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2010 Cranial Academy Proficiency Examination Case

ID: A.M. a 34 year-old Caucasian female DOB: 7/21/1974

CC: "Migraine headaches from Arnold Chiari Syndrome"

HPI: 34 year-old female with a history of hypothyroidism, GERD and Thoracic Outlet Syndrome presented 2/10/2009 to my private OMT practice suffering from migraine headaches for more than eight years. The headaches began four months after the birth of her only child when she was 26 years old. The daily headaches are right-sided, at times with auras, lights, double and blurred vision as well as pinprick sensations on her head. She described the pain as at times pounding, throbbing and pulsing along with a tight band around her head. She experiences severe nausea with vomiting on average 6 times on most days. She is more sensitive to light than sound and her ears feel "plugged up." Her sleep is disrupted at 1-2am by onset of headaches and she often wakes up in the morning with a headache. Her pain ranges from 4/10 to >10/10 at worst, although she has to work despite the pain. She is worse from repetitive noises such as tapping as well as chemicals, perfumes, bleaches, dyes and even strong natural scents such as lavender or rosemary. She uses Norco to take the edge off of the pain. In the past, she has tried Vicodin, Neurontin, Imitrex, Inderal, Prozac, Lyrica and Elavil without improvement. She was originally diagnosed with Trigeminal Neuralgia at age 27 and botox was tried without effect. Later she was diagnosed with migraines. Five months prior to her initial presentation in my office she first learned that her migraines were caused by a Chiari 1 Malformation, diagnosed by MRI in 2008, although past records included an MRI in 2002 which showed mild "Arnold Chiari malformation."

Past History

Medical: Hypothyroidism diagnosed 4 months after the onset of her migraines; Hiatal Hernia with GERD and Esophagitis, Thoracic Outlet Syndrome, Hepatitis C diagnosed at age 26, s/p PEG Interferon, now with undetectable viral load.

Surgical: Planned Cesarean Section with Tubal Ligation with concurrent Cholecystectomy in 2001

Social: Married with 8 year old daughter, works as a nanny. Patient's father was drug dealer, patient suffered from significant sexual abuse from a very young age. Addicted to methamphetamine age 10-24, heroin from age 20-24. Tobacco use from early teens to age 24. Entered substance abuse recovery program at age 24 and has been drug free since, except for prescription medicines.

Allergies: Penicillin which caused "huge hives and trouble breathing" and Codeine had similar but milder reaction.

Medications: Norco 10/325 TID, Synthroid 75mcg QD, Prilosec 20mg QD, Benadryl 50mg PRN insomnia

Trauma History: Sternal fracture at age 13, fall on tailbone as a younger child, sexual abuse in early childhood.

Birth History: she was the 1st of 2 children for her mother, may have been breech with cord around neck

OB/GYN History: G5P1041, only delivery via planned cesarian section for concurrent cholecystectomy and tubal ligation, due to gallbladder problems during pregnancy. Birth control – s/p tubal ligation.

Diet: 2 cups coffee per day, some soda, 5-6 glasses of water per day, regular fruit and salads, denies eating fast food

Exercise: uses treadmill approx. 3x per week

Family History: limited, both parents adopted, Father deceased. Mother, siblings healthy.

Review of Systems

HEENT: double vision, blurred vision, more sensitive to light, with ringing and sensation of plugged ears as in HPI;

no sore throat, some neck stiffness Respiratory: no shortness of breath, cough or wheezing

CV: no palpitations, no chest pain GI: heartburn, nausea, vomiting; no constipation, diarrhea, or bloody stool

GU: no burning on urination Neuromusculoskeletal: red/purple fingers at times

Psychological: sleep difficulty waking during night from headaches

Reproductive: heavy periods with cramping/pain

Physical Exam

Vitals: afebrile, BP 128/74 P 72 R13

General: heavy set female, cooperative and in no acute distress, non-toxic

HEENT: normocephalic, atraumatic *Neck:* supple without thyromegaly or lymphadenopathy

Heart: regular rate and rhythm, without murmurs *Lungs:* clear to auscultation

Abdomen: soft, non-tender, non-distended, positive bowel sounds

Extremities: no clubbing, cyanosis, or edema

Neurological: alert and oriented x 3, cranial nerves II-XII intact, motor 5/5 in all four extremities, sensation intact,

DTRs brisk 3+ and symmetric x 4 extremities

Musculoskeletal: gait balanced and easy, full range of motion in all extremities

Osteopathic Structural Exam: Cranium: Flexion pattern cranium, SBS compression and poor amplitude of PRM.

