The Role of Spontaneous Venous Pulsations in the Diagnosis of Adult Chiari Malformation

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Citation

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Abstract

Objective: To perform neuro-ophthalmologic evaluations in patients with symptomatic Adult Chiari Malformations (ACM).Methods: Observational case series of 56 patients with symptoms and neuro-imaging consistent with ACM. Comprehensive histories and complete neuro-ophthalmic examinations were performed. Findings: Sixteen men and forty women were evaluated. The mean age was 41 years old (range 8 - 73 years old). The most common visual complaint was transient visual obscurations lasting seconds (n= 34). Pain or fullness behind the eyes was present in 32 patients. Forty-eight patients had headaches. Other frequent symptoms included neck pain, dizziness, facial pain, and numbness. Fundus examination was remarkable for absent spontaneous venous pulsations in one or both eyes in 28 patients. Interpretation: Adult Chiari Malformation (ACM) presents with characteristic symptoms that may be overlooked or misdiagnosed. Neuro-ophthalmic evaluation is helpful in ruling out other entities and identifying characteristic absent venous pulsations.

INTRODUCTION

The diagnosis of tonsillar herniation has increased as the use of Magnetic Resonance Imaging (MRI) has become widespread. Hans Chiari first described the various alterations in the posterior fossa that became known as the Chiari Malformations. Type I was defined as elongation of the cerebellum into cone shape projections into the spinal canal. Chiari's original cases all had hydrocephalus. The first case of a Chiari Type I malformation in an adult without hydrocephalus was described by Aring in 1938. Larger series were published in the 1960s and 1970s and the term Adult Chiari Malformation (ACM) was coined for type I malformations in adults without hydrocephalus. [1]

The sagittal views provided by MRI demonstrate tonsillar ectopia of at least five millimeters (ACM) in 0.56-0.77 % of scans performed (Figure 1). The prevalence of symptomatic ACM is more difficult to characterize. The disorder occurs more commonly in women (3:1 gender ratio). The mean age of onset is approximately 25 years of age. The delay in diagnosis is at least five years in most studies. Symptoms include headache, imbalance, transient visual obscurations, intracranial noises and retro-orbital ache (Table 1) [2,3,4,5,6,7,8,9,10,11,12] . Mueller and Oro, in a study of 265 patients, found that the five most frequently reported symptoms were headache (98%), dizziness (84%), difficulty sleeping (72%), weakness of an upper extremity (69%) and neck pain (67%). This study included both patients with and without syringomyelia. [12] Reported clinical signs have varied widely (Table 2). [13,14,15,16,17,18,19,20,21,22]

Figure 1

Table 1: Reported Symptoms of Adult Chiari Malformation (ACM)

Ocular:
Transient visual obscurations
Retrobulbar ache
Photophobia
Diplopia
Visual field loss

Otologic:
Dizziness/ Vertigo
Tinnitus
Decreased hearing
Ear pressure
Vertigo
Hyperacusis

Lower Brainstem:
Dysphagia
Dysarthria
Sleep apnea
Syncope
Shortness of breath
Palpitations

Cerebellar:
Unsteady gait
Poor coordination
Tremor
Impaired fine motor skills

Sensory:
Occipital headaches
Cervical pain
Facial and acral numbness
Paresthesias
Dysethesias

Other:
Weakness
Fatigue
Diminished memory
Nausea/Emesis

Figure 2

Table 2: Reported Clinical Signs of Adult Chiari Malformation (ACM)

Bilateral disc edema
Absent venous pulsations
Afferent dysfunction
Cranial nerve palsies
Nystagmus
Sensoineural hearing loss
Abnormal vestibular testing
Impaired gag
Vocal cord paralysis
Hypoglossal nerve palsy
Spinal nerve palsy
Dysmetria
Ataxia
Analgesia
Impaired proprioception
Weakness
Spasticity
Hyper-reflexia
Associated reported findings
Scoliosis
Glossopharyngeal neuralgia
Neurologic deterioriation following neck injury

Ophthalmic findings in ACM have been poorly characterized but have included papilledema, decreased acuity, extraocular muscle palsy, convergence or divergence paresis, skew deviation, nystagmus and absent venous pulsations. [21] Kumar and colleagues performed vestibular evaluation on 77 confirmed ACM patients symptomatic of hearing loss, dizziness, and tinnitus and found horizontal spontaneous nystagmus in over one third of patients. Other findings included vertical upbeat nystagmus, downbeat nystagmus, saccadic dysmetria, optokinetic nystagmus, and smooth-pursuit impairment. [22]

