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Case Report
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Superior Vena Cava Obstruction with Thrombolytic Therapy in Primary Antiphospholipid Syndrome

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Abstract

We report a case of superior vena cava obstruction of a 39 years old female patient with primary antiphospholipid syndrome presented with upper limb pain and swelling, which improved on thrombolytic therapy, we review patients with Antiphospholipid syndrome and management with thrombolytic therapy.

Keywords: Superior Vena Cava (SVC); Antiphospholipid Syndrome (APS); MVC

Background

Superior vena cava syndrome results from the obstruction of blood flow through the superior vena cava and is most often due to thoracic malignancy. However, benign etiologies are on the rise secondary to more frequent use of intravascular devices such as central venous catheters and pacemakers. There also are other rare non neoplastic diseases that can cause SVCS, for example, fibrosing mediastinitis [1], which is a rare chronic inflammatory condition similar to retroperitoneal fibrosis, use for indwelling catheter and hypercoagulable condition as primary antiphosplpid syndrome which is very rare condition.

Case Description

A 39 years old female presented with fever and shortness of breath for 14 days for which Total leucocytic shift 16 with shift to the left, hemoglobin 9.5 MCV 69, CRP 161, chest X ray was free on admission, ECG showed normal sinus rhythm, Echocardiography showed from right atrial mass 2.7 cm × 1.9 cm, moderate mitral and tricuspid regurgitation second day after admission she started to suffer from pain and swelling of both upper limb. Duplex upper limbs revealed bilateral total occlusion of subclavian, axillary and brachial veins. Proximal part of right IJV was totally occluded and left side was partially occluded. By examination her blood pressure was 120/60, regular heart rate and rhythm and oxygen saturation 97% her neck showed visible dilated non pulsating veins. Cardiovascular exam showed pansystolic murmur over apex with no thrill and another at lower part of sternum increases with inspiration. Lung auscultation showed diminished air entry at the right base (Figure 1).

During hospital stay lupus anticoagulant and anticardiolipin IgM were positive, workup against infective endocarditis was done to exclude secondary infection of right atrial mass and it was negative, tumor markers were negative, CT chest revealed right sided pleural effusion, left lower lobe consolidation ,enlarged pretracheal, supracarinal and aortocaval lymph nodes. TEE and CMR revealed right atrial mass 2.7 cm × 2.9 cm × 1.7 cm most likely thrombus and another mass in right atrial appendage 1.0 cm × 0.7 cm × 0.7 cm, moderate mitral and tricuspid regurgitation, pulmonary hypertension ,patent non occluding thrombi within SVC (Figures 2-4).

Patient underwent venous angiography for which catheter was inserted through venous access to right cephalic vein and 1.5 million IU of streptokinase was given and then she was maintained on heparin infusion with adjustment to a PTT 50-70, venography was done and revealing no improvement so a decision was given to give second dose streptokinase on fifth day she was given second bolus 250,000 and maintained on 100,000 IU/h for 10 h stopped due to fever (drug reaction), patient underwent venous angiography follow up which revealed marked improvement in venous blood flow. She was discharged home safely on aspirin 75 mg once and warfarin with proper adjustment to target INR (Figure 5).

Outcome

She had smooth course with improvement to her symptoms and upper limb edema and was

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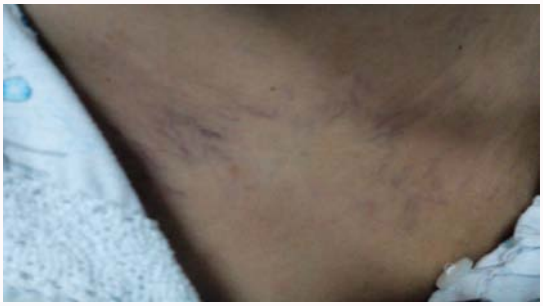

Figure 1: Dilated visible veins on patient neck.

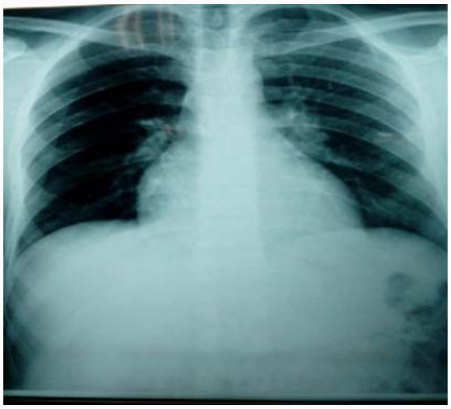

Figure 2: Chest X-ray.