The core-link was extremely tight from C2, pulling inferiorly on the tentorium cerebelli and superiorly from the sacrum. There was significantly diminished motility of the ventricles as well as decreased amplitude of CSF fluctuation. Right temporal bone internally rotated. Right sphenoid greater wing inferior. Right S-S pivot compression. Cerebellum inferior with flattening of the tentorium cerebelli. Right condylar compression with opisthion rotated to the left. Falx cerebelli tension through foramen magnum. Inferior tension through the falx cerebri, flattening the parietals into extreme external rotation.

Cervical: significant paraspinal muscle spasm with right rotation throughout the cervical vertebrae, except C6-C7 which were RISI. The OA is EsrRI.

Thoracic: diaphragm increased inhalation phase with increased kyphosis of thoracic spine and flexion at T1.

Chronic right-sided T5-9 facilitation evidenced by tight, ropy, muscles.

Lumbar: flattening of L1-3 lordosis and L-S compression, Left psoas spasm

Sacrum: S2 intraosseous strain with decreased primary respiratory motion and decreased ROM with thoracic respiration. *Pelvis:* Right anterior innominate *Ribs:* right 1st rib elevated in inhalation. Right ribs 5-9, inhalation dysfunction.

Upper extremities: Right clavicle depressed with sterno-clavicular compression.

Right worse than left Radial-Ulnar interosseous membrane tension. *Lower Extremities:* Left hip externally rotated, Left posterior fibular head.

Abdomen: stomach elevated and rotated internally toward the midline, liver internally rotated toward the midline.

Imaging:

Brain MRI Non-Contrast 10/18/2008 (see attached image)

1. Chiari I malformation
2. Small CSF signal intensity structure just below the skull base at the foramen magnum, displacing the medulla in an anterior leftward direction. This has not caused significant compression deformity or edema within the medulla. Remainder of exam normal.

Brain MRI Non-Contrast 06/16/2002

- Tonsillar ectopia/mild Arnold-Chiari malformation

MRI/MRA Bilateral Upper Extremities with and without Contrast 11/14/2008

- Marked extrinsic compression of both subclavian veins on hyperabduction, with mild distention of both cephalic veins, no distention of neck veins, and no venous thrombosis.
- Right anterior scalene muscle has mildly broadened insertion on the first rib
- Mild hypertrophy and mild to moderately broadened insertion of the left anterior scalene muscle.
- No stenosis of costoclavicular interval, subclavius-serratus space, or retropectoralis space bilaterally. No extrinsic compression or intrinsic stenosis of arterial tree.
- No cervical disc disease, central canal stenosis or neural foraminal stenosis.

Assessment

- 1) Chiari I Malformation
- 2) Chronic Migraines
- 3) Thoracic Outlet Syndrome
- 4) Hiatal Hernia with GERD
- 5) Hypothyroidism
- 6) Somatic Dysfunction of Cranium, Cervical, Thoracic, Lumbar, Pelvis, Sacrum, Ribs, Upper and Lower Extremities, and Abdomen

Treatment

Osteopathic Manipulative Treatment to 7-8 regions

The patient was managed pharmacologically by her primary care physician, so she is seeing an OMT specialist for Osteopathic treatment. At the initial visit, the aim was the resolution of tension through the core-link that was limiting motion throughout the cranium and pulling the cerebellum inferiorly. BLT was utilized in the lower and upper extremities, pelvis and diaphragm. Then, a partial release of the Occipito-Atlantal joint utilizing balanced membranous tension (BMT) and Lumbo-Sacral decompression as well as intraosseous strain at S2 segment. Venous Sinus Drainage Technique was applied to begin to address the sutural and dural tension. HVLA techniques were avoided. Due to the complexity of the case, treatment proceeded slowly.

Course of Treatment:

Patient followed-up four weeks later (3/12/09) and stated that for 3-4 days the headaches were 50-60%

better and was able to reduce the Norco. Numbness in her hands had also improved although not as much. At this time, work with the core link was continued, addressing lesions in the OA and AA joints using Balanced Ligamentous Tension techniques. The internally rotated right temporal bone, in contrast to the rest of the mechanism which favored inhalation, presented a difficult lesion in the cranium and improved motion was established using BMT at both the Sphenosquamous and Occipitomastoid articulations.

Due to work and travel constraints, the patient was not able to follow up for 6 weeks (4/30/09), at which time she presented with a severe flare-up of her thoracic outlet syndrome with pain and numbness and tingling into her right arm. She stated that she had only had 2 full migraines since last visit, although severe tension headaches were present daily. Structural exam revealed anterior and middle scalene spasm drawing the right upper ribs superiorly while the right clavicle was compressed at the sterno-clavicular articulation. Focus of treatment was BLT to the externally rotated left hip, counterstrain to address scalene tenderpoints as well as continued work with the core-link utilizing Balanced Membranous Tension techniques. She had complete relief of her upper extremity symptoms following OMT.