The cause of ACM is multifactorial. Craniocephalic disproportion is common. The skull dimensions can be congenitally small as seen with craniosynostosis. Increased intracranial pressure, as found in idiopathic intracranial hypertension, is also associated with ACM. Lumbar punctures or lumboperitoneal shunts can cause tonsillar descent. [23,24,25,26,27,28,29]

ACM becomes symptomatic due to two distinct effects: 1) direct compression of nervous tissue and 2) a valve effect by the tonsils at the foramen magnum. [30] The direct compression leads to lower brainstem and cerebellar signs. The valve effect alters normal neural hydrodynamics. Normally, cerebral blood volume increases with systole and the relative brain engorgement is accommodated by the flow of cerebrospinal fluid into the spine. In ACM, intracranial pressure transiently increases and brain compliance is altered. Not surprisingly then, many of the symptoms are similar to IIH and even worsen with Valsalva maneuver. [23,24,25,26,27,28,29,30,31,32]

Approximately 3500 surgical decompressions are performed per year in the US for symptomatic ACM. Arora et al. reported that the most dramatic symptomatic improvements in a group of Chiari I patients following surgical decompression were spasticity, neck pain, and cerebellar signs. [33]

METHODS

This is a prospective observational case series of consecutive patients referred for standard neuro-ophthalmic evaluation of ACM. Inclusion criteria included tonsillar descent of at least five millimeters and characteristic symptoms.

A comprehensive neuro-ophthalmologic evaluation was performed that included best-corrected visual acuity, color vision (pseudoisochromatic color plates), pupillary size and function, anterior segment and posterior segment examination. The extraocular motility examination included assessment for cranial nerve dysfunction, nystagmus and vestibular ocular reflex impairment. In addition Humphrey visual fields (HVF 30-2) and Heidelberg Retinal Tomography (HRT) were performed to detect subclinical optic nerve dysfunction. (All examinations were performed by KPC).

FINDINGS

56 patients with symptomatic Adult Chiari Malformation (ACM) were evaluated. The gender ratio was consistent with previous studies (40 women: 16 men). The mean age was 41.2 years old with a range of 8 to 73 years. This mean age is older than previous reports.

The most common visual symptom was transient visual obscurations (TVOs) lasting seconds (n= 35). The TVOs were unilateral (n = 3) or bilateral (n = 31), for a total of 65 affected eyes. Pain or fullness behind the eyes was present in 57 eyes of 32 patients, with 7 patients having unilateral discomfort. The discomfort increased with eye movement in some cases. 14 patients had floaters in the absence of any vitreous detachment. Neck pain (n = 40), dizziness (n = 35), numbness (n= 26) and facial pain (n = 12) were also common complaints. A wide variety of other symptoms were reported (Table 3). The severity of symptoms varied from moderate to extremely limiting.

Figure 3

Table 3: Neurologic Symptoms in Our ACM Series

Symptom	No. of patients	Percentage
Transient visual obscurations (TVOs)	34	60.7%
Bilateral	31	55.4%
Unilateral	3	5.6%
Retrobulbar ache	32	57.1%
Bilateral	25	44.6%
Unilateral	7	12.5%
Headache	48	85.7%
Neck Pain	40	71.4%
Dizziness	35	62.5%
Numbness	26	46.4%
Facial Pain	12	21.4%
Visual Field Defects	0	0.0%
Diplopia	0	0.0%

Persistently decreased vision due to afferent dysfunction was not found. Fundus examination was remarkable for lack of venous pulsations in 51 eyes of 28 patients (50%), 1+ SVP in 24 eyes of 17 patients (7 unilateral) and 2 + SVP in 22 eyes of 14 patients (Table 4)(Figure 3) . One patient had 3+ SVP in one eye. Eight patients had dampened SVPs evoked only with digital pressure or Valsalva maneuver. Mild hyperemia with telangiectatic vessels was noted in 3 patients. No patients demonstrated disc edema, exudates, gliosis or reverse vessel taper. 3 patients demonstrated nystagmus, but no patients demonstrated cranial nerve deficits.