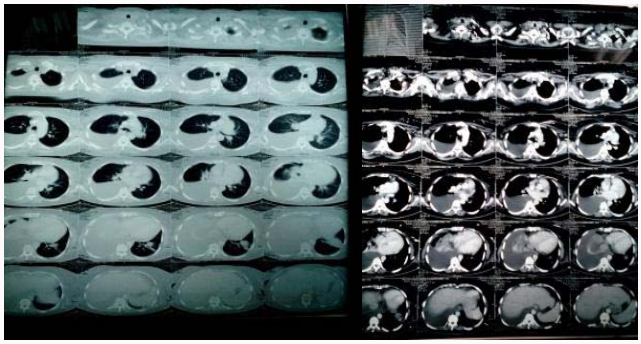

Figure 3: CT chest of female patient revealed enlarged pretracheal and supracainal and aortocaval lymph node and right sided pleural effusion.

discharged home at stable condition on regular follow up to her anticoagulation.

Discussion

Benign Superior vena cava syndrome accounts for less than 10%. Half of them due to fibrosing mediastinitis and use of indwelling catheters which increase incidence of thrombosis while other causes are rare.

Antiphospholilpid syndrome is a thrombophilic syndrome in which venous or arterial thrombosis or both, may occur in patient with antiphospholipid antibodies positive, the diagnosis of definite antiphospholipid syndrome must include at least one clinical event of arterial or venous thrombosis and at least one of the laboratory criteria [lupus anticoagulant or anticardiolipin IgG or IgM antibodies, this was reported in three case at Iran Journal of Immunology [2], Spain [3] and Israel [4].

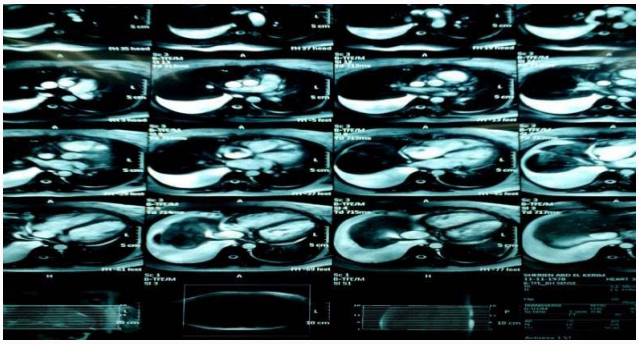

Figure 4: MRI heart showed right atrial mass.

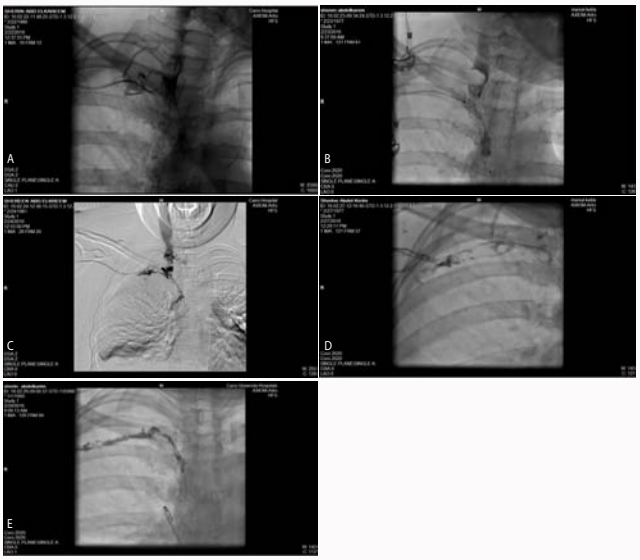

Figure 5: Showed the five days venous angiography were patients received thrombolytic therapy and was maintained on heparin infusion.

Traditionally, the presence of soft thrombus in an occlusion should require prompt attempts to relieve the clot burden before deciding to treat more invasively. The presence of a thrombus should be considered in patients with acute onset of symptoms, occlusions that can easily be traversed by a wire, and when SVCS are secondary to central venous catheters. In these patients, a trial of systemic anticoagulation or, in the acute setting, thrombolysis can be attempted by using thrombolytic agents delivered at the site of the clot through a catheter, which allows more effective local action. Reports of systematic administration of thrombolytic agents have also been described [5]. By reducing the thrombus load, the length of the obstruction can often be reduced, thus reducing the number of stents needed [6]. However, thrombolysis is most effective if it is started within 2 to 5 days of onset of symptoms and tends to be ineffective if started after 10 days [7]. Gray et al. [8] found that 88% of the cases had a successful outcome when thrombolytic treatment was commenced within 5 days. The success rate dropped to 25% when it was started after 5 days [8]. They also observed that catheter-related SVCS demonstrated a better response to thrombolysis, which was attributed to the earlier detection of symptoms secondary to catheter dysfunction, the absence of extrinsic compression, and the ability to infuse the thrombolytic agent directly to the site of the thrombus *via* the existing catheter [10]. Caution should be exercised in patients with cystic fibrosis because an increased risk of hemoptysis and

gastrointestinal bleeding has been reported in this patient population [5].

Newer techniques that use pharmacomechanical thrombolysis tools, such as the Trellis Thrombectomy System (Bacchus Vascular, Santa Clara, CA) and Trellis infusion system (Covidien, Santa Clara, CA), have shown promising results in reducing the thrombus load [9,10].

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