You're the Flight Surgeon

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You are a flight surgeon on a military base. During a routine clinic, a 35-yr-old, left-handed male Air Force helicopter pilot presents complaining of left arm swelling, weakness, and intermittent paresthesias into his left hand over the last week. He notices the symptoms primarily while lifting the collective (horizontal lever that controls the pitch of the rotor blades) with his left arm during routine flights. The maximum exertion required for this action is less than 15 lb of force. He reports working out regularly, including weight lifting, although he doesn't recall a specific vigorous workout prior to the onset of his symptoms. He denies any other injuries or trauma to his arm, shoulder, or neck, although states his neck is mildly sore since painting several rooms in his house 5 d ago. He denies any recent surgeries, periods of immobility, or prolonged periods of travel.

On chart review, his last visit was 2 mo ago for a routine flight physical. At that time there were no significant abnormalities noted, including no musculoskeletal complaints. His past medical history is unremarkable and he has had no prior surgeries. He takes no medications other than a daily vitamin. His family history is notable for a brother who survived marginal B cell lymphoma. His younger brother died of a brain tumor at 5 yr old. There is no family history of blood clots or other clotting disorders. His immunizations are up to date and appropriate for his age and military service with no vaccinations in the last 2 mo. He smokes (one pack per day for the last 10 yr) and is a social drinker. Occupationally, he has been a search and rescue helicopter pilot for the military for the last 12 yr with a total of 2000 flight hours. He denies having problems lifting the collective previously.

His review of systems is unremarkable. He denies fevers, weight loss, night sweats, dyspnea, or chest pain. On physical exam, his vital signs are within normal limits. He is alert, oriented, and in no distress. He has mildly decreased breath sounds bilaterally at the lung bases, but no wheezing. His heart sounds are normal without murmurs or extra heart sounds. His musculoskeletal exam shows normal range of motion in his bilateral upper extremities as well as 5/5 grip strength, biceps, and triceps strength. Pain in his left shoulder worsens with maximum overhead extension. He has moderate swelling that starts in his left arm from his mid bicep down to his fingers. His left distal radial pulse is normal. His upper extremities are normal to light touch with intact reflexes.

1. What is the MOST reasonable first step in his evaluation?

A. Chest X-ray.
B. Chest computed tomography (CT) without contrast.
C. Labs – complete blood count, prothrombin time/partial thromboplastin time/international normalized ratio, erythrocyte sedimentation rate.
D. Upper extremity ultrasound.

ANSWER/DISCUSSION

1. D. Differential diagnosis for this patient's current symptoms includes vascular, musculoskeletal, or anatomic/structural problems. The most concerning and potentially life threatening of these possibilities is a vascular obstruction or deep venous thrombosis (DVT), which could lead to limb pathology as well as pulmonary embolism (PE). Given the relatively acute nature of the unilateral swelling, a DVT diagnosis is possible, making the most appropriate initial evaluation an upper extremity ultrasound.[3,6] A chest X-ray or a chest CT scan without any pulmonary symptoms is less likely to identify the underlying pathology given the patient's signs and symptoms. Laboratory studies might be necessary, as the evaluation continues to help determine any underlying thrombotic tendencies, but are not likely to impact the initial treatment plan.

In this patient's case, his swelling triggers an ultrasound, which demonstrates a left subclavian venous thrombus with partial thrombosis of the axillary vein. He has no other DVTs. Primary spontaneous upper extremity DVT is a relatively rare event, comprising only 1–4% of all DVTs.[12] Secondary causes of upper extremity DVT, such as the sequela of a vascular catheterization or paraneoplastic thrombosis, make up a significantly larger percentage of upper extremity DVTs.[11] PE is concomitantly found in 13–17% of cases of upper extremity DVTs, which is higher than the rate of 8% of PE found in conjunction with lower extremity DVTs.[3] Given the high index of suspicion of PE with upper extremity DVTs, this patient has further evaluation with CT angiography and is found to have an asymptomatic, lower left lobe PE.

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2. What is the most accurate diagnosis for this patient at this time?

A. Venous thoracic outlet syndrome (VTOS).
B. Lymphadema secondary to undiagnosed paraneoplastic thrombosis.
C. Lymphoma.
D. Previously undiagnosed thrombophilia.

