Title and Author(s)

Title: Chiari 1 Malformation

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Introduction or Patient History

A 51 year old female patient presented with symptoms of fatigue, headache, pain, and weakness. The patient first had a CT Scan of the cervical spine done at the same facility where the MRI was done at the University of Pennsylvania Hospital (HUP). The result of the CT Scan of the neck was negative. An MRI was ordered of the Head and Cervical spine to r/o Chiari Malformation. Also, no previous MRI was done on this patient.

Patient Preparation and Scan Set up

The MRI was performed in an outpatient facility at HUP on a Siemens Symphony Maestro 1.5 Tesla MRI. The patient was screened by using a questionnaire that asked specific questions to the history of the patient in relation with MRI. It was done before entering the room complying with MRI safety guidelines. A multi-channel head and phased array spine coil was used for the procedure. The patient was given earplugs as required by the safety guidelines for hearing protection due to noise created by the scanner. The patient was positioned supine (*flat on her back*) with her head going first into the scanner. Furthermore, to make the patient more comfortable a wedge that goes under the patient knees to give support to the lower back of patient as well as a blanket was provided. Also, the patient was given a call bell to squeeze in case the patient doesn't feel comfortable inside the scanner.

MR Imaging Parameters

MRI sequences were: Sagittal T1- weighted, Axial Flair, Axial fast spin echo T2-weighted. The flow dynamics were performed using a cardiac gating and 2D phase contrast cine with sequences in the sagittal plane with VENC values of 30, 20, 10 cm/sec. The cervical spine sequences were; Sagittal T1, Sagittal T2 and Axial fast spin echo T2-weighted images.

* Sequences of the select images at the end of article.

Siemens Symphony Maestro 1.5 Tesla MRI Syngo 2004A 4VA25A

Series	Sequen-ces	Types	TR	TE	FOV	S/T	Matrix	NX	Band width
Series 1	Multi-Plane Loc.		63.0 ms	6.7 ms	22 cm	20 mm	256/ 256	1	590
Series 2	Fl2d Multipla-ne *	Cine	63.0 ms	6.7 ms	22 cm	20 mm	384/ 269	1	110
Series 3	Sag T1	SE	551.0 ms	16.0 ms	22 cm	5.8 mm	384/ 269	1	110
Series 4	Ax. T2 *	TSE	3730.0 ms	103.0 ms	22 cm	5 mm	256/ 156	1	210
Series 5	Ax Flair	Flair	9870.0 ms	153.0 ms	22 cm	5 mm	256/ 153	1	210
Series 6	Loc.		24.0 ms	6.0 ms	20 cm	9 mm	256/ 128	1	130
Series 7	Sag. T1	Flair	1620.0 ms	13.0 ms	16 cm	3.5 mm	384/ 192	1	130
Series 8	Sag. T2 *	FSE	4000.0 ms	116.0 ms	16 cm	3.5 mm	384/ 192	2	190
Series 9	Ax. T2	2D	4300.0 ms	129.0 ms	16 cm	3.7 mm	256/ 192	2	150

Findings and Discussions

Chiari malformation is considered a congenital condition although acquired forms of the condition have been diagnosed. There are several types of severity the higher the type number the more life-threatening. For example:

Type I – it may or not cause any symptoms at all to the patient and usually is found by doing another procedure for something else to the patient. Here are some of the symptoms or signs:

- Headaches, by coughing, sneezing or straining
- Neck pain
- Vomiting
- Swallowing
- Slurred speech

Type II – the symptoms with a degree of herniation of the cerebellum pushing down the foramen magnum into the spinal canal.

- Respiratory problems
- Loss of upper and lower strength
- Downward eye movements
- Gag reflex

Type III – it is more serious because is life-threatening complications.

- Mental impairment
- Paralysis of the arms and legs
- Myelomeningocele
- Syringomyelia
- Spina bifida

Type IV – Especially infants born with it usually don't survive infancy.

- Cerebellum fails to develop normally

According to the report, "This patient was found to have Chiari I Malformation with some swelling of the cerebellum tonsils. Also there was no intercranial hemorrhage, extracerebral fluid collection, midline shaft or mass effect. Furthermore, there was no evidence of hydrocephalus or syrinx." (*Radiologist findings)* The swelling didn't stop CSF flow to the spinal canal anteriorly or posteriorly. The patient will be periodically checked to see if any more changes occur. Moreover, the patient at this point will get treatment with medications. Since Chiari I Malformation at this stage doesn't require surgery of the posterior cranial area. Chiari Malformation is a congenital condition of the cerebellum, it is part of the brain that controls balance. Again the symptoms are headache, coughing, and straining. The symptoms can develop at any age. But usually occur in childhood or after the age of 30. Neurosurgeons take a more conservative approach before doing surgery because they try to use other methods before having to do surgery. The surgery involves decompression and enlargement of the foramen magnum in the base of the skull. Other methods maybe the use of a shunt or by creating a path outside the spinal cord into some other body cavity where the fluid could be drained. These types of procedures report a 70% success rate in dealing with the symptoms. In relation to modalities that are used in diagnosing this disease a CT Scan is very helpful with Chiari II Malformation in diagnosis of hydrocephalus and shunt malfunctioning. MRI is very helpful due to the different views anatomically and excellent images contrasts acquired.

