Misdiagnosis in a 12 Year-Old Female with Right Aberrant Subclavian Artery and Difficult Swallowing

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INTRODUCTION

A left aortic arch with aberrant right subclavian artery is the most common congenital aortic arch anomaly. Aberrant right and left subclavian arteries have been imaged on MRI/MRA/MRV of the brachial plexus in patients presenting with symptoms of TOS: arm and hand pain with paresthesias; hand muscle weakness, headache; autonomic vascular and temperature changes; whooshing sounds in the ear, ear pressure pain; jaw pain, lower extremity pain, and visual "floaters". A 12-year old girl presented with complaints

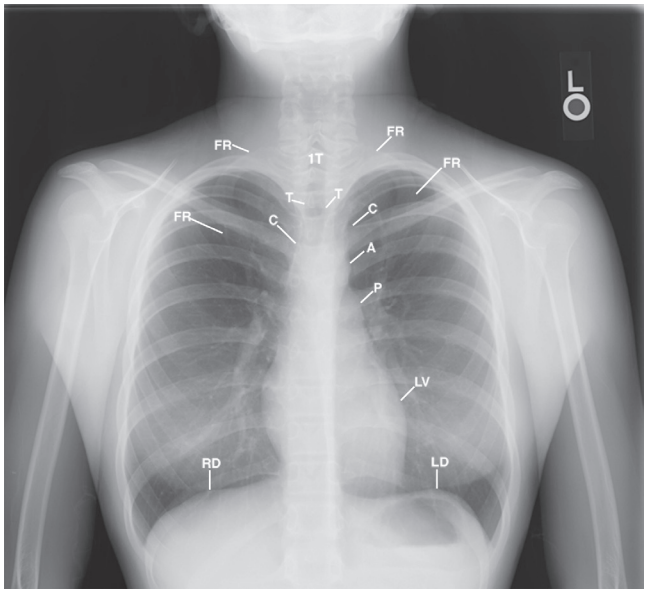

FIGURE 1. Posterior Anterior Chest Radiograph

The Posterior Anterior chest radiograph displays thin subcutaneous tissues; forward rounding of the shoulders, right elevated shoulder as compared to the left shoulder; heads of the clavicles (C) low over the posterior 5th ribs, right lower than left.

A-Aorta, C-Clavicle, FR-First rib, LV-Left ventricle, P-Pulmonary artery, -Left and Right Hemidiaphragm (LD, RD) and T-Trachea, 1T-First thoracic vertebra.