ANSWER/DISCUSSION

2. A. This patient has VTOS based on his clinical presentation: a young, active person with an upper extremity DVT that occurred in the absence of any predisposing factors. Thoracic outlet syndrome (TOS) is a spectrum of disorders involving the compression of the subclavian vessels or the brachial plexus as these structures traverse the thoracic outlet, which is the anatomic area from the supraclavicular fossa to the axilla that passes between the clavicle and the first rib.[7] The underlying pathology of most VTOS is a congenital or acquired musculo-ligamentous abnormality in the anterior thoracic outlet that causes recurrent microvascular damage and subsequent thrombosis of the subclavian vein.[2,8,11] VTOS represents only 3–5% of all TOS.[7] Venous obstruction is much more common than arterial obstruction.[12] VTOS often presents with acute swelling, whereas the most common type of TOS, neurogenic TOS, frequently presents with an indolent course of intermittent paresthesias, pain, or muscle weakness.[7] However, VTOS can be positional or cause intermittent or partial obstruction leading to inconsistent and intermittent symptomatology.[2] Therefore, the diagnosis is often delayed after significant evaluation and treatments. Acute, complete vascular obstructions are often identified by a blue, swollen, heavy, or aching upper extremity. Chronic obstructions have less swelling and dilated veins form in the neck, anterior chest, or axilla as collateral circulation.[2,12]

3. Which of the following is a common risk factor for VTOS?

A. History of humerus fracture.
B. Females between 15 and 25 yr old.
C. Athletes (especially those involved in repetitive overhead activities).
D. Exposure to prolonged flights.

ANSWER/DISCUSSION

3. C. The major risk factors for VTOS include male sex, age in the fourth decade of life, individuals who are physically active or have occupational or athletic activities that involve repetitive overhead movement, a personal history of thrombophilia, or a history of congenital or acquired abnormalities in the thoracic outlet region.[2,8] The most common subtype of VTOS, called Paget-Schroetter syndrome, is effort-induced subclavian vein thrombosis.[2] These patients can have "herald" events characterized by intermittent symptoms of subocclusive thrombotic events that spontaneously resolve.[11] These events are often triggered by upper body physical activity and dehydration.[11] A history of repetitive activities of the affected limb is found in 80% of cases with symptomatic VTOS.[1] With manipulation of the vasculature, such as with venous catheters or pacemaker wires, patients are at significantly increased risk of axillosubclavian DVT, although these cases do not represent true VTOS cases.[11]

This patient has several risk factors for VTOS, including sex, age, recent repetitive overhead activities (painting his house), and being overall physically active.

4. What is the best treatment option for this patient given his desire to fly again in the future?

A. Oral anticoagulation 12 mo, then discontinue the medication and observe for 6 mo for recurrence.
B. Undergo balloon venoplasty of his subclavian vein.
C. Thrombolytic therapy to dissolve DVT rapidly followed by endovascular stent placement.
D. Undergo surgical decompression (remove first left rib and scalene muscle).

ANSWER/DISCUSSION

4. D. There are several treatment options, depending on the severity of stenosis with variable success rates, complications, and residual long-term effects. Conservative treatment includes physical therapy, limb elevation, oral anticoagulation, compression of the affected extremity, pain control, and activity limitations.[1,8] These noninvasive treatment options are more commonly employed with nonspecific, neurogenic TOS or those patients with VTOS who are not surgical candidates.[4] While these methods do not cause further damage to the vessel, there is the significant possibility of residual disability, symptoms, and recurrent thrombosis.[8,11] Evidence suggests that 39–68% of VTOS patients treated conservatively will have permanent disability, persistent symptoms, and recurrent thrombus.[1,2]

Minimally invasive methods are also useful in removing the acute clot. Catheter-directed thrombolysis is most effective if done within 14 d of clot formation, but the likelihood of long-term thrombosis recurrence ranges from 23–33%.[8] Angioplasty prior to surgical decompression is ineffective with a high rate of recurrent stenosis or thrombosis, due to continued extrinsic mechanical compression of the vein.[1,11] In VTOS cases, angioplasty is generally reserved for residual stenosis postsurgery. Endovascular stents have very poor outcomes with high recurrent thrombosis rates.[12]

