Division of Vascular Surgery,
UPMC Heart and Vascular Institute

VASCULAR rounds

WINTER 2016

In This Issue

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Editor's Letter

The current issue of *Vascular Rounds* presents a collection of six unusual vascular vignettes that are not directly related to atherosclerotic disease, often misconstrued as the only underlying pathology that affects our blood vessels. We have listened to some of your comments requesting more case presentations, especially those that are somewhat obscure and controversial and go beyond the usual menu of vascular topics. We hope you will be interested in this format that we intend to provide once a year to add to the usual write-ups about new therapies and devices for the treatment of more common vascular problems.

With this issue of *Vascular Rounds* we also celebrate the one year anniversary of this publication intended to keep you informed of recent developments in vascular care. We have received feedback from some of you but would like to encourage all of our readers to let us know how we are doing. We are interested to know if you find *Vascular Rounds* to be of benefit to your practice and education. We are especially keen to know of perceived deficiencies and any suggestions to make this circular of more interest to you. You will find a link at the bottom of this page to a quick anonymous online survey that we hope you will take the time to complete. We respect your time, and promise it is short and to the point.

Best Regards,

Michel S. Makaroun, MD
Co-Director, UPMC Heart and Vascular Institute
Professor and Chief, UPMC Division of Vascular Surgery

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UPMC LIFE CHANGING MEDICINE

Nutcracker Syndrome as a Cause of Abdominal Pain

Eric Hager, MD
Assistant Professor of Surgery, UPMC Division of Vascular Surgery

Nutcracker syndrome is an unusual cause of flank pain and, on occasion, hematuria. The following case presented with flank pain and thigh varicosities.

Case Report

A 50-year-old healthy female presented to the office with a history of progressive left flank pain, exacerbated in the supine position. The pain was described as a dull ache or gnawing sensation radiating to the left hip. Over the months prior to the office consultation, the pain began to worsen with activity and prohibited her from her routine exercise regimen. She denied a history of weight loss or trauma and she was up to date on all cancer screenings. Her recent lab work was unremarkable aside from microscopic hematuria by urinalysis. Her physical exam was unremarkable aside from proximal left thigh varicosities which were non-tender. She underwent an abdominal duplex which revealed left renal vein compression between the aorta and superior mesenteric artery consistent with Nutcracker syndrome (Figure 1). The left ovarian vein was dilated and demonstrated retrograde flow into the pelvic and proximal thigh varicosities.

She underwent a venogram through a right femoral approach and we were able to successfully select the left renal vein and ovarian vein with a catheter and deploy several Interlock coils (Boston Scientific, Natick, Mass.) to stagnate flow and induce thrombosis (Figure 2). We then utilized intravascular ultrasound and identified the area of stenosis of the left renal vein. A 12mm self-expanding S.M.A.R.T.® CONTROL® stent (Cordis Corp, Miami Lakes, Fl.) was then deployed and the resulting venogram revealed proper stent placement with dramatic improvement of the venous stenosis and brisk flow into the vena cava (Figure 3). In the post anesthesia care unit the patient was able to lay supine with no abdominal discomfort. She was placed on dual antiplatelet therapy and discharged on postoperative day one in good condition.

She was seen back in the clinic for follow-up at one month and six months with complete symptom relief and had resumed all activity. An abdominal duplex was performed and revealed a widely patent renal stent with no evidence of external compression or stenosis (Figure 4). The gonadal vein was thrombosed and there was no reflux noted in the decompressed thigh varicosities.

Overview

Nutcracker syndrome is caused by symptomatic compression of the left renal vein between the superior mesenteric artery and aorta. This compression can cause reflux and dilatation in the pelvic, lumbar, adrenal,

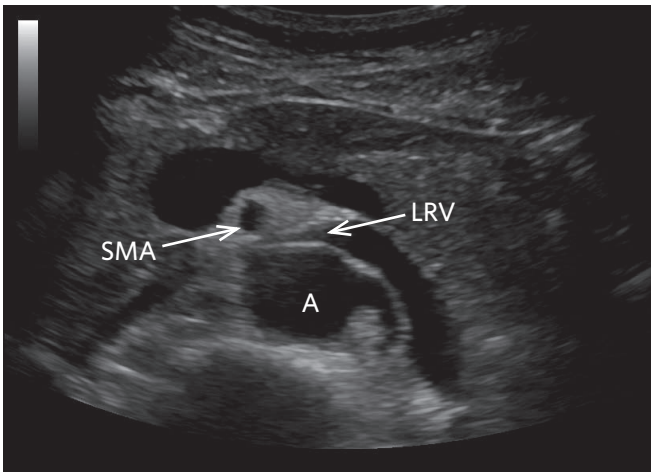

Figure 1:
Duplex ultrasound showing compression of the left renal vein (LRV) between the aorta (A) and superior mesenteric artery (SMA).

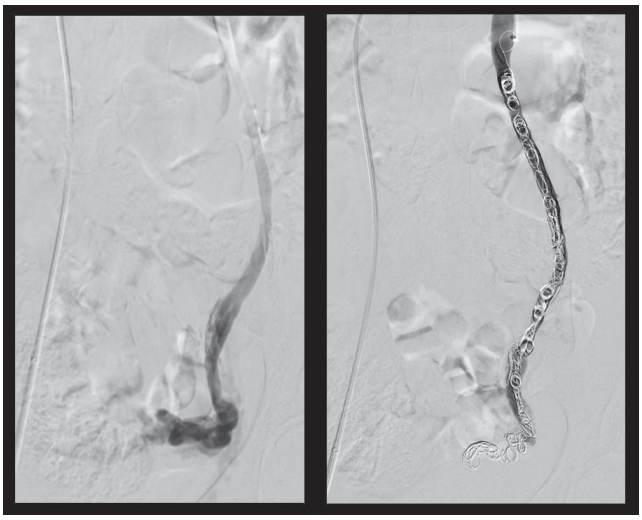

Figure 2:
Venogram demonstrating the refluxing ovarian vein before and after coil embolization.

and hemiazygos veins, which are all tributaries of the renal vein. Symptoms typically manifest as flank pain, pelvic congestion, and hematuria.