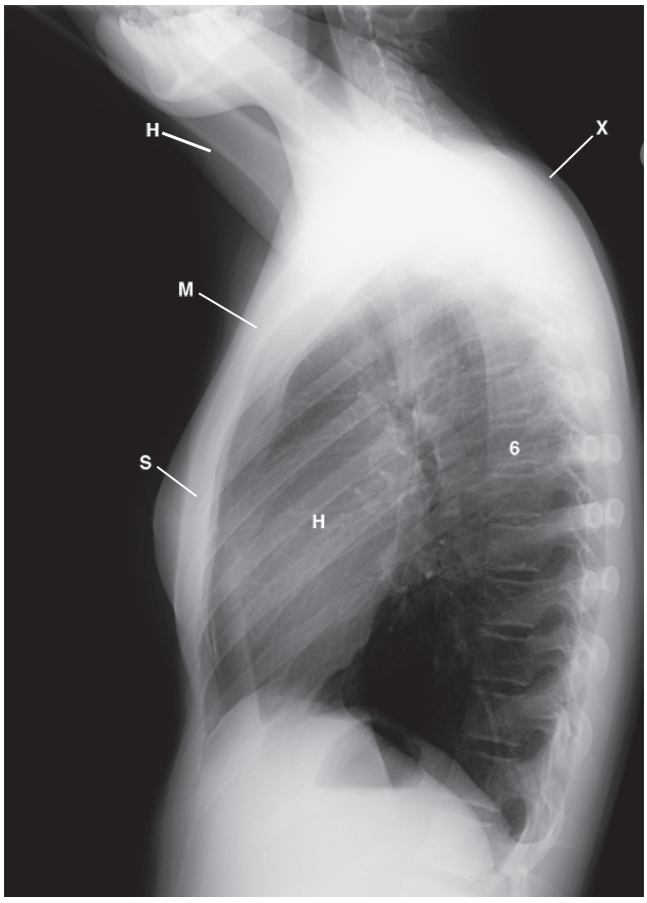

FIGURE 2. Lateral Chest Radiograph

The lateral chest radiograph displays the increased slopes of the first ribs (not labeled) backwardly displacing the manubrium (M) and sternum (S), mild kyphosis of the cervicothoracic spine accentuated by the anterior bowed body of the sternum, and rounding of the shoulders (X)

H-Heart, M-Manubrium, T-Trachea, X-Rounding of the shoulders, 6-Sixth thoracic vertebra

of problems swallowing solid foods since age seven years. She recently developed pain in the right chest wall with inspiration, in addition to common thoracic outlet syndrome (TOS) complaints of headache, dizziness, hand pain, tingling and numbness, muscle pains in the neck and shoulders and back, along with jaw pain and whooshing sounds in the ears.[1,2]

CLINICAL HISTORY

The clinical history provided by her mother indicated that she began having trouble swallowing prior to the scheduled bilateral MRI/MRA/MRV of the brachial plexus. At age 4 years, she was diagnosed with asthma and had to take medication. At age 5 years, she was diagnosed with juvenile arthritis because she presented with right wrist pain that progressed to involve the joints of the right fingers. She was initially evaluated by her pediatrician who referred her to an orthopedist who obtained x-rays of her right and left wrists. A right dorsal lunate subluxation was found. She was then referred to an adult rheumatologist who evaluated her laboratories for systemic inflammation. Labs were all negative. CBC revealed a white count of 6.7, hemoglobin of 14, hematocrit of 40.2, and a platelet value of 279. Her differential was N 47, L 41, M 6–5, E 5.1 and B 0.3. The rheumatologist ordered a wrist guard and had her wear it during school as well as during physical education. Her mother observed she complained much less with the help of the wrist guard. She complained of difficulty when combing her hair, difficulty holding a pencil or pen in school. The pain never awakened her at night.

Past Medical History: Significant for Asthma, Atopic dermatitis, Allergic rhinitis. Immunizations were up to date. PPD was negative and Chicken pox status: vaccinated.

Social History: She had no siblings, pets, and did not report travel.

Allergies: Amoxicillin gave her a rash and pollens.

Review Of Systems: Positive for Asthma; atopic dermatitis: 3–4 episodes of diffuse chest pain unrelated to exercise; "dizzy" sensations in her legs; positive joint pain in the bilateral wrists and right finger.

Physical Examination: Vitals: Temperature 36.7, blood pressure 99/45 mm Hg, heart rate 60 beats/min, respiration

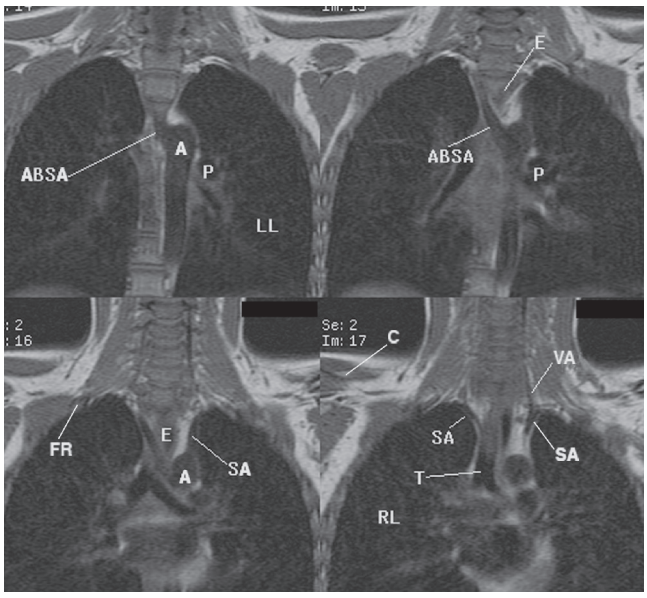

Figure 3. This is a coronal 4/4 T1-Weighted MRI

Image that displays the origin of the dilated aberrant right subclavian artery (AB.S.A) image 14, ascending from the aorta (A) posterior to the esophagus (E) image 15, from the descending aorta, images 14–17.