The only definitive treatment for VTOS caused by anatomic abnormalities is surgical outlet decompression, which involves complete removal of the first rib and frequently scalenectomy.[8] Surgical decompression successfully establishes vein patency and resolution of symptoms in 95–100% of cases.[2] The complications of a thoracic outlet decompression surgery include pneumothorax, hemothorax, chylothorax, injury to the subclavian neurovasculature or thoracic duct, or failure to fully decompress the thoracic outlet.[4,7]

The surgery is often sequentially combined with catheter-directed thrombolysis, balloon angioplasty, or oral anticoagulation to obtain rapid vessel patency and remove the mechanical obstruction.[1,7,8,12] Interventions undertaken earlier are more effective than delayed treatments. One study showed thrombolysis within 7 d of symptom onset followed by surgery within 30 d had significantly reduced recurrent thrombosis and improved symptoms long term.[1] A retrospective study of 312 episodes of effort-associated thrombosis

showed that if the patient was not treated within 6 wk of the onset of symptoms, most patients remained symptomatic even after decompression surgery.[12]

With any treatment listed, there can be residual venous defects from the chronic damage that may require vascular repair.[8]

5. What is the most likely aeromedical disposition for this pilot?

A. Permanently disqualified for flight duties.
B. Return to flying status with waivers.
C. Grounded during anticoagulation use with 12-mo observation period to ensure no repeat DVTs and then return to flying status without restrictions.

ANSWER/DISCUSSION

5. B. PEs and acute vascular occlusions in VTOS can compromise flight safety because they can cause acute incapacitation. Additionally, the pain and swelling that occur with VTOS can endanger the aircrew if they degrade an airman's ability to effectively perform during a critical phase of flight. Therefore, both conditions are disqualifying for military aviators. VTOS itself is not listed as a specific disqualification in any flying physical standards. The U.S. Air Force Medical Standards Directory lists repeated deep venous thrombosis and pulmonary embolisms as disqualifying for all flying classes.* Oral anticoagulation medications are not authorized for flight duties in the U.S. Air Force without a waiver.† According to the U.S. Air Force Waiver Guide, a service member must be treated for at least 3 mo (or longer as indicated by clinical guidelines) with anticoagulation prior to waiver consideration.[9] Furthermore, if the airman requires continued, prolonged anticoagulation, the airman cannot fly high-performance or single-pilot aircraft. Airmen also require a thrombophilia evaluation.

If this pilot were in a different military service, the flying requirements would be slightly different. The U.S. Navy and U.S. Army have more detailed waiver requirements to confirm resolution of any thrombus and rule out inheritable thrombophilias.[10,13,14] Specifically, the U.S. Navy will not waive recurrent episodes of DVT/PE[16] and the U.S. Army requires case-by-case evaluation for any recurrent thrombosis or those that require surgical treatment.[14] Detailed service-specific criteria can be found in that service's waiver guide.

According the Federal Aviation Administration Guide for Aviation Medical Examiners, civilian airmen with thromboembolic events, even if still on oral anticoagulation, can be considered for certification after case review by the Federal Air Surgeon.[5] The required documentation for consideration of a special issuance includes hospitalization records, family history of thromboembolic disease, neoplastic workup if indicated, thrombophilia evaluation, and international normalized ratio if currently taking warfarin. An Aviation Medical Examiner can reissue an airman medical certification if the airman meets certain guidelines and the necessary interval history is submitted.

This patient is placed on oral anticoagulation and undergoes surgical resection of his first left rib and scalenectomy. He subsequently discontinues anticoagulation and is rehabilitated with physical therapy to recover the majority of the strength in his left arm. He has minimal residual left shoulder pain that does not affect his performance as a pilot. He is granted an indefinite waiver without limitations. Should he have a recurrence, he will be grounded and require a new waiver submission.

This case highlights the uncommon but important condition of VTOS. Having an increased index of suspicion for patients with recurrent shoulder pain, upper extremity paresthesias, weakness, or swelling could shorten the time to correct diagnosis and reduce unnecessary workup and treatments.

