SMRT Student Scope Submission

Title and Author
Title: X-linked adrenoleukodystrophy

Author: Lisa Hellerich

E-mail/Contact Information: lhelleri@unmc.edu

Supervisor/Education Coordinator: Adam Stevens

E-mail/Contact Information: ajstevens@unmc.edu

Affiliation: UNMC

Date of Submission: April 17, 2008

Introduction or Patient History

An 8 year old male was scheduled to have an MRI brain with and without the use of a contrast agent. No previous imaging noted. The patient was being seen primarily for behavioral regression including visual and gait disturbances. He was also having problems with incontinence.

Patient Preparation and Scan Set Up

The exam was performed on a 1.5 Tesla (T) Philips MR Scanner. The patient was screened to ensure that no ferrous object was on or in him. After completion of the screening process, the patient was changed into a hospital gown and taken to our radiology nurses to have his IV started. The patient was then taken into the scan room and positioned supine on the table head first into the magnet. The patient then was given head phones to protect his hearing and to allow the technologist to communicate to the patient during the scan. He was also given a cushion under his knees to aide in his comfort and a squeeze ball in case he needed anything throughout the scan. The head coil was used and placed over the patient's head and then centered inside the magnet.

MR Imaging Parameters

Seven sequences were performed for the routine brain, including post contrast imaging. 5 cubic centimeters (cc) of MultiHance (gadobenate dimeglumine) were administered for the post contrast images.

Sequence	FOV	Slices	TR	TE	Spacing	Matrix
Survey	250	3-plane	15	5.2	10 skip 10	256x256
T1 Sag. SE	240	21	566	15	5 skip 1	256x256
FLAIR Ax.	240	26	11,000	140	5 skip 1	256x256
DWI Ax.	240	24	3,156	75	5 skip 1	112x256
T2 Ax.	240	26	6,142	100	5 skip 1	384x512
CONTRAST PATIENT						
T1 Ax.	240	26	704	15	5 skip 1	256x256
T1 Cor.	240	26	704	15	5 skip 1	256x256
T1 Sag. SE	240	21	566	15	5 skip 1	256x256

Findings and Discussion

The patient was diagnosed with x-linked adrenoleukodystrophy (ALD). This is one of a group of genetic disorders called leukodystrophies that cause damage to the myelin sheath. The loss of myelin and the progressive dysfunction of the adrenal gland are the primary characteristics of ALD. X-linked ALD is the most common form, which involves an abnormal gene located on the x-chromosome. Women have two x-chromosomes and are the carriers of the disease, but since men only have one x-chromosome and lack the protective effect of the extra x-chromosome, they are more severely affected. Childhood form of ALD is the most severe. The onset occurs in ages 4-10 years. Common symptoms are usually behavioral changes such as abnormal withdrawal or aggression, poor memory, and poor school performance.

Prognosis for patients with ALD is generally poor due to progressive neurological deterioration. Death usually occurs within 1-10 years after the onset of symptoms. Treatment with adrenal hormones can be lifesaving. Symptomatic and supportive treatments for ALD include physical therapy, psychological support, and special education. Evidence suggests that a mixture of oleic acid and erucic acid administered to boys with x-ALD can reduce or delay the appearance of symptoms. Bone marrow transplants can provide long-term benefit to boys who have early evidence of x-ALD, but the procedure carries risk of mortality.

Conclusion

This case was interesting to me; therefore, I researched the genetic disorder ALD. When scanning a brain that has a tumor or other abnormality it is necessary to scan in all three planes post contrast. This gives the radiologist more information to use for proper diagnosis. ALD is a rare inherited disorder that leads to progressive brain damage, failure of the adrenal gland and eventually death. Onset of childhood ALD occurs between the ages of 4 and 10. Up to the point of onset, development is normal. The most common initial symptoms are difficulty in school, behavioral disturbance, impaired vision, or impaired hearing. After initial symptoms appear, the health of the patients deteriorates rapidly. The average time between the initial symptoms and the vegetative state or death is approximately 2 years.