She returned three weeks later (5/25/09) and remarked that her vision had been remarkably free of visual effects since after the initial visit, and she had had only one episode of ear ringing. Both of these had been occurring 2-3x per week. Her daily multiple episodes of vomiting had diminished to one episode in five days, the most recent one triggered by bending over. Specific attention to the sagittal suture and parietal notch was begun, as well as Counterstrain and BLT to the cervical, thoracic, lumbar, and pelvic regions and all extremities.

Three weeks later, she returned (6/15/09) and stated that she had only had five days of migraines and no episodes of vomiting. On exam, she had significantly decreased core-link tension and with improved inherent motion of the right temporal bone. At this visit, the cerebellum responded to treatment and elevated, relieving congestion of CSF fluctuation and improving motility of the neural tube. The lower cervical somatic dysfunctions were resolved with gentle articulatory techniques as well as BLT.

She did not return for 2 months, as she experienced the onset of severe anxiety in the interim. Her primary doctor felt that she was bipolar and placed her on both Lexapro 10mg QD and Xanax 2-3mg in divided doses per day as needed for the anxiety. She reported that she also began to lose weight and incidentally, had had no migraines since the previous treatment.

Over the course of the next year, she has had very few migraines, usually related to some sort of physical strain such as a difficult car ride, or visit to an amusement park with her daughter (against my advice). However, she quickly responded to OMT and become pain-free, particularly at the point during treatment when she perceives “the back of her head relaxing” which coincides with the relaxation of the tentorium and elevation of the cerebellum. Because of her history of addiction and struggles with anxiety and thyroid regulation, she has not eliminated her medicines, although she currently is taking only 5mg of Lexapro, <2 mg of Xanax total per day, and 1-2 Norco tablets per day, along with 75mcg of Synthroid. The current plan is to taper off her Norco first.

Discussion

Chiari I malformation (CM-1) is also known as the adult Chiari malformation which indicates a clinically significant cerebellar tonsillar herniation with or without hydrocephalus. This is a separate clinical entity from type II “Arnold-Chiari malformation” in infants which is associated with spina bifida and herniation of parts of the brainstem as well as cerebellum. In his review of the literature in 2001, Bejjani concluded that the amount of herniation visible on MRI does not always correlate with level of symptomatology as there are cases with >5mm herniation who are asymptomatic and cases with less who are symptomatic.ⁱ From an Osteopathic perspective, diminished motion of the PRM may be proportional to symptoms.

Kaplan and Oksuz reviewed 73 patients with CM-1 and found 15% had migraines of which 2/3rds had chronic migraines. Therefore, approx, 10% of this CM-1 population suffered from chronic migraines compared with 2 to 3% of the general population, according to a study cited in this article. Chronic migraines are defined as migraines occurring on at least 15 days per month, for more than three months, in the absence of medication overuse. The group with CM-1 and chronic migraines were found to have significantly earlier age of onset of pain, more headache days per month and significantly higher baseline pain intensity, exacerbation of pain intensity, nausea, vomiting, and pain aggravated by physical activity than a control group with chronic migraines in the absence of CM-1.ⁱⁱ This patient suffered from severe chronic migraines consistent with the CM-1 population.

Aydin, et al pursued a morphometric analysis of patients with CM-1 and found an overall decrease in mean measurements including lengths of the supraocciput, the clivus and the posterior cranial fossa as well as height of the posterior cranial fossa (foramen magnum to splenium of corpus collosum) compared with a healthy control group. The one measurement which was larger in the CM-1 group was the A-P diameter of foramen magnum. The larger diameter of foramen magnum could be seen as a compensation for the cerebellar herniation. Unfortunately, this study did not involve lateral measurements.ⁱⁱⁱ From an Osteopathic perspective, this indicates that CM-1 may be more common in Flexion-type craniums, or perhaps cases with inferior vertical strains, both of which would have shorter A-P dimensions. This would be consistent with flattening of the tentorium cerebelli during the inhalation phase of the PRM.^{iv}

Standard treatments for chronic migraines involve the reduction of pain medicine to avoid the transition to medication overuse headaches, and the use of topiramate (Topamax) and botulinum toxin.^v This patient failed botulinum therapy as well as other anti-seizure medications, although she had not been on topiramate. The standard treatment for symptomatic, refractory Chiari I malformation is surgical decompression.^{vi} In this case, the patient was seeking non-pharmacological and non-surgical options.