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REFERENCES

1. Bamford RF, Holt PE, Hinchliffe RJ, Thompson MM, Loftus IM. Modernizing the treatment of venous thoracic outlet syndrome. Vascular. 2012; 20(3):138–144.
2. Bailey CJ, Illig KA. Contemporary management of axillosubclavian vein thrombosis. Interv Cardiol (Lond). 2013; 5(4):453–463.
3. Chin EE, Zimmerman PT, Grant EG. Sonographic evaluation of upper extremity deep venous thrombosis. J Ultrasound Med. 2005; 24(6): 829–38.
4. Colli BO, Carlotti CG, Jr. Thoracic outlet syndrome: II. Diagnosis and treatment. Contemp Neurosurg. 2001; 23(10):1–5.
5. Federal Aviation Administration. Protocol for thromboembolic disease. In: Guide for aviation medical examiners. Washington (DC): Federal Aviation Administration; 2015:260. [Accessed on 2 Dec. 2015]. Available from http://www.faa.gov/about/office_org/headquarters_offices/avs/offices/aam/ame/guide/media/guide.pdf.
6. Gelfand EV, Piazza G, Goldhaber SZ. Venous thromboembolism guidebook. Crit Pathw Cardiol. 2002; 1(1):26–43.
7. Kuhn JE, Lebus VGF, Bible JE. Thoracic outlet syndrome. J Am Acad Orthop Surg. 2015; 23(4):222–232.
8. Moore R, Wei Lum Y. Venous thoracic outlet syndrome. Vasc Med. 2015; 20(2):182–189.
9. Mulagha EH, Van Syoc D. Deep venous thrombosis/pulmonary embolism (May 14). In: Air Force waiver guide. Wright-Patterson AFB. OH: U.S. Air Force School of Aerospace Medicine; 2015: 277–283. ([Accessed 2 Nov. 2015], Available from http://www.wpafb.af.mil/afrl/711hpw/usafsam.asp.
10. Naval Aerospace Medical Institute. 3.23 Thrombophilia/venous thrombosis/pulmonary embolism. In: U.S. Navy aeromedical reference and waiver guide. Pensacola (FL): Naval Aerospace Medical Institute; 2015.

* U.S. Air Force. Section H: heart and vascular disqualifying conditions, H49 & H50. In: Medical standards directory. 2013:27. [Accessed 2 Nov. 2015]. Available from https://kx2.afms.mil/kj/kx4/FlightMedicine/Documents/Medical%20Standards%20Directory%20(MSD)/MSD%202013-Dec-2.pdf to those with access.

† U.S. Air Force. Official Air Force aerospace medicine approved medications. [Accessed 2 Nov. 2015]. Available from https://kx2.afms.mil/kj/kx4/FlightMedicine/Pages/standards.aspx to those with access.

[Accessed 2 Nov. 2015]. Available from http://www.med.navy.mil/sites/nmotc/nami/arwg/Pages/AeromedicalReferenceandWaiverGuide.aspx.
11. Thompson JF, Winterborn RJ, Bays S, White H, Kinsella DC, Watkinson AF. Venous thoracic outlet compression and the Paget-Schroetter syndrome: a review and recommendations for management. Cardiovasc Intervent Radiol. 2011; 34(5):903–910.
12. Tsekouras N, Comerota AJ. Current trends in the treatment of venous thoracic outlet syndrome: a comprehensive review. Interv Cardiol (Lond). 2014; 6(1):103–15.
13. U.S. Army. Standards of medical fitness. Washington (DC): Department of the Army; 2011. Army Regulation 40-501. Retrieved on 2 Nov 2015 from https://kx2.afms.mil/kj/kx4/FlightMedicine/Pages/armyhome.aspx. Available to those with access.
14. U.S. Army Aeromedical Activity. Venous thrombosis (ICD9 453.8/415.1). In: Flight surgeon's aeromedical checklists. Ft. Rucker (AL): U.S. Army Aeromedical Activity; 2014. [Accessed 2 Nov. 2015]. Available from http://www.rucker.amedd.army.mil/assets/documents/pdf/Army_APLs_28may2014.pdf.