollis. He demonstrated 5/5 strength in all four extremities with normal muscle bulk and tone, and sensation was intact on all extremities. Gait and balance were normal, and there was no dysmetria. Deep tendon reflexes were 2+ in the bilateral upper and lower extremities, with normal flexor plantar responses. He appeared nondysmorphic, was talkative and fluent, and playful and interactive, following simple instructions with age-appropriate behavior. The patient and his family denied eye pain, headache, nausea, vomiting, gait abnormalities, decreased activity level, weight loss, decreased appetite, recent trauma, or delay in developmental milestones. He had a history of multiple viral upper respiratory infections but no rashes, tick bites, joint pains, or history of torticollis. His family medical history was notable for pituitary mass, intraocular lymphoma, and Hashimoto’s disease, colon cancer, ventricular septal defect, and hyperlipidemia. Diagnostic laboratory analyses were normal, including complete blood count, complete metabolic panel, C-reactive protein, erythrocyte sedimentation rate, Lyme antibodies, and blood lead level.

Noncontrast computed tomography (CT) and magnetic resonance imaging (MRI) of the brain demonstrated low-lying cerebellar tonsils with 13 mm of ectopia beneath the basion-opisthion line and crowding of the cisterna magna, consistent with CM1 (Fig. 3). There was no hydrocephalus. The pons, petrous temporal bone, abducens, and other CNs appeared otherwise normal with no postcontrast enhancement or edema. MRI of the orbit demonstrated prominent CSF in the optic nerve sheath with normal-appearing lateral and medial rectus muscles. There was no posterior globe flattening or optic nerve head protrusion. There was no syringomyelia on MRI of the spine.

Patching the right eye for 1 hour per day was initiated to prevent amblyopia of the left eye. Imaging and laboratory work ruled out, or made unlikely, neoplastic, infectious, and inflammatory etiologies. Increased intracranial pressure in the setting of CM1 was suggested, and
surgical decompression of the posterior fossa was discussed. Given the absence of other classic Chiari symptoms and the atypical presentation not definitively attributable to CM1, the patient was discharged with outpatient observation.

Over the next 2 weeks, he developed a larger compensatory face turn, worsened esotropia, and an abduction deficit grading of −4 (no movement from the primary position toward the lateral canthus). MRI was repeated, with additional constructive interference in steady state (CISS) sequences revealing a prominent left anterior inferior cerebellar artery (AICA) and left vertebral artery (VA) crossing near the cisternal segment of the left abducens nerve in the prepontine cistern. The possibility of a compressive etiology for the palsy was considered.

Risks and benefits of PFDD were discussed with the family as an option to potentially restore normal craniospinal CSF dynamics in the posterior fossa and indirectly decompress the AICA–CN VI complex. Given the atypical nature of this suggested pathophysiology, however, PFDD was offered with caution, and the family was counseled that if the CN palsy persisted, it could ultimately be attributable to either an incorrect etiological hypothesis or an inadequate nerve recovery from prolonged traction or pressure from the AICA and posterior fossa intracranial pressure. Alternatives to PFDD were also discussed, including continued observation, lateral rectus muscle surgery, and even a high-risk microvascular decompression via direct skull base approach. The family obtained a second opinion from pediatric neurosurgery at another children’s hospital, who agreed with our assessment, and the patient was scheduled for surgery.

Surgery
The patient underwent PFDD via the standard approach at our institution (suboccipital craniectomy; C1 laminectomy; exploration of the arachnoid planes, basolateral cisterns, and obex, with expansile allograft duraplasty). Intradural findings included tonsillar hypertrophy and pale, gliotic tonsillar tips. These were separated gently until free-flowing CSF was seen coming from the obex and choroid plexus, without tonsillar coagulation. There were no complications, and neurophysiological monitoring of somatosensory evoked potentials remained stable.