The majority of patients that present with Nutcracker syndrome are tall, athletic females in their third decade of life[1]. The symptoms are heterogenous and can range from asymptomatic microhematuria to incapacitating flank and/or pelvic pain. Oftentimes the diagnosis of nutcracker syndrome is one of exclusion as the majority of symptoms are gynecologic or urologic in nature. An astute clinician must have a high degree of suspicion in order to make the diagnosis. Once common causes of flank pain and hematuria are ruled out, duplex ultrasonography should be utilized to provide anatomic and hemodynamic data which can assist in the diagnosis. Calculating the peak velocity ratio (> 4) between the compressed and dilated venous segments may suggest nutcracker syndrome. In addition, reflux and dilation through the collateral ovarian and pelvic veins may be identified. Computed tomography with intravenous contrast timed for the venous phase may help with the delineation of the anatomy and aid in surgical planning.

Patients with severe symptomatic nutcracker syndrome can be managed in several ways. Observation is typically recommended for patients under 18 years of age due to the high likelihood that the lesion will spontaneously regress as the patient grows and the acute SMA-aorta angle increases thereby reducing the amount of vein compression[2]. While the patient ages, there has been suggestion that angiotensin inhibitors may improve orthostatic proteinuria that may be present[3]. For symptomatic adult patients, the options include open surgical reconstruction or endovascular therapies to alleviate the venous compression.

Surgical options include left renal vein transposition, renal vein bypass, auto-transplantation of the kidney, and nephrectomy[4]. Outcomes from these are generally good with the left renal vein transposition being the most commonly performed. Hartung et al. reported that 17 of 18 patients who underwent transposition became asymptomatic with minimal surgical complications[5]. Endovascular techniques have evolved and are now considered viable alternatives to open surgery and carry highly favorable results. Patients undergo venography with cannulation of the left renal vein. In women with pelvic congestion syndrome and demonstrated reflux in the ovarian vein, coil embolization may be performed. Using intravascular ultrasound, precise size measurements may be taken and the compressed vein may be stented. A recent retrospective analysis from China reported symptom relief in 59 of 61 patients with stented stenotic renal veins at a median five year follow-up[6]. Although the long term outcomes are unknown with endovascular techniques, many patients elect for the minimally invasive approach versus the open surgical options.

This case report is a demonstration of the endovascular treatment of nutcracker syndrome with a highly positive outcome. The patient presented with classic flank pain and pelvic congestion which was treated with coil embolization of the refluxing ovarian vein and stenting of the compressed renal vein. Her symptoms have resolved and her six month follow-up imaging reveals a patent stent with a successfully thrombosed ovarian vein.

References

1. K. Ahmed, R. Sampath, M.S. Khan. Current trends in the diagnosis and management of renal nutcracker syndrome: a review. Eur J Vasc Endovasc Surg, 31 (2006), pp. 410–416
2. Y. He, Z. Wu, S. Chen et al. Nutcracker syndrome—how well do we know it? Urology, 83 (2014), pp. 12–17
3. T.S. Ha, E.J. Lee. ACE inhibition can improve orthostatic proteinuria associated with nutcracker syndrome Pediatr Nephrol, 21 (2006), pp. 1765–1768
4. S.M. Said, P. Gloviczki, M. Kalra et al. Renal nutcracker syndrome: surgical options Semin Vasc Surg, 26 (2013), pp. 35–42
5. O. Hartung, D. Grisoli, M. Boufi et al. Endovascular stenting in the treatment of pelvic vein congestion caused by nutcracker syndrome: lessons learned from the first five cases. J Vasc Surg, 42 (2005), pp. 275–280
6. S. Chen, H. Zhang, H. Shi et al. Endovascular stenting for treatment of nutcracker syndrome: report of 61 cases with long-term follow-up J Urol, 186 (2011), pp. 570–575

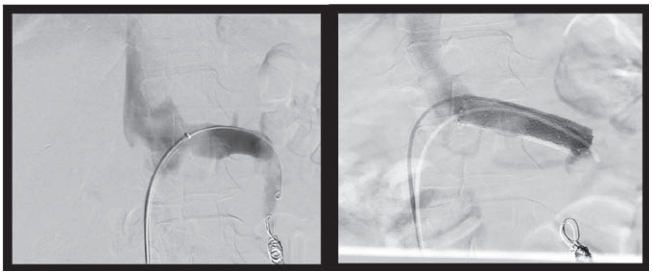

Figure 3:
Venogram demonstrating the left renal vein before and after stenting.

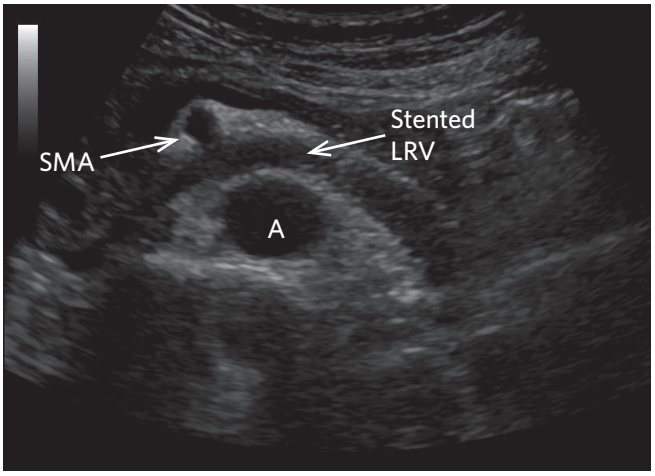

Figure 4:
Duplex ultrasound showing a patent renal stent. Left renal vein (LRV) between the aorta (A) and superior mesenteric artery (SMA).