A-Aorta, C-Clavicle, FR-first rib, LL-left and RL-right lung, P-pulmonary artery, SA-subclavian artery, T-trachea, VA-left vertebral artery

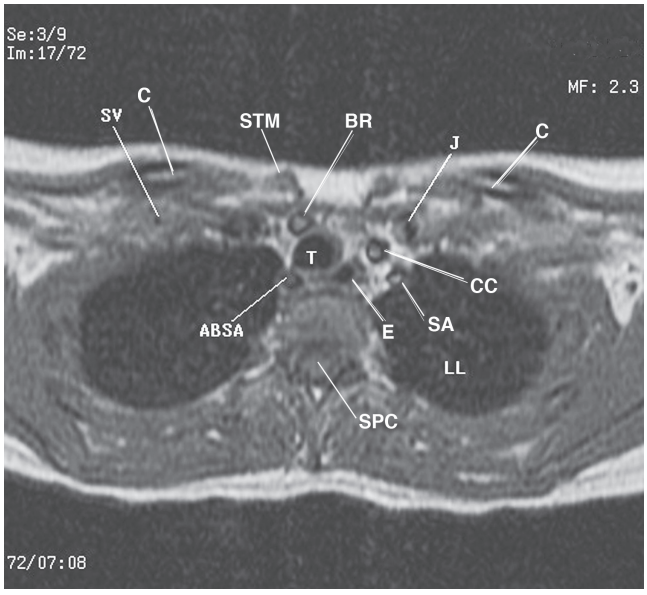

FIGURE 4

This a transverse T1 weighted image that displays the bulbous expanded right subclavian vein (SV) over the right first rib (not labeled) as compared to the left subclavian vein on the left first rib (not labeled), left lower than right reflecting impedance to venous return. Observe the aberrant right subclavian artery (AB.S.A) posterior right lateral to the trachea (T) on the vertebral body (not labeled). BR-high proton dense brachiocephalic artery, C-clavicle, E-esophagus, J-left internal Jugular vein, CC-left common carotid artery, LL-left lung, SA-left Subclavian artery, SA-Subclavian artery, SPC-spinal canal with the spinal nerve, STM-sternocleidomastoid muscle, T-Trachea.

16 Mm., height 128 cm and weight 25.9 kg. She was positive for bowel sounds, minimal pain with movement of the right wrist and pain on flexion and extension; minimal pain with movement of the left elbow and wrist.

Impression: Right lunate dislocation. Differential diagnosis to include juvenile idiopathic arthritis. Would recommend MRI and conservative management.

At 7 years of age, she complained of pain over her right chest wall. She needed more saliva when she had to drink fluids.

At age 8 years, she had trouble swallowing pills. She could not swallow nor cough up pills, a significant finding as her mother recalls. She could actually hear her having difficulty in swallowing liquids.[3]

At 11 years of age, she began having headaches. Topamax (25 mg 1 time a day) was prescribed for her headaches. Four months later, Topamax was increased to 25 mg twice a day due to her increasing painful headaches. She had been doing well for several weeks. Headaches increased in frequency and severity, although she continued taking Topamax. She developed dizziness and lightheaded sensations when getting up out of bed and or lying down, pain in her neck, shoulders, back, feet, and an increased frequency of headaches along with jaw pain and whooshing sounds in the ears.[4] She had never experienced these symptoms before.