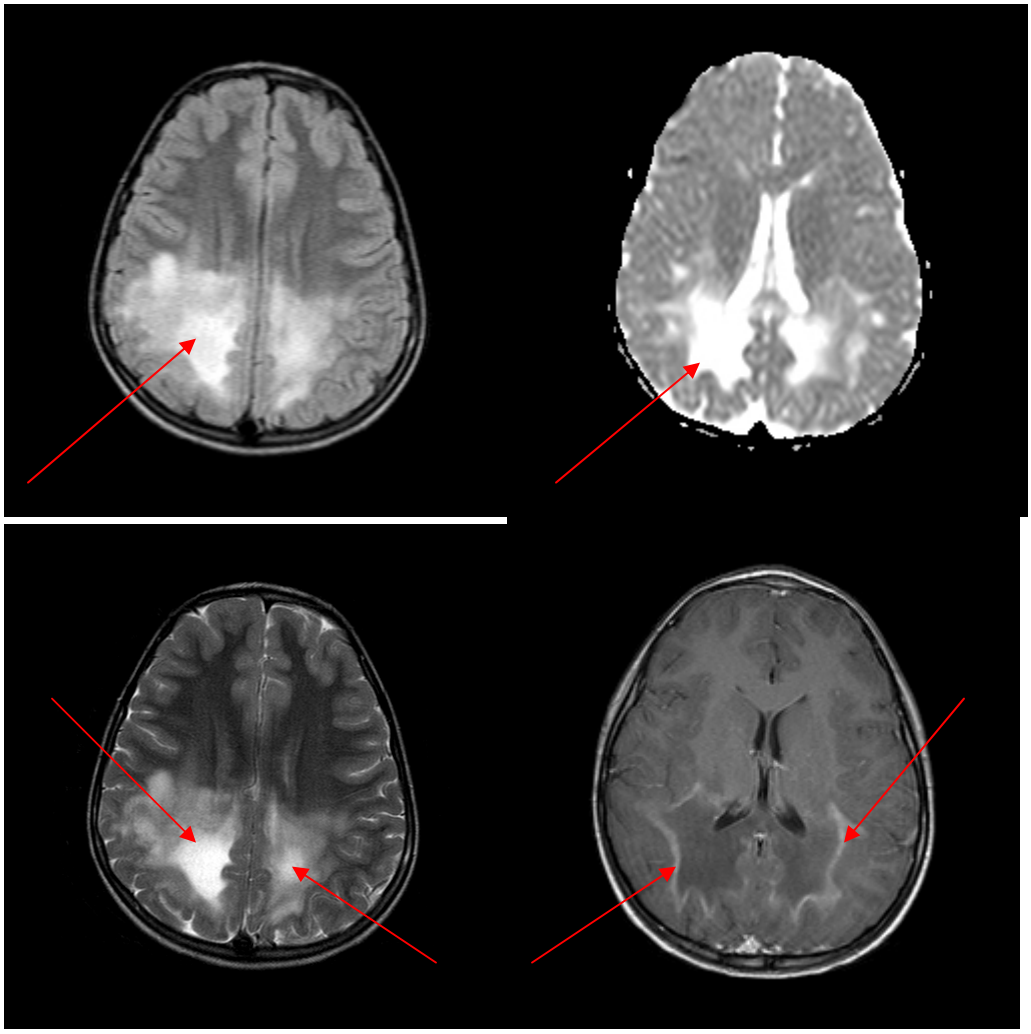
References

Adrenoleukodystrophy (ALD): Wikipedia. Available at: http://en.wikipedia.org/wiki/Lorenzo_Odone#Famous_patients. Accessed March 2008.