Neurogenic Thoracic Outlet Syndrome

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Co-Director, Aortic Center

Thoracic outlet syndrome (TOS) is well documented in the medical literature but remains a diagnostic dilemma. The inherent challenges of making the diagnosis include vague, nondescript patient symptoms involving the upper extremity, persistent patient frustration due to pain and discomfort, conflicting physical exam findings, and the absence of a definitive diagnostic study. This case report describes the successful diagnosis, treatment, and recovery of patient with neurogenic TOS.

Case Report

A 16-year-old female was referred to our institution after nine months of right upper extremity and shoulder discomfort along with the complaint of having a "dead arm." The patient was a talented high school swimmer who had been competing nationally since the age of 12. The patient was evaluated by orthopedic and sports medicine specialists for her right arm discomfort and was initially treated with rest and physical therapy which focused on strengthening her acromioclavicular joint, biceps, and rotator cuff. Radiographic imaging and electrophysiology testing were unremarkable. Despite six months of abstinence from swimming, corticosteroid injections, continued physical therapy, and chiropractic care the patient's symptoms persisted and her quality of life deteriorated.

She was referred to the UPMC Division of Vascular Surgery for a second opinion. On evaluation the patient's history, posture, and physical exam findings were

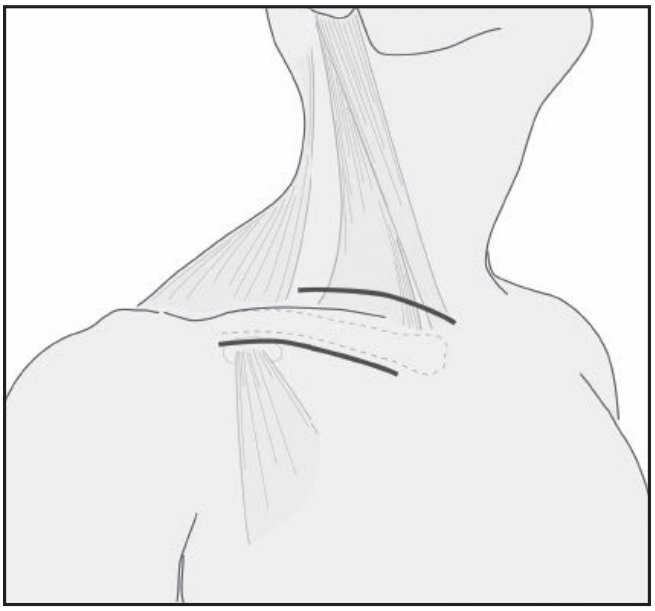

Figure 1:
Supra or infra-clavicular surgical exposure of the thoracic outlet.

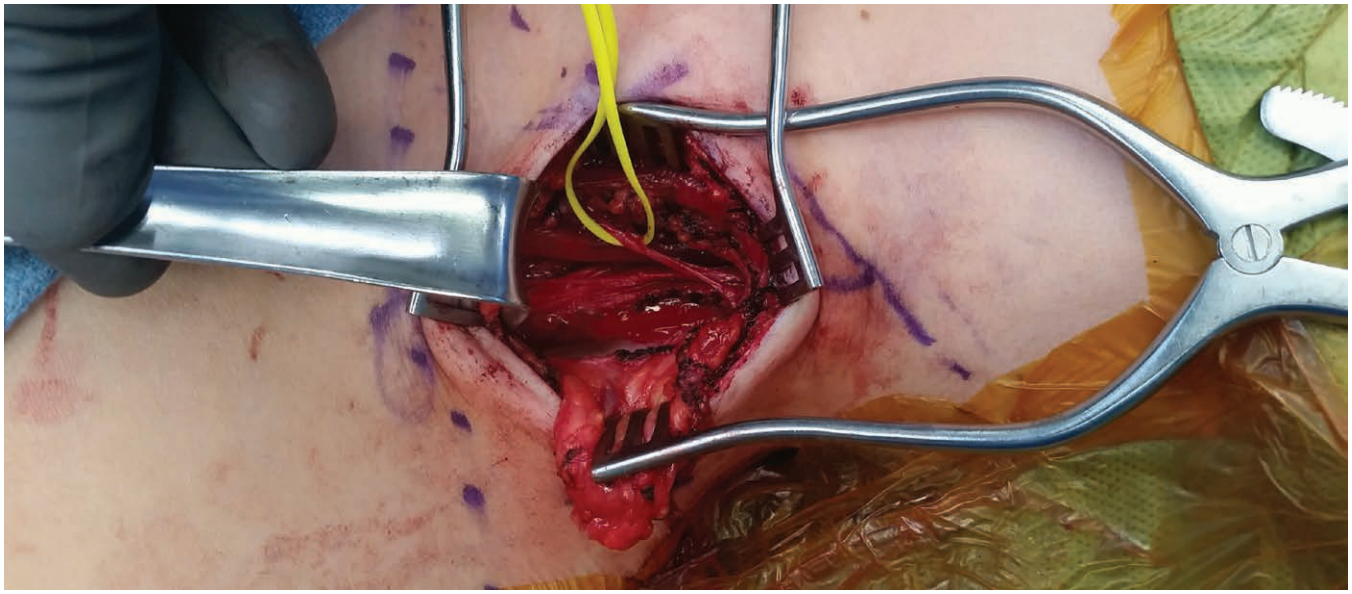

Figure 2:
Intraoperative photo of a hypertrophied middle scalene and anomalous scalene minimus muscles. Phrenic nerve is located by the vessel loop.

suggestive of neurogenic thoracic outlet syndrome. Her history was remarkable for the fact that she was a talented butterfly swimmer who lifted weights regularly up until 12 months ago. Her right arm was weaker compared to the left, she was right handed, and she had no prior history of chest or neck trauma. On examination she was physically fit with a slouching posture and tight trapezius muscles. Standard neurogenic thoracic outlet exertional maneuvers were performed and were found to be positive. She showed no features of pectoralis minor syndrome or venous or arterial TOS based on physical examination and duplex imaging. Cervical spine and chest radiographs did not reveal a cervical rib or abnormalities of the first rib or clavicle.