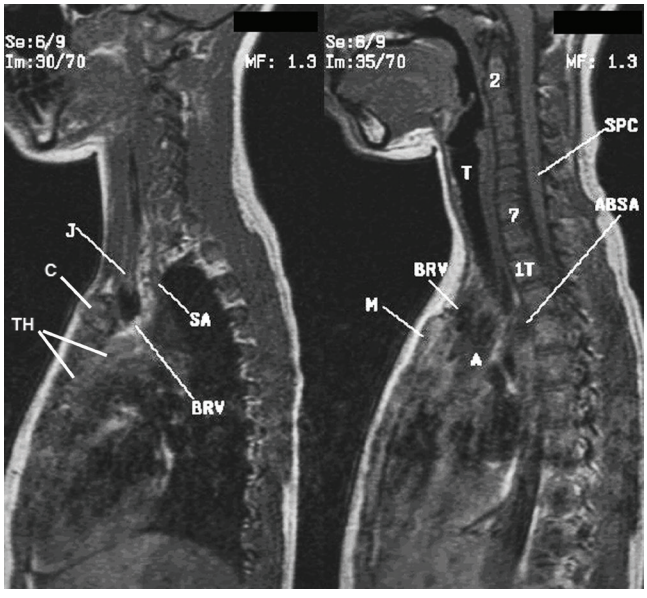

FIGURE 5

This a T1 weighted image sagittal sequence, image 30/70, that displays the speckled gray proton dense thymus gland (TH) compressed against the ascending aorta (not Labeled), gray proton dense left brachiocephalic (BRV) compressed by the anterior rotated clavicle (C) compressing the fascial plane of the ascending first division of the left subclavian artery (SA). Image 35/70 displays the aberrant right subclavian artery (AB.S.A), manubrium (M) compressing the left brachiocephalic vein (BRV) against the aorta (A). J-internal jugular vein, SPC, Spinal cord, T, trachea, 2–7, cervical vertebrae, 1T-first thoracic vertebra.

Her mother discussed her daughter's problem with a co-worker that had been previously evaluated at UCLA for thoracic outlet syndrome. The co-worker suggested that she should see Dr. James D. Collins in radiology. The mother was desperate, so she self-referred her daughter and paid cash for bilateral MRI/MRA/MRV of the brachial plexus.

Because of suspected thoracic outlet syndrome, bilateral MRI/MRA/MRV of the brachial plexus was requested to detect sites of brachial plexus compression.

Posterior anterior, lateral chest and anterior posterior cervicothoracic spine radiographs were obtained prior to the bilateral MRI/MRA/MRV of the brachial plexus.

Physical examination prior to her scheduled MRI revealed a thin narrowed chested female leaning forward. Vital signs were as follows: blood pressure in the right arm 100/70 mm Hg; pulse, 70 beats/min, temperature, 36.3 C; weight 36.3 kg; carotid pulses were palpable bilaterally; right carotid pulse had an early systolic bruit. Bilateral arms overhead in the sitting position dropped her blood pressure in the right arm to 60/40 triggering complaints of pain over the right neck, shoulder with tingling and numb sensations down into the 4–5th digits of the right hand (ulnar distribution). She became lightheaded and dizzy.

METHODS AND MATERIALS

Plain chest radiographs (posterior-anterior and lateral) were obtained and reviewed prior to the MRI. The procedure was discussed and the patient examined. Respiratory gating was applied throughout the procedure to minimize motion artifact. The patient was supine in the body coil, arms down to the side, and imaging was monitored at the MRI station. Magnetic resonance images were obtained on the 1.5 Tesla GE Signa MR scanner (GE Medical Systems, Milwaukee, Wisconsin). A body coil was used and intravenous contrast agents were not administered. A water bag was placed on the right and the left side of the neck to increase the signal to noise ratio for high-resolution imaging. A full field of view (44 cm) of the neck and the thorax was used to image the supraclavicular fossae. Contiguous (4 mm) coronal, transverse (axial), oblique transverse, sagittal, AER (of the upper extremities) T1-weighted images, and 2-dimensional time-of-flight MRA were obtained. If there was clinical evidence of scarring, tumor and/or lymphatic obstruction, fast-spin echo T2-weighted images would have been selectively obtained. The parameters for acquiring each sequence have been published.[5]

MRI/MRA/MRV FINDINGS

The PA chest radiograph displayed the elevated forward rotated right shoulder as compared to the smaller left; anterior