Follow-Up
Postoperatively, the left CN IV palsy was 50% improved by postoperative day (POD) 2 with a restored ability to abduct the left eye past midline. Postoperative CISS MRI demonstrated successful posterior fossa decompression and a larger space between the AICA loop and the left CN VI (Fig. 4). By POD 14, the patient demonstrated complete resolution of his left CN VI palsy. He had a slight residual small-angled intermittent esotropia. He exhibited good vision in both eyes with no amblyopia or head turn. His mother reported that his overall gaze and head position were approaching baseline (Fig. 5). At his latest neurosurgical follow-up 5 months postoperatively, he continued to exhibit full abduction, and his mother reported that his gaze and posture were completely back to baseline.

Patient Informed Consent
The necessary patient informed consent was obtained in this study.

Discussion
Observations
We present the first reported pediatric case of a unilateral, isolated abducens nerve palsy in the setting of concurrent radiographic CM1 and possible microvascular compression, with no hydrocephalus or clinical papilledema and no classic Chiari symptoms. PFDD was performed, ultimately leading to full symptom resolution and radiographic improvement of the AICA-abducens proximity seen preoperatively. We discuss the possibility that PFDD enabled an “indirect decompression” of the cisternal abducens-AICA complex by restoring normal craniospinal CSF flow and thereby relieving compression on the left abducens nerve. This is illustrative of the complex CSF dynamics present in CM1. Moreover, the absence of papilledema on clinical examination cannot rule out subclinical ICP effects leading to unique clinical presentations.

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FIG. 3. A: Preoperative midsagittal noncontrast CT scan demonstrating 13 mm of downward herniation of the cerebellar tonsils. B: Preoperative midsagittal T1-weighted MRI with a similar degree of downward tonsillar herniation. C: Axial T2 CISS MRI demonstrating an aberrant vessel loop thought to be arising from the left AICA and contacting the left CN VI.

FIG. 4. A: Postoperative midsagittal T1-weighted MRI with significant posterior fossa decompression and improvement in tonsillar herniation through the foramen magnum. B: Axial T2 CISS MRI demonstrating decompression of the left CN VI with an increase in the space between the nerve and aberrant vessel loop.
Studies have demonstrated that the AICA or other vessel branches traverse the ventral surface of the abducens nerve’s cisternal segment in up to 75% of cases. Others examining the origin of the abducens nerve from the pontomedullary sulcus through Dorello’s canal have also found frequent instances of contact between the abducens nerve and the AICA. Symptomatic neurovascular compression of the abducens nerve is rare. A few reported cases have implicated compression from the AICA or a dolichoectatic VA. Given the difficult skull base approach to the preptontine cistern and the rarity of the condition, microvascular decompression of the abducens nerve is seldom undertaken, in contrast to more commonly decompressed pathologies such as hemifacial spasm and trigeminal neuralgia.

In our case, the radiographic finding of AICA and VA proximity to the cisternal abducens nerve was critical in the clinical decision-making but only in the context of suspected CM1-induced flow abnormalities contributing to possible neurovascular compression. Therefore, the surgery chosen was not a direct microvascular decompression but rather an “indirect” decompression via PFDD. The long course of the abducens nerve leaves it vulnerable to ICP changes, and several cases of nonlocalizing abducens nerve palsy related to CSF dynamics have been reported in association with increased or decreased ICP. These include cases of CSF diversion and hydrocephalus, as well as several cases of central nervous system hypotension following lumbar puncture.

Lessons
We present the first reported pediatric case of a unilateral, isolated abducens nerve palsy in the setting of radiographic CM1 and a tight AICA–CN VI microvascular complex. Despite the absence of classic Chiari symptoms, frank papilledema, or hydrocephalus, PFDD was associated with complete symptom resolution and radiographic resolution of the AICA–CN VI complex. We discuss the possibility of PFDD acting to normalize CSF flow dynamics and indirectly decompress CN VI. This discussion is illustrative of the complex CSF dynamics within CM1, which can lead to unique clinical presentations.

References


Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Shao, Anderson, Svokos. Acquisition of data: Doberstein, Kozel, Shao, Anderson, Harappanahally, Svokos. Analysis and interpretation of data: Doberstein, Shao, Anderson, Harappanahally, Svokos. Reviewing submitted version of manuscript: Doberstein, Kozel, Shao, Anderson, Harappanahally, Svokos. Approved the final version of the manuscript on behalf of all authors: Doberstein. Administrative/technical/material support: Svokos. Study supervision: Shao, Svokos.

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