The patient was educated on neurogenic TOS and enrolled in an intense three month physical therapy regimen focusing on posture reconditioning, upper extremity range of motion, and strengthening and conditioning of the trapezius, rhomboid, and deltoid muscles. The patient responded to physical therapy at three months but had recurrent symptoms when reinitiating her exercise regimen. A scalene block alleviated her symptoms and supported the diagnosis of neurogenic TOS.

The patient elected to undergo TOS decompression, first rib resection, scalenectomy, and brachial plexus neurolysis via a supraclavicular approach (Figure 1). Intraoperative photos show the impressive hypertrophy of the patient's middle scalene muscle and an anomalous scalene minimus muscle which enveloped the entire brachial plexus (Figure 2). After performing a scalenectomy, first rib resection, and neurolysis, the skeletonized nerve roots were isolated (Figure 3). To prevent or limit the development of perineural scar tissue, a hyaluronidate-based absorbable film was wrapped around the brachial plexus. The patient's postoperative course was unremarkable, paresthesias resolved immediately, and she was discharged on postoperative day three. In follow-up, she returned to school the next week, completed six additional weeks of physical therapy, and returned to noncompetitive swimming at eight weeks. She started training with her swim team at three months and was once again swimming competitively at six months.

Overview

In many cases, the diagnosis of neurogenic TOS fails to be made in a timely manner. Delayed or inaccurate diagnoses often contribute to the patient's psychosocial concerns and potentiation of symptoms. Both surgical and conservative therapies have proven to be acceptable treatment options for patients with neurogenic TOS. A comprehensive history and thorough physical examination combined with select imaging modalities can successfully diagnose neurogenic TOS in most cases. Long-term surgical results, even in the competitive athletes are very favorable with symptomatic recurrence in a small number (10 to 20 percent).

Further Reading Suggestions

Sanders RJ, Hammond SL, Rao NM. Diagnosis of thoracic outlet syndrome. J Vasc Surg 2007;46(3):601-4.

Balci AE, Balci TA, Cakir O, Eren S, Eren MN. Surgical treatment of thoracic outlet syndrome: effect and results of surgery. Ann Thorac Surg 2003;75(4):1091-6.

Illig KA, Thompson RW, Freischlag JA, Donahue DM, Jordan SE, Edgelow PI. Thoracic Outlet Syndrome. Springer-Verlag London 2013.

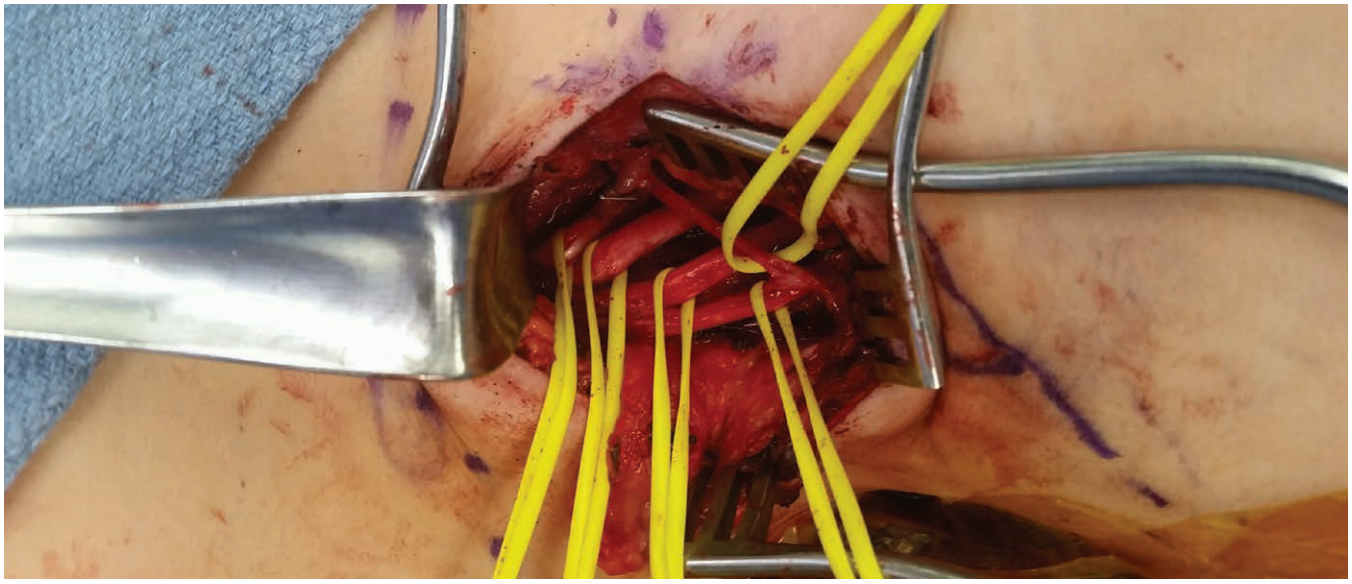

Figure 3:
Intraoperative photo after scalenectomy, first rib resection, and neurolysis. Note the skeletonized brachial plexus with successive vessel loops. The phrenic nerve is located by the vessel loop.

Arterial Duplex Examination Can Diagnose Anatomically Remote Conditions

Steven Leers, MD
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Duplex arterial examinations are often limited to the diagnosis of abnormalities related to accessible structures and the area being interrogated. As such, intracranial or intrathoracic pathologies are not usually amenable to superficial ultrasound exams. Some findings on routine evaluations can occasionally suggest remote pathology and vascular laboratories have to be alert to such subtle findings.

Occasionally, waveforms from the femoral vessels or the neck suggest intrathoracic pathology such as heart failure or valve abnormalities that can be pursued when previously undiagnosed. Whenever diffuse waveform abnormalities are seen, this should suggest a probable remote cause for the findings. For example, with severe aortic stenosis, all carotid waveforms and velocities will be abnormal without significant plaque seen. In the femoral vein, continuous low flow without respiratory variation suggests thrombus proximal to the vein (May-Thurner Syndrome). The following is a very unusual case of a carotid duplex scan that led to the diagnosis of an aortic dissection.