Figure 4

Table 4: Neuro-Ophthalmic Signs in Our ACM Series

Optic Nerve Dysfunction
Decreased visual acuity
Abnormal color vision
Visual field defect
Optic disc edema
Evidence of prior optic disc edema
Lack of spontaneous venous pulsation
Cranial neuropathy
Nystagmus

INTERPRETATION

Spontaneous venous pulsations are best seen with a direct ophthalmoscope or hand held lens (90 or 78 diopter) at the slit lamp. A vein overlying the optic disc, usually at the depth of the cup, is observed for brief collapse of the walls (pulsations). The physiology behind the pulsations is debated (Table 5). Pulsations are increased in patients with significant elevated intraocular pressure. Mydriatics have historically been noted to increase pulsations, but this is now controversial. In normals, digital pressure or Valsalva maneuver results in a transient increase in pulsations following release of the pressure or termination of the Valsalva. Intra-ocular hypotension and increased intracranial pressure have been associated with absent pulsations. Jugular pressure causes transient increase in intracranial pressure and has been noted to lead to suppression of venous pulsations.

Figure 5

Table 5: Theories on the Pathophysiology of the Retinal Venous Pulsation ,,

Name/Author	Description	Evidence
Classical theory(Baillart)	Intraocular pressure rises and exceeds venous pressure during systole, causing veins to collapse. RVP then exceeds IOP, making the vein expand.	
Jacks/Miller 2003	Pressure gradient variations along the retinal vein, secondary to difference in pulse pressure between the intraocular space and CSF, cause pulsations as the retinal vein traverses the lamina cribrosa. The gradient is between the central retinal vein and the intraocular retinal veins.	Increased ICP causes SVPs to stop. As ICP rises, the intracranial pulse pressure rises to equal the intraocular pulse pressure.
Meyer-Schwickerath	IOP oscillates at a pressure significantly higher than external pressure. Increasing the resistance in the vessel as it leaves the vessel, or increasing pressure upstream from the veins, increases vessel pressure required for pulsation.	Modeling experiments
Conrad	Vessels collapse when the difference between chamber and exterior pressure exceeds a certain amount.	Modeling experiments
Levine	Assume: 1) Inflow to retinal veins is constant. 2) Outflow varies during cardiac cycle because IOP is oscillating (and transmitting to CRV) at a higher amplitude than the CSF is oscillating. The radial blood flow distending the veins obeys a diffusion equation.	Flow within the vessels remains constant during the cardiac cycle.
Coccius	Veins collapse during systole (when arteries expand) due to transcapillary pulse transmission not occurring. Blood flows into eye and the resultant IOP compresses the veins.	
Modified theory of Baillart	Transmural vein pressure gradient periodically changes direction. Peripheral veins are surrounded by retinal fibers and are less likely to collapse. Blood flow velocity is higher in the CRV than in smaller vessels, producing a Bernoulli effect that approximated the CRV to IOP	

Hedges and colleagues further characterized the optic nerve head characteristics that limit the ability to detect venous pulsations in normals. If the veins were easily visible and a normal cup to disc ratio was present, spontaneous venous pulsations were present in 73%, if the arteries or gliosis partially obscured the veins or the cup to disc ratio was less than 0.1, spontaneous venous pulsations were present in 43 %. If the arteries or gliosis completely obscured the veins or the optic nerve is congenitally elevated and full, absent pulsations were very prevalent.

Adult Chiari Malformation (ACM) is diagnosed by performing sagittal MRI in patients with suspicious symptoms. Because the combination of vague symptoms can be difficult to objectively quantitate, many patients' symptoms are mistakenly labeled as psychogenic. This delay to diagnosis accompanies an increasing patient frustration. In this series of symptomatic patients with Adult Chiari Malformation, the most common visual symptoms were transient visual obscurations and retro-orbital ache. The most common objective finding was lack of venous pulsations even with Valsalva maneuvers. One possible explanation for this could be that the increased intracranial pressure in ACM leads to suppressed spontaneous venous pulsations.

A complete neuro-ophthalmic evaluation was helpful to exclude other diagnoses and confirm characteristic findings. A team of subspecialists, including a neuro-ophthalmologist, neurologist, neuro-otologist, and neurosurgeon best manages adult Chiari malformation.

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