Introduction

Superior vena cava syndrome (SVCS) is obstruction of blood flow through the superior vena cava (SVC). It is a medical emergency which requires immediate diagnostic evaluation and therapy. Apart from carcinomas causing external compression, infections and indwelling central venous devices, it is rarely caused by thrombosis which may also be associated with pulmonary embolism. Venous thrombosis in the form of Deep Vein Thrombosis (DVT) affects about 2 in 1000 deliveries and this incidence increases to 3.24 per 1,000 deliveries in postpartum period but it usually involves veins of pelvis and legs. Superior vena cava thrombosis in pregnancy and accompanied by peripartum cardiomyopathy is extremely rare condition and only a few cases have been reported.

We describe a case of superior vena cava thrombosis with peripartum dilated cardiomyopathy in a patient with history of caesarean section few days back and a normal thrombophilia screen.

Case Report

A 30 years multigravida female with body mass index (BMI) of 23 kg/m\(^2\), underwent emergency caesarean section due to transverse lie in labor 10 days back in a local hospital. She was referred to our hospital with cough, severe breathlessness at rest, orthopnea with pain in neck and arms. Clinical examination revealed signs of heart failure. Echocardiography showed ejection fraction of 15%, with no right ventricular strain. A diagnosis of peripartum cardiomyopathy was made. Doppler ultrasound of neck veins showed bilateral internal jugular vein thrombosis. Subsequent multislice CT examination showed thrombosis of superior vena cava and both internal jugular veins (with collateral formation) and pulmonary embolism. There were no mediastinal abnormalities on the CT scan. Her thrombophilia screen and CT scan brain was normal. She was managed in collaboration with cardiologist. Following treatment with subcutaneous enoxaparin therapy and warfarin her symptoms of upper limb pain improved. She responded very well to medical therapy for heart failure with marked improvement of NYHA functional class.

were raised at 24 mg/L. Coagulation profile was normal with INR of 1.00. Thrombophilia screen including protein-C, protein S, anti-thrombin III deficiency, factor V leiden, anticardiolipin antibodies, antinuclear antibodies and lupus anticoagulant all came out to be normal. Brain natriuretic peptide levels in blood were raised at 1989 pg/mL.

ECG showed sinus tachycardia with T-wave inversion from lead V1-V3. Her chest X-ray revealed cardiomegaly with pulmonary oedema and pleural effusion on the right side. Echocardiography showed a dilated left ventricle with an ejection fraction of 15% with global hypokinesia. There was no evidence of right ventricular dysfunction or significant rise in pulmonary artery pressures; measured at 26 mmHg. Doppler ultrasound of neck veins revealed thrombosis of caudal half of both internal jugular veins.

Subclavian and axillary veins were patent. Doppler ultrasound of both the pelvic and leg veins was normal. Cardiac multislice CT (MSCT) revealed bilateral internal jugular vein thrombosis with the thrombus in right internal jugular vein extending into brachiocephalic vein and proximal superior vena cava (Figure 1 and 2). Pulmonary embolism and infart was noted in right mid and lower zone. As a part of her heart failure evaluation, right heart catheterization studies were done through right femoral vein approach. The occlusion of the SVC was confirmed by injection of contrast into the SVC. CT scan brain showed a normal study.

She was managed in collaboration with cardiologist and initially kept on subcutaneous enoxaparin with warfarin overlap and shifted to warfarin only when INR reached 3.0. Her heart failure was managed on optimal medical therapy including a combination of diuretics, ACE inhibitors and beta blockers. Her baseline 1 year mortality on the seattle heart failure