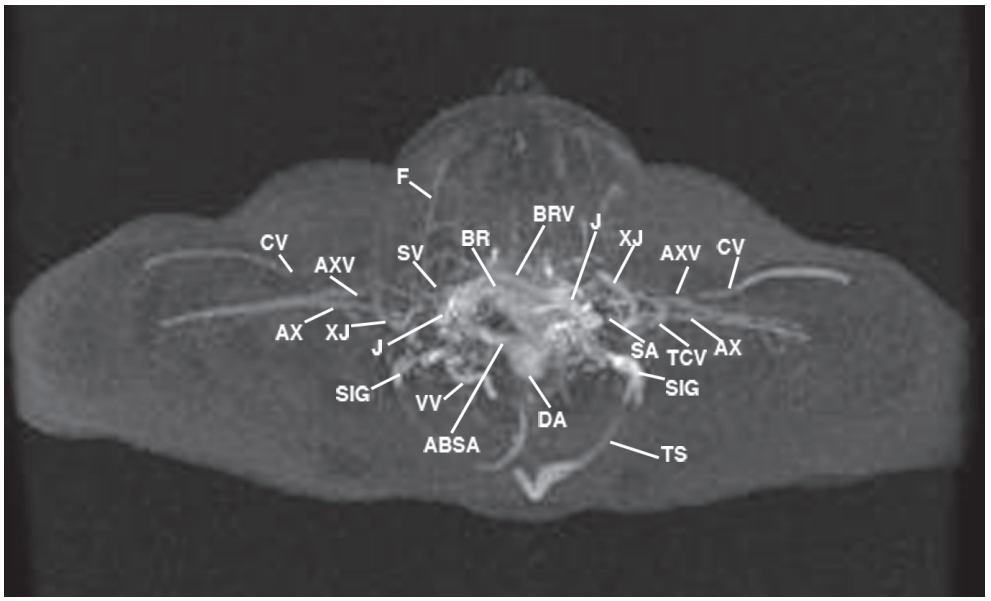

Figure 6. 2D Time of Flight (TOF) MRA / MRV Stacked Image

The stacked image displays the dominant forward right shoulder; longitudinal sinus (not labeled) draining into the dominant left transverse (TS) into the sigmoid sinus (SIG) compared to the decreased signal of the right transverse sinus (not labeled); backward displaced mildly compressed left subclavian artery (SA) and proximal dilatation of the axillary vein (AXV) reflecting greater impedance to venous return on the left; small left cephalic vein (CV) providing collateral circulation to the left axillary vein; near parallel rotation of the right axillary artery (AX) and vein (AXV) reflecting the forward rotation of the right shoulder as compared to the left; dominant left internal jugular vein (J) as compared to the right; aberrant right subclavian artery (AB.S.A) exiting the descending aorta (DA) and the transverse cervical vein (TCV) providing collateral venous return to the left subclavian vein.

BR- Brachiocephalic artery, BRV- Brachiocephalic vein, F-Facial vain, SV- Subclavian vein, VV-Vertebral vein, XJ-external jugular vein.

rotated heads of the clavicles over the posterior 5th intercostal spaces, right lower than left; arms at her side accentuating the thin subcutaneous tissues and narrow thorax, and normal cardiomediastinal structures and lungs.

The lateral chest radiograph displayed the anterior bowed body of the sternum; increased slope of the ribs; mild kyphosis of the thoracic spine; backward displaced the manubrium accentuating rounding of the shoulders, and normal cardiomediastinal structures and lungs.

AP cervicothoracic spine radiograph 15 degrees angle to the chin, arms at the side, displayed head and neck leaning right accentuating the low right first rib over the posterior low second rib as compared to the left.