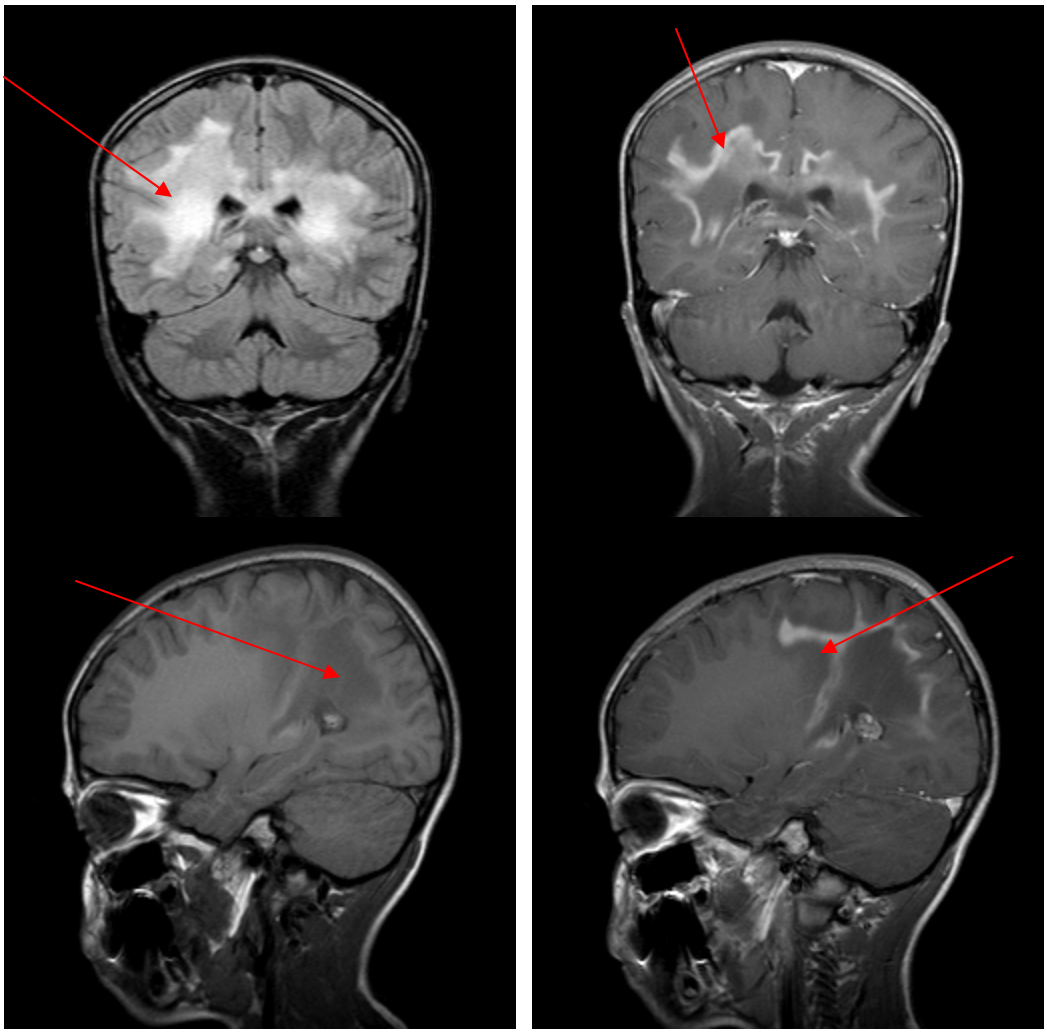
NINDS Adrenoleukodystrophy Information Page. National Institute of Neurological Disorders and Stroke. Available at: http://www.ninds.nih.gov/disorders/adrenoleukodystrophy/adrenoleukodystrophy.htm. Accessed March 2008.

Fact Sheet: X-Linked Adrenoleukodystrophy (X-ALD). United Leukodystrophy Foundation. Available at: http://www.ulf.org/types/XALD.html. Accessed March 2008.

Images



Images: Top left image is FLAIR AXIAL
Top right image is Diffusion weighted image (DWI) AXIAL
Bottom left image is T2-weighted AXIAL image
Bottom right image is T1-weighted AXIAL (post contrast)



Images: Top left image is a FLAIR CORONAL
Top right image is a T1-weighted CORONAL (post contrast)
Bottom left image is a T1-weighted SAGITTAL
Bottom right image is a T1-weighted SAGITTAL (post contrast)