Case Report

A 79-year-old African American female presented to the emergency department after a syncopal episode. When she awoke, she was neurologically intact and completely asymptomatic. Her past history was positive for hypertension and dyslipidemia. An echocardiogram showed good left ventricular function and a cardiology consultation suggested hypovolemia. No other abnormalities were noted. Because of the neurologic event, a carotid duplex scan was requested.

Velocity analysis was not diagnostic of any severe stenosis but revealed a widened waveform suggestive of proximal disease.

The B mode scan revealed an elevated flap in the right common carotid artery that suggested a dissection (Figures 1 and 2). Since a similar abnormality was also noted on the left and flow velocities were unusually widened in both common carotid arteries it was suspected that the flap was an extension of an aortic dissection, which is not expected in a currently asymptomatic patient. CT angiography was recommended and documented a complex type A dissection involving the aortic arch and extending into the carotid artery (Figure 3). The patient was brought immediately to the operating room for surgical treatment.

Overview

Findings on both grayscale imaging and flow velocity patterns can often suggest remote pathology to the ultrasound technologist interrogating the carotid or femoral vessels. These should not be discounted as they can bring attention to an undiagnosed medical condition. In this case, the carotid ultrasound findings suggested a previously unexpected and life-threatening diagnosis and directed emergency therapy.

Further Reading Suggestion

Rohren EM, Kliewar MA, Carroll BA, Hertzberg BS. A Spectrum of Doppler Waveforms in the Carotid and Vertebral Arteries. AJR 2003; 181:1695-1704 0361-803X/03/1816-1695.

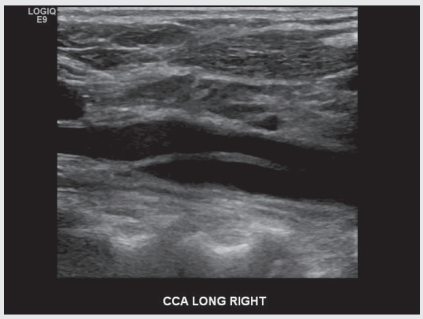

Figure 1:
Ultrasound image demonstrating dissection flap in the right common carotid artery (long axis).

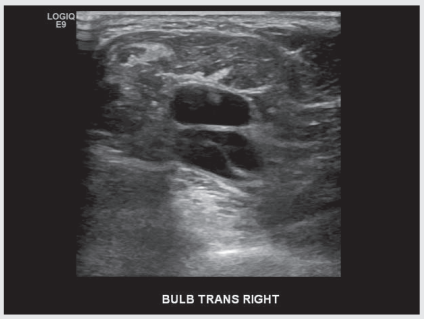

Figure 2:
Ultrasound image demonstrating dissection flap in the right common carotid artery (short axis).

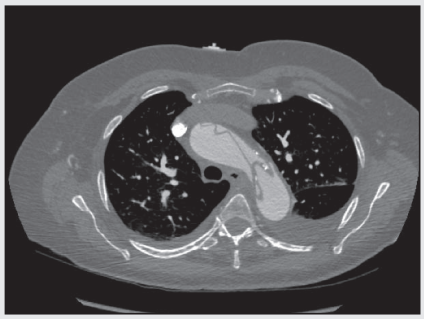

Figure 3:
CT angiography demonstrates complex aortic dissection involving the aortic arch.

Native Aortic Coarctation in an Adult Patient

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Professor of Surgery, UPMC Division of Vascular Surgery
Chief of Vascular Surgery, UPMC Presbyterian

Raymond E. Eid, MD
UPMC Vascular Surgery Fellow

Aortic coarctation usually presents in relatively young individuals with upper extremity hypertension and lower extremity symptoms. The following is a rather late presentation of this congenital abnormality.

Case Report

A 62-year-old female presented to her primary care physician with generalized fatigue, progressive shortness of breath, and dyspnea on exertion.

Her past medical history is significant for severe hypertension (maintained on three different antihypertensive medications), and hyperlipidemia. Her physical examination was remarkable for faint femoral pulses bilaterally.

A transthoracic echocardiogram demonstrated a normal ejection fraction with normal right ventricular function, but showed severe mitral regurgitation. She underwent a cardiac catheterization to evaluate her valvular heart disease and coronary artery disease prior to her mitral valve replacement. The ascending aorta could not be accessed from a femoral approach, and the interventional cardiologist instead used the right radial artery to access the heart. The catheterization revealed normal coronary arteries. An aortogram was performed because of the initial difficulty and showed a dilated aortic root with severe coarctation of the proximal segment of the descending aorta with a pressure gradient of 68mmHg across the stenosis. This was further confirmed with a computed tomography angiography for better evaluation of the extent of coarctation (Figure 1A).

The patient was then referred to the UPMC Division of Cardiac Surgery for mitral valve replacement and simultaneous treatment of her coarctation. After a multidisciplinary discussion, the cardiac and vascular teams decided to proceed with an endovascular repair of the aortic coarctation prior to the valve replacement. This would allow shorter operative time and enhanced hemodynamics, primarily afterload reduction of the left ventricle and better perfusion of her lower extremities, prior to her valve replacement.

As planned, she was taken to the operating room angiography suite with the vascular surgery team. Through a femoral access, she underwent an aortogram that confirmed the coarctation and the elevated gradient across the stenosis (Figure 1B). This was treated with an endovascular aortic endograft with favorable results (Figure 1C). Her gradient following stent deployment normalized (< 5 mmHg). She was taken the next day by the cardiac surgery team for her mitral valve replacement. She tolerated both procedures well and was discharged home on postoperative day five.

Overview

Aortic coarctation is a narrowing of the aortic isthmus, the site of insertion of the ductus arteriosus, distal to the left subclavian artery. It constitutes five to eight percent of congenital vascular malformations and is most common in males. This condition generally results in left ventricular afterload failure. It is typically congenital however it may be rarely acquired secondary to Takayasu's arteritis or severe atherosclerosis.