Mutiplanar coronal MRI sequence displayed thin subcutaneous tissues; head and neck leaning left away from the forward elevated right shoulder, and drooping right shoulder as compared to the left; proximal dilated aberrant right subclavian artery taking origin from the posterior arch of the aorta ascending posterior to the esophagus compressed as the gray proton dense second division of the right subclavian artery with binding nerve roots within the scalene triangle (Figure 3); transverse MRI sequence cross referencing the coronal sequence to display the aberrant right subclavian artery right lateral to the esophagus and posterior lateral to the trachea on the thoracic vertebra ascending into the scalene triangle with bilateral costoclavicular compression of the bulbous expanded subclavian veins on the first ribs reflecting impedance to venous return (Figure 4); high proton dense right axillary vein reflecting greater impedance to venous return on the right as compared to the left; left and right transverse oblique sequence confirming the above findings on the coronal and transverse sequences; left sagittal sequence (image 30/70) displaying the backward displaced manubrium compressing the hazy gray proton dense thymus gland against the pulmonary artery and ascending aorta as the first division of the left subclavian artery enters the scalene triangle; aberrant right subclavian artery ascending posterior to the esophagus accentuating costoclavicular compression of the left gray proton dense brachiocephalic vein against the aorta, image 37/70 (Figure 5).

2D time-of-flight (TOF) MRA/MRV stacked image (1/168) (Figure 6) displays the high proton dilated dense aberrant right subclavian artery as it exits the aorta giving origin to the very small right vertebral artery;

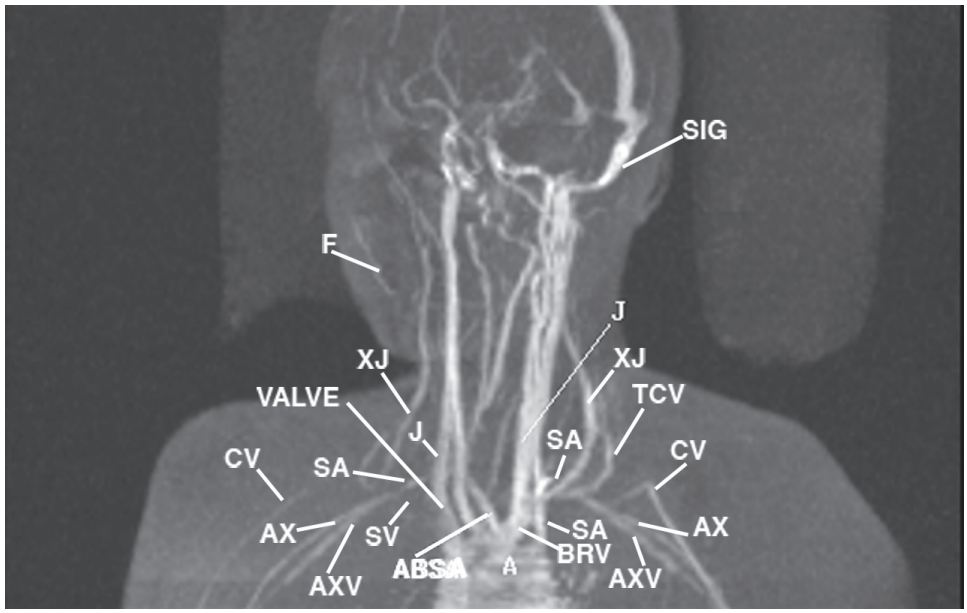

Figure 7. Coronal 3D reconstructed MRA / MRV

This 3D reconstructed image displays the head and neck leaning right; increased signal intensity of the dilated left external jugular vein (XJ) compared to the smaller right external jugular vein; mild compression of the second division of the right subclavian artery (SA) lateral to the right smaller internal jugular vein (J) with "cone shaped" compressed inferior bicuspid valve (valve) within the right internal jugular vein; compressed right subclavian vein (SV), proximal dilatation of the high proton dense left brachiocephalic vein (BRV) reflecting compression against the aorta (A) and the right common carotid artery (not labeled) anterior and separate from the aberrant right subclavian artery (AB.S.A); ascending high proton dense left subclavian artery reflecting costoclavicular compression at the second division of the artery; compression of the diminished signal intensity of right subclavian vein (SV) as drains into the right brachiocephalic vein. Observe the dilated right vertebral vein (not labeled) impeding venous return into the right brachiocephalic vein.