Conclusions

It appears that people who have Arnold Chiari Malformation don't realize they have this disease. Generally it is diagnosed as an incidental finding when they are having some other procedure. The Chiari Malformation statistically affects about 1 in 1,000 people in this country. Adults have different choices when dealing with this disease. One of the methods in dealing with this disease is surgery of the base of the skull. By making an enlargement of the foramen magnum to release pressure from the tonsils of the cerebellum that are pushing down on them. Of course, medications also play a role with this disease by helping patients to deal with the discomfort. Newborns have less chance surviving childhood because the cerebellum is not fully developed. There are combinations of modalities that contribute in the fight against this disease. Someday, new research involving stem cells and with new technologies, we should be closer to discover a cure for this congenital disorder.

References

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   Office of Communication and Public Liaison
   National Institute of Health
   Bethesda, MD 20892
   http://ninds.nih.gov/disorders/chiari.htm

2. Chiari Malformation
   http://www.Neurosurgerytoday.org

3. Arnold-Chiari Malformation
   National Organization for Rare Disorders at P. O. Box 1968, Danbury, CT 06813-1968; Phone (203) 744-0100; www. Rarediseases.org, WebMD.com 4/19/2005

4. Chiari Malformation
   Cleveland Clinic Foundation
   http://cms.clevelandclini.org/neuroscience/

Images

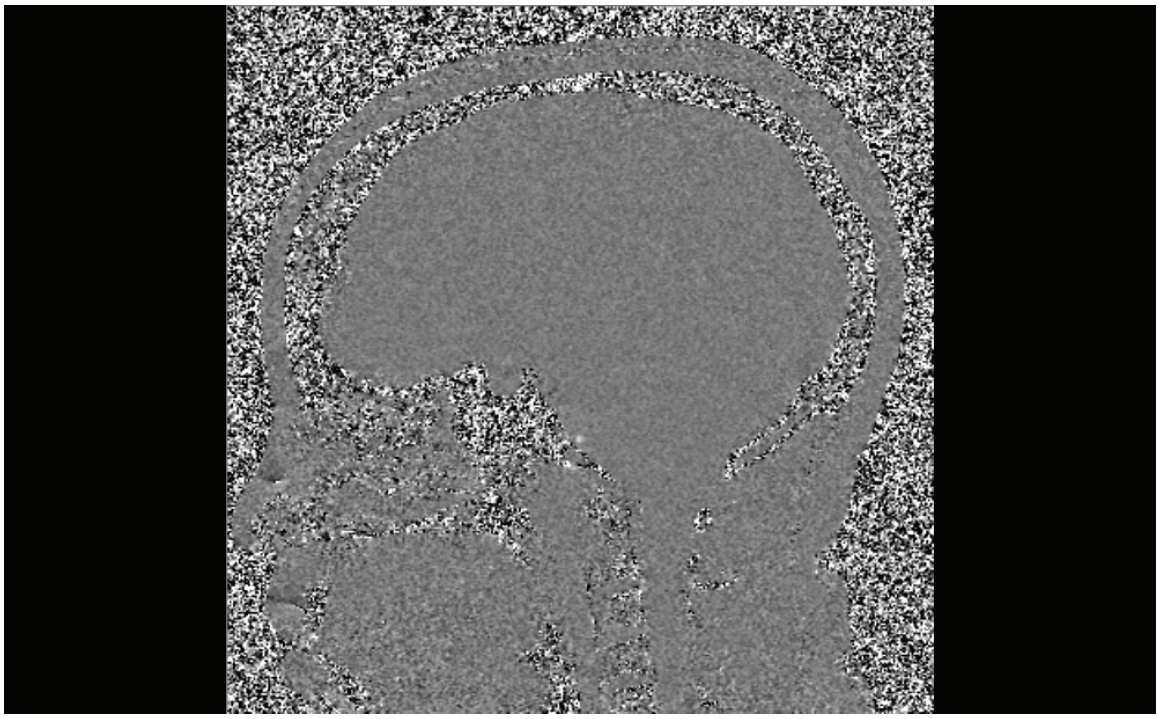

Image 1
*Fl2dl Sag. Brain Cine

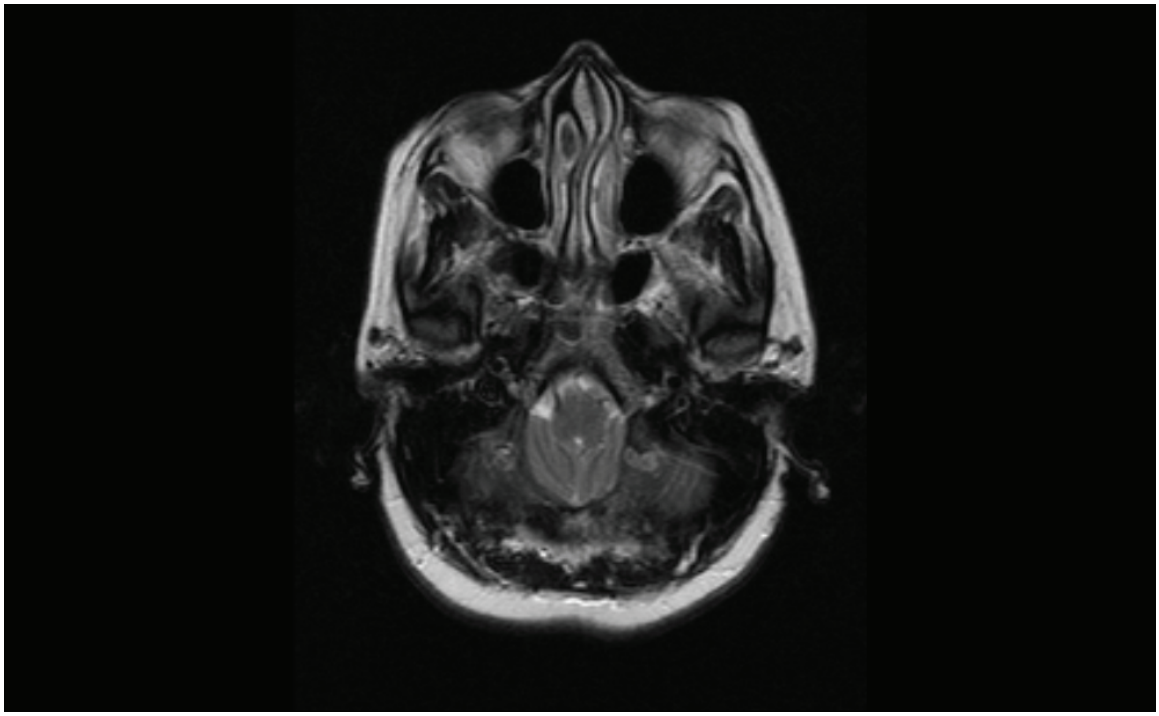

Image 2
AxialT2

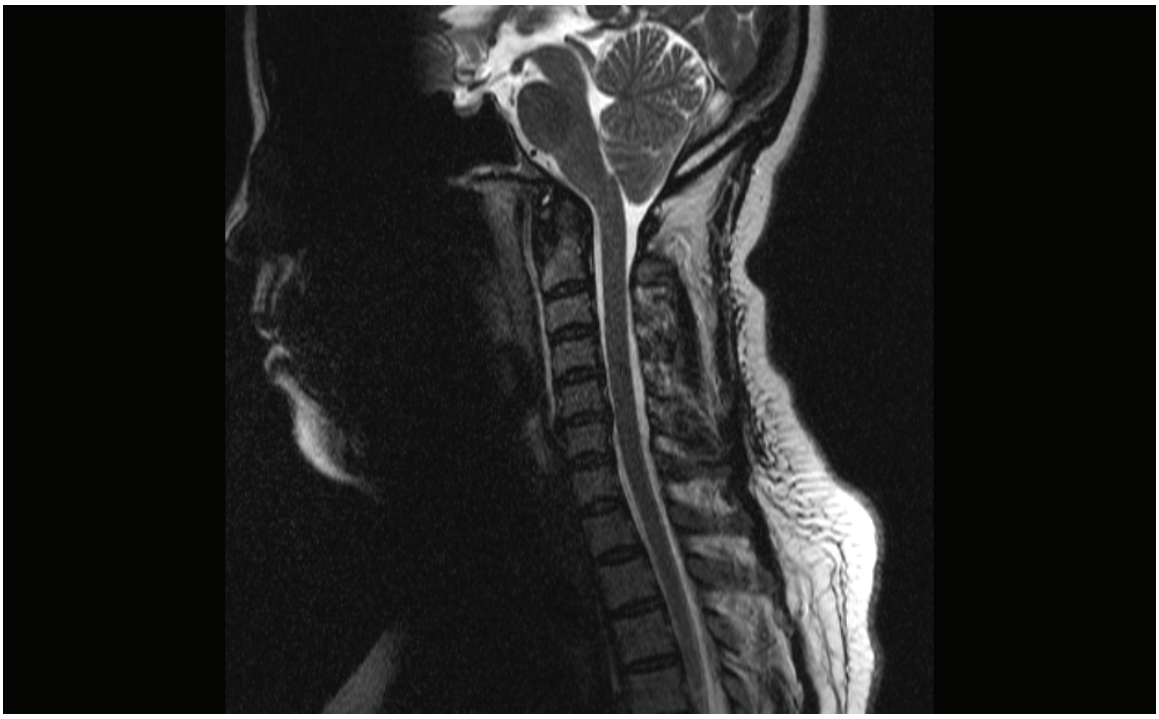

Image 3
Sag. T2