There are multiple forms, simple vs. complex, depending on the extent of the coarctation. It is associated with bicuspid aortic valves, intracranial arteries, and acquired intercostal artery aneurysms.

Hypertension is the classic presenting symptom in previously undiagnosed adults. Most patients are asymptomatic secondary to compensatory mechanisms, including left ventricular myocardial hypertrophy and the development of collateral blood flow through the intercostal, internal mammary, and scapular vessels. If hypertension is severe and poorly controlled, patients may complain of headache, epistaxis, heart failure, or aortic dissection. In addition, some patients may complain of lower extremity claudication, primarily on exertion.

The American College of Cardiology (ACC) and American Heart Association (AHA) guidelines recommend treatment of aortic coarctation when gradient > 20 mmHg, or < 20 mmHg with radiological evidence of significant collaterals, or in cases of hypertension and > 50 percent narrowing of the aortic diameter at the site of coarctation. If untreated, the average life expectancy is 35 years of age, with 75 percent mortality by 46 years of age. This is due to severe systemic hypertension, accelerated coronary artery disease, stroke, aortic dissection, and heart failure.

Summary

Our patient represents an unusual late presentation of aortic coarctation. At UPMC, we recommend corrective intervention for all patients who meet one criterion of the ACC/AHA guidelines mentioned previously. When indicated, intervention should be attempted as early as possible for optimal results. Both open and endovascular interventions have good results, and a decision should be made after multidisciplinary discussion considering the age of the patient and the morphology of the underlying pathology. Endovascular repair of aortic coarctation is feasible, safe, and effective not only in pediatric but also in adult patients, and is associated with minimal morbidity and mortality.

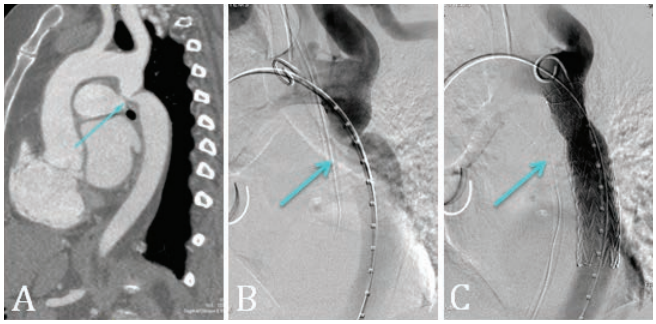

Figure 1:
CT angiogram (A) and initial intraoperative aortogram (B) showing the aortic coarctation. Completion angiogram (C) following deployment of the aortic stent graft, with resolution of the stenosis.

An Unusual Case of Claudication and Leg Ischemia in a Teenager

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Raymond E. Eid, MD
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Popliteal artery entrapment syndrome (PAES) is a relatively rare condition that occurs as a result of compression of the popliteal artery by surrounding muscles and tendons during exercise, usually as a result of an abnormal anatomic structure. Affected individuals often present with vascular claudication, however it can also lead to acute limb ischemia. Diagnosis is usually suspected by loss of pedal pulses during active plantar flexion and occasionally dorsiflexion, and confirmed by imaging with MRA or CTA. The following is a very unusual case where the offending structure was not related to abnormal muscles or slings but a bony abnormality.

Case Report

A 16-year-old healthy male presented with a sudden onset of pain in his left calf at rest. He subsequently developed calf ecchymosis. He was initially evaluated by an orthopedic surgeon who ordered an ultrasound and an MRI of his knee. This revealed swelling and possible tendon rupture. However, there was a concern for a possible band compressing the popliteal artery on the MRI and he was referred to the UPMC Division of Vascular Surgery for further evaluation. He is otherwise very active and is a competitive soccer player with no previous similar episodes or leg claudication.

His vascular exam was normal, with no signs of ischemia with provocative maneuvers. His imaging studies were re-evaluated and no clear compression of the artery was seen. He was managed conservatively considering the low suspicion for PAES and the presence of a ruptured tendon.

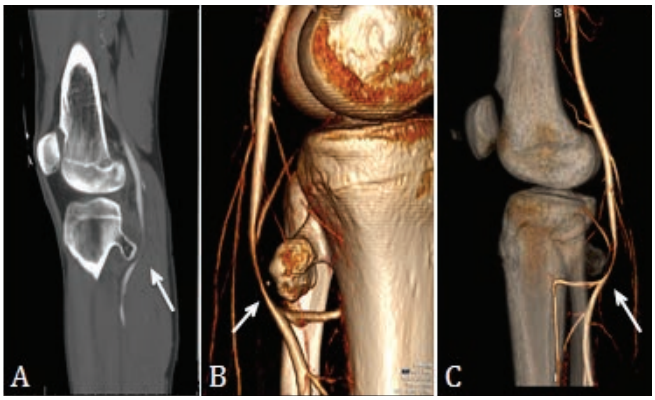

Figure 1:
CT angiogram (A) with 3-D reconstruction (B and C) showing the compression of the popliteal artery by the osteochondroma.

His symptoms continued to improve, however he presented again four months later with unprovoked and persistent numbness in his left foot. This was again not associated with ischemic changes or symptoms. His vascular exam at his second visit was unchanged and normal, with intact motor function. An arterial duplex ultrasound was performed in the office. This revealed external compression of the left popliteal artery at rest and during plantar flexion. A CT angiogram was obtained and demonstrated an approximately 70 percent compression of the infrageniculate popliteal artery by an osteochondroma extending posteriorly from the proximal tibia (Figure 1). There was worsening of the compression with plantar flexion. No abnormal slip of the gastrocnemius muscles around the popliteal artery was present to suggest classical popliteal entrapment.

The patient underwent surgical resection of his osteochondroma. This was performed through a posterior popliteal approach. The popliteal vessels and neurovascular bundle were mobilized and the bone was resected. The artery appeared normal with no evidence of narrowing or scarring. No vessel reconstruction was required. He was discharged the next day and was seen one month later in the office and was doing well with complete resolution of his symptoms and no evidence of vascular compression on repeat ultrasound imaging.