AX-Axillary artery, AXV-Axillary vein, BRV-Brachiocephalic vein, CV-Cephalic vein, SIG-Sigmoid sinus, TCV, transverse cervical vein.

right vertebral vein draining the high proton dense right sigmoid sinus; high proton dense longitudinal sinus/ superior sagittal sinus draining into the dominant left transverse, sigmoid and left internal jugular vein; high proton dense impedance to venous return in the right facial vein as compared the left facial vein; asymmetric right and left cephalic veins and sigmoid sinuses; 3D reconstructed, image 7/168 (Figure 7), dominant high proton dense left internal jugular vein, as compared to the smaller right, draining into the proximal dilated high proton dense left brachiocephalic vein and the high proton dense aberrant right subclavian artery ascending and decreasing signal intensity into the compressed second division of the artery and the "cone shaped" compressed inferior bicuspid valve in the smaller right internal jugular vein.

Bilateral vertebral venous plexus diverts venous drainage from the spinal cord medial to the asymmetric vertebral arteries, image 11/168 (not displayed) decreased signal intensity of the second division of the right subclavian artery reciprocally increasing signal intensity into the high proton dense third division of the subclavian artery paralleling the proximal right axillary vein as it decreases signal intensity into the compressed right subclavian vein, images 12/168–15/168, (not displayed).

Bilateral abduction external rotation of the upper extremities coronal sequence displayed the posterior inferior rotation of the clavicles with the subclavius muscles and the posterior anterior medial rotation of the coracoid processes with attached muscles enhancing costoclavicular compression of the draining veins within the neck, supraclavicular fossae with lymphatics and compression of the subclavian and axillary arteries with binding nerves, evident by the intermediate gray proton dense right subclavian vein on the first rib, dilated valveless right anterior jugular vein as compared to the left and the proximal dilatation of aberrant subclavian artery posterior to the esophagus exiting the aorta (Figure 8).

Bilateral abduction external rotation (AER) of the upper extremities in the supine position triggered complaints over the right anterior chest wall and tingling and numbness of the right arm.

MRI/MRA/MRV CONCLUSIONS

1. Thin narrow thorax.
2. Bilateral round shoulders, right greater than left.
3. Dilated origin of an aberrant right subclavian artery coursing posterior to the esophagus ascending compressed into the scalene triangle with binding nerve roots.
4. Very small right vertebral artery as compared to the left.
5. Compressed inferior bicuspid valve within the right internal jugular vein as compared to the dominant left internal jugular vein.
6. Bilateral costoclavicular compression (laxity of the sling/erector muscles - trapezius, levator scapulae, serratus anterior muscle) of the bicuspid valves of the draining veins within the neck, supraclavicular fossae and compression of the subclavian and axillary arteries with binding nerves, right greater than the left.
7. Bilateral coronal abduction external rotation (AER) of the upper extremities captured images and triggered complaints of pain over the right anterior chest wall and tingling with numbness of the right arm.

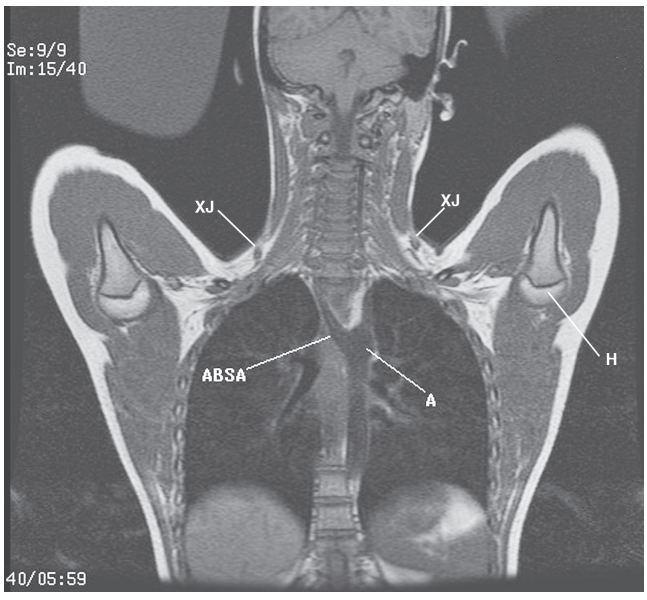

Figure 8. Coronal abduction external rotation of the upper extremities (AER) MRI Sequence

This abduction external rotation (image 15/40) displays head and neck leaning left with the elevated left shoulder as compared to the drooping painful right shoulder; costoclavicular compression of the subclavian arteries (not labeled) veins on the asymmetric first ribs (not labeled) and the enhanced dilatation of aberrant right subclavian artery (AB.S.A) as it exits the descending aorta (A). Observe the asymmetric external jugular veins (XJ) reflecting impedance to venous return, left greater than right. H-head of the left humerus.