The resolution of chronic migraines in the setting of Chiari 1 malformation in this case was dependent on treatment of the Reciprocal Tension Membrane (RTM). Normal function is based on freedom of motion of the bony attachments throughout this mechanism. Particular attention was paid to the relationship of the cervical attachments of the spinal dura to the attachments around foramen magnum. Normalization of the motion of the tentorium cerebelli allows for greater freedom of the cerebellum which relaxes pressure on the tonsils and nearby cranial nerves, including the vagus. This led to a decrease or elimination of symptomatology in this patient.

In patients with migraines, Sutherland drew attention to the “sidetip” of the sphenoid towards the side with pain. This would have significant effect on the pituitary as well as the dura attached to the sphenoid.^{vii} Magoun emphasized the importance of not only the sphenoid but the temporal and its sutures with the sphenoid and occiput as well as the dura and the vasculature that travels with it including the venous sinuses and the middle meningeal artery in migraines. He also highlighted dural pull on the vagus at the jugular foramen causing nausea, vomiting and other digestive complaints.^{viii} With Chiari I malformation, cerebellar tonsil herniation could increase pressure on the vagus as well. The role of the temporal bones is fundamental because of the attachments of the tentorium cerebelli and its close relationships to neighboring bones. The tent crosses the parietal bone at the parietal notch between the mastoid and squamous portions of the temporal bone. The fixed border of the tent runs from the petrous ridge to the posterior clinoid process on the sphenoid body. The lateral angle of the occiput is linked to the mastoid portion of the temporal at the most superior section of the occipito-mastoid suture.^{ix} Thus, freedom of motion of the tentorium cerebelli depends on free motion of the temporal bones and its neighbors, which will then allow the depressed cerebellum rise.

The etiology of Chiari I Malformation is not well understood. Aydin, et al, came to the conclusion that:
...underdevelopment of occipital somite originating from the paraxial mesoderm during the intrauterine life is the main cause of CMs. Downward herniation of normal-sized hindbrain is secondarily induced by overcrowding in the PCF. Displacement of CSF probably contributes to symptoms, and formation of the syringomyelia is directly a result of the obstruction at the foramen magnum.^x

In this case, it seems that the nuchal cord may be implicated, however the the role of intrauterine trauma in the development of CM-1 is not well explored in the medical literature. The effects on CSF fluctuation are consistent with dysfunction of the PRM.

This case raises many other topics of discussion beyond the scope of this paper. The function of the three diaphragms, as articulated by Viola Frymann^{xi}, reveals a relationship between the dysfunction of the thoracic diaphragm in GERD/Hiatal hernia and the dysfunction of the tentorium cerebelli in Chiari I malformation, both disease process were present in this patient. The relationship of pituitary function to thyroid function via biomechanics of the sphenoid and the core-link as well as decreased motion of the cervico-thoracic junction due to thoracic outlet syndrome could be explored in relation to Chiari I malformation. Finally, the history of substance abuse, the current medications and the possibility of medication overuse headaches are alternate possible explanations for her chronic migraines other than the obvious Chiari I malformation. However, regardless of ultimate cause, osteopathic treatment gave her great relief with the resolution of somatic dysfunction, making medication overuse a less important culprit. Given the choice of surgery, it is clear that a therapeutic trial of OMT presents a much safer and potentially more effective approach in the treatment of chronic migraines in the setting of Chiari I malformations.



Figure 1: Non-contrast Brain MRI, Mid-Sagittal View, dated 10/18/2008.

Endnotes

- i Bejjani GK: Definition of the adult Chiari malformation: a brief historical overview. *Neurosurgery Focus*. July 2001 11:1-8.
- ii Kaplan Y, Oksuz E: Chronic migraine associated with the Chiari I malformation. *Clinical Neurology and Neurosurgery*. 2008 110:818-822.
- iii Aydin S, Hanimoglu H, et al: Chiari type I malformations in adults: a morphometric analysis of the posterior cranial fossa. *Surgical Neurology*. 2005 64:237-241.
- iv Magoun HI: *Osteopathy in the Cranial Field*. Journal Printing Company. Kirksville, MO. 1976. p. 36
- v Diener HC, Holle D, Dodick D: "Treatment of Chronic Migraine." *Current Pain and Headache Reports*. November 16, 2010 [Epub ahead of print].
- vi Bejjani, 1-8
- vii Sutherland WG: *Contributions of Thought*. 2nd Ed. SCTF, Inc. Fort Worth, TX. 1998. p. 100
- viii Magoun, pp.282-283
- ix Ibid. p. 28
- x Aydin. P. 240
- xi Frymann, VM: The core-link and the three diaphragms. A unit for respiratory function. *AAO Yearbook*, 68:13, 1968. Reprinted in *The Collected Papers of Viola M. Frymann, DO*. American Academy of Osteopathy. Indianapolis. 1998