Overview

Lower extremity ischemia or claudication in teenagers and young adults is rare. Common causes include PAES, cystic adventitial disease of the popliteal artery, and musculoskeletal causes such as tendinitis or stress fractures. PAES is a result of compression of the popliteal artery by muscles or fibers with abnormal insertions and anatomy. It is most commonly seen in young and middle-aged men, and can manifest itself with significant disability and impairment.

According to the popliteal vascular entrapment forum classification, PAES is classified into six types. Type I is characterized by an atypical course of the popliteal artery, type II occurs as a result of an abnormal insertion of the medial head of the gastrocnemius muscle, type III occurs as a result of an accessory slip of gastrocnemius/fibrous bands arising from the medial head of the gastrocnemius muscle, type IV occurs when the popliteal artery passes underneath the popliteus muscle, type V is primarily venous entrapment, and type VI encompasses other variants including occasionally no clear abnormality. The abnormal anatomy of the popliteal artery and its surrounding tendons causes extrinsic compression which ultimately leads to irreversible scarring and narrowing of the artery. If left untreated, this results in occlusion, or distal embolization.

Unfortunately, delayed diagnosis due to young patient age, low index of suspicion, and lack of atherosclerotic risk factors is common.

Clinical diagnosis is made with a careful history, physical examination, and imaging studies, but most importantly a high index of suspicion and recognition that even young adults can have arterial claudication. Imaging studies during active plantar flexion or passive dorsiflexion of the foot will frequently demonstrate compression of the popliteal artery and can include arterial duplex ultrasonography, magnetic resonance angiography (MRA), or computed tomography angiography (CTA). Other causes of arterial compression can also be identified by these studies, including adventitial cysts or as illustrated in this case, a bony abnormality. Occasionally, symptoms can be associated with elevated compartment pressures and hypertrophied muscles.

The most common presenting symptom is intermittent claudication in a young or middle-aged athletic patient with no atherosclerotic risk factors.

Once the diagnosis is made, treatment options should be discusssed promptly to release the external compression and preserve popliteal arterial flow before irreversible changes to the vessel wall occur.

Summary

Our patient represents a very atypical case of PAES . Although his compression is less dynamic and not related to fibromuscular structures as seen in traditional PAES, the presenting symptoms and goals of management are similar.

At UPMC, we recommend taking all claudication and ischemic symptoms in young patients seriously and considering PAES as a possible etiology. Early diagnosis and treatment has favorable outcomes with full functional recovery and avoidance of arterial occlusion and the need for late bypasses.

Upcoming Events

Friday, April 22, 2016

3rd Annual Advances in Cardiovascular Care Symposium
Cumberland Woods Village (next to UPMC Passavant) – Allison Park, Pa.

This one-day CME event will feature a range of case-based discussions presented by the UPMC Heart and Vascular Institute faculty and primary care providers. There will be a variety of cardiovascular topics, including transcatheter aortic valve replacement (TAVR), minimally invasive mitral valve surgery, stroke prevention for patients with atrial fibrillation, heart failure and cholesterol management medications, and vascular surgery updates.

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Save the Date: Aug. 26-27, 2016

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To learn more, contact Mark Byrne at 412-802-3031 or byrneme@upmc.edu.

Disseminated Intravascular Coagulation due to Persistent Endoleak after Endovascular Aneurysm Repair

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Disseminated intravascular coagulation (DIC) is a syndrome characterized by the systemic activation of blood coagulation, which generates intravascular thrombin and fibrin, resulting in the thrombosis of small- to medium-sized vessels and ultimately organ dysfunction and severe bleeding. DIC may present as a complication of infection, solid cancers, hematological malignancies, obstetric diseases, trauma, aneurysms, and liver diseases, each of which presents characteristic features related to the underlying disorder. The following is a case of DIC resulting from a very unusual source.

Case Report

A 76-year-old male was transferred to UPMC Presbyterian with a one month history of increasing bruising and a 10-day oozing from his tongue and oral cavity after a tongue biopsy and cauterization (biopsy was negative for malignancy). His blood workup suggested severe DIC with markedly decreased platelets to 80x109/L, fibrinogen < 60mg/dl (normal 200-375mg/dl), D-dimer 55,000ng/ml (normal < 250ng/ml), prolonged PT and PTT times, hemoglobin 6.6 g/dl, and normal liver function tests.

His past medical history included hypertension, hyperlipidemia, and stage 4 chronic kidney disease. He had a partial left nephrectomy for a benign neoplasm in 2012 and an endovascular aneurysm repair with a GORE EXCLUDER® AAA Endoprosthesis (Gore Medical, Flagstaff, Ariz.) device in 2009. His aneurysm shrunk down to 4.4cm during the three year follow up, but a year ago (September 2014) it was found to have increased in size reaching 6cm. A significant mixed type I and II endoleak was identified and using CO2, the inferior mesenteric and two lumbar arteries were coiled, and a proximal cuff and a right distal extension were deployed and sealed the endoleaks. Of note, the left renal artery was occluded. At the six month ultrasound follow up the aneurysm was stable in size at 6cm.

During his current admission for bleeding, the patient underwent an extensive workup including a haptoglobin test and reticulocyte count to rule out hemolysis and a peripheral smear to rule out leukemia. Flow cytometry was negative. Malignancy biomarkers, CEA, and PSA were also negative. Non-contrast CT of the chest, abdomen, and pelvis did not show any malignancy, however, the aortic aneurysm had increased in size to 6.6cm. An ultrasound scan could not identify any endoleaks.

The patient received 10 units of cryoprecipitate and four units of packed red blood cells; within three days his symptoms resolved and hematologic tests normalized. The patient was discharged with a diagnosis of unclear DIC, but a suspicion that the expanding AAA may be related.