DISCUSSION

The PA and lateral chest radiographs displayed mild kyphosis of the thoracic spine asymmetrically increasing the slopes of the first ribs, right lower than left, backward displacing the manubrium accentuating rounding of the shoulders and normal cardiomediastinal structures and lungs.

Multiplanar MRI captured images that displayed thin subcutaneous tissues; narrow thorax; dilated compressed aberrant right subclavian artery ascending posterior to the compressed esophagus into the right subclavian artery with binding nerve roots greater than on the left; diminished arterial flow to the right neck, supraclavicular fossa with dilated gray proton dense lymphatics as compared to the left; small compressed right internal jugular vein as compared to the dominant left internal jugular vein; diminished venous return from right axillary vein as compared to the left; dilated impedance to venous return from the dominant right vertebral vein impeding venous return from drainage into the right brachiocephalic vein as compared to the left; very small right vertebral artery as compared to the dominant left, and the patient leaning away from the painful right shoulder as compared to the left.

The 2D TOF MRA/MRV stacked and 3D reconstructed images cross-referenced the coronal, transverse, transverse oblique and sagittal sequences.

Bilateral abduction external rotation (AER) coronal sequence of the upper extremities captured functional anatomic images that triggered complaints of pain over the right anterior chest wall, and tingling with numbness of the right arm only.

Following the diagnosis of an aberrant right subclavian artery as above, she was referred to a vascular surgeon at UCLA who repeated her physical examination and concluded that the MRI of the brachial plexus showed a dilated origin of an aberrant right subclavian artery coursing posterior to the esophagus, ascending compressed into the scalene triangle with binding nerve roots. She also had a very small right vertebral artery compared to the left. There was bilateral costoclavicular compression of the bicuspid valves of the draining veins within the neck, supraclavicular fossa, and compression of the subclavian and axillary arteries with binding nerves, right greater than left.

The impression by the vascular surgeon was an aberrant right subclavian artery with symptoms suggesting dysphagia lusoria. She also had symptoms and findings suggestive of right upper extremity TOS with moderate impairment as gauged by her Quick DASH score. The surgeon discussed the symptoms with her mother in relation to her anatomy. Given her age and size he thought it would be preferable to delay any surgical intervention if possible. He remarked that surgery for correction of her dysphagia would require anterior scalene muscle division that would benefit her

right upper extremity TOS symptoms and therefore, referred her to a pediatric surgeon for consultation.

TAKE HOME MESSAGE

The PA and lateral chest with the AP cervicothoracic spine radiographs are necessary to detect any osseous or soft tissue anomalies prior to the MRI/MRA and the MRV of the brachial plexus. Anomalous vascular structures impede venous with lymphatic return and arterial flow as in this patient with an aberrant right subclavian artery. Any decrease in venous return increases intrathoracic, intracranial, and intra abdominal pressures and triggers patient complaints.[6] It would seem that she was misdiagnosed. Her mother was very concerned and did not know what to do. Were it not for discussion with others, she would not have considered the possibility of TOS. The history of difficulty in swallowing was the clue that should have linked the diagnosis to right aberrant subclavian artery, that would have been displayed by a **simple barium swallow**.[7] The referring physicians did not consider an aberrant subclavian artery in the differential diagnosis as a cause for difficult swallowing in a preteen patient. Magnetic resonance imaging details sites of landmark anatomy and costoclavicular compression impeding arterial, venous and lymphatic flow without the need for contrast agents. The diagnosis was confirmed immediately on viewing the coronal sequence at the imaging console. When using magnetic resonance imaging, you never know what anomaly will be displayed.[8]

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