Fifteen days later, lab tests indicated that the DIC had recurred and transfusions were again initiated. An abdominal duplex scan revealed the AAA to have increased to 7cm with perigraft flow indicating an endoleak. At this point, and as there was no other clear etiology for the DIC, management of the endoleak was entertained. One of the challenges was his chronic kidney disease but he was willing to start dialysis earlier if needed, seeking to have his problem resolved

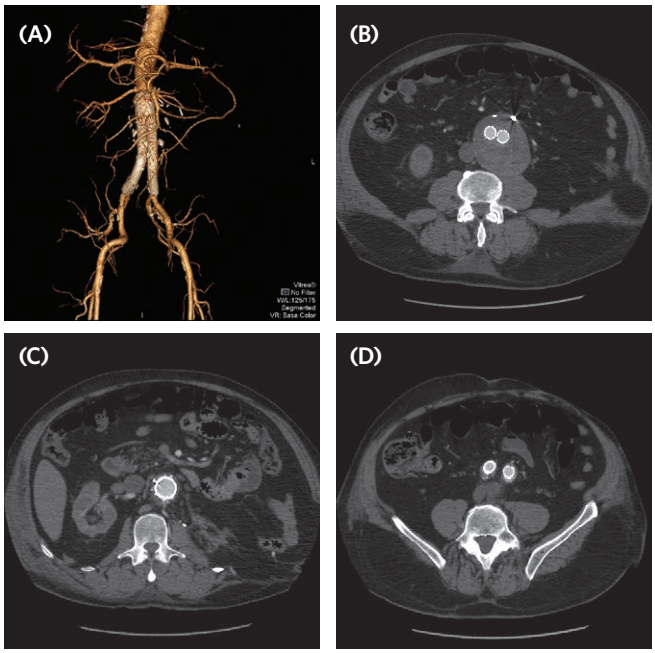

Figure 1:
CT Angiogram showing (A) an endograft in place and (B) an expanding 7cm aneurysm with poor apposition of all attachment points: (C) proximal aortic neck, (D) right and left iliacs with suspicion of type IB endoleak.

sooner rather than later. A CTA was performed and showed poor graft apposition in the neck and both iliac arteries with a suspicion for bilateral type IB endoleaks. (Figure 1). He was taken to the operating room and had bilateral limb extensions to the external iliac arteries and a proximal cuff which covered the right renal artery to increase the proximal sealing zone with no contrast used. Perfusion to the right kidney was maintained using a covered stent into the renal artery overlapping with the proximal cuff (chimney technique, Figure 2). Postoperatively all laboratory parameters and peripheral smears gradually improved and eventually normalized. His kidney function returned to his baseline and he did not require any dialysis.

Literature Review

DIC related to an endoleak after an endovascular aneurysm repair is very rare and very few such cases have been reported in medical literature. All cases were a result of large, delayed type I or III endoleaks with significant aneurysm sac expansion, with or without concomitant liver disease. It is hypothesized that turbulent flow and a large cavity stimulates the process of clot formation and lysis. Denuded endothelium and tissue factor release leads to activation of coagulation factors, excess generation of thrombin, chronic consumption of clotting factors, and simultaneous excess plasmin generation and fibrinolysis of the clots. This is the first case we have encountered at UPMC that illustrates that possibility.

Although specific recommendations for an endoleak induced DIC management are not available, the cornerstone of DIC treatment is providing treatment for the underlying disorder, in this case managing the endoleak. Supportive measures that are used until the underlying disorder is identified and treated include the administration of platelet concentrates and fresh frozen plasma in those with active bleeding or those at high risk of bleeding. The threshold for transfusing platelets depends on the clinical state of the DIC patient, usually when the platelet count is $\leq 50 \times 109$ /L. Heparin and derivatives (e.g. low-molecular-weight heparin) can interrupt the vicious cycle of thrombin generation and ameliorate the DIC and can be used sparingly as needed. Low-molecular-weight heparin is preferable to unfractionated heparin. After the endoleak management, serial follow-up of platelet counts, fibrinogen, and D-dimer levels are necessary to ensure resolution of DIC, as platelets and fibrinogen can rise as acute-phase reactants due to surgical intervention alone.

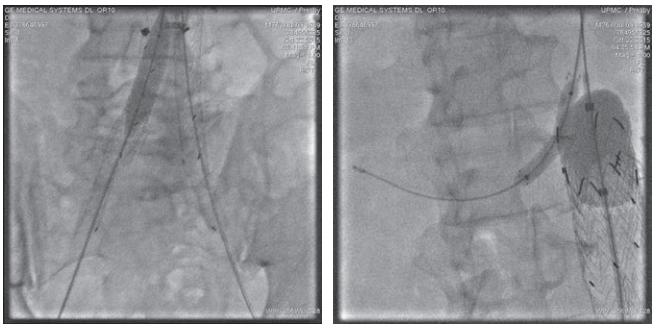

Figure 2:
Intraoperative imaging indicating bilateral iliac limb extension into the external iliac arteries and proximal aortic cuff placement with right renal artery stenting (ballooning of aortic cuff and renal stent).

Further Reading Suggestions

Patel AS, Bell R, Hunt BJ, Taylor PR. Disseminated intravascular coagulation after endovascular aneurysm repair: resolution after aortic banding. J Vasc Surg 2009;49:1046-9.

Keo HH, Diehm N, Baumgartner I, Schmidli J, Do DD. Disseminated intravascular coagulopathy caused by endoleak type I: successful treat-ment by endovascular stent-graft extension. EJVES Extra 2006;12:68-70.

Higashiura W, Kichikawa K, Sakaguchi S, Kubota Y, Nagata T, ishimine K, et al. Deteriorating consumptive coagulopathy with type III endoleak following endovascular repair for abdominal aortic aneu-rysm associated with liver cirrhosis. J Endovasc Ther 2007;14:421-5.

Nienaber JJ Duncan AA, Oderich GS, Pruthi RK, Nichols WL. Operative and nonoperative management of chronic disseminated intravascular coagulation due to persistent aortic endoleak. J Vasc Surg 2014;59:1426-9

Wada H, Matsumoto T, Yamashita Y. Diagnosis and treatment of disseminated intravascular coagulation (DIC) according to four DIC guidelines. J Intensive Care 2014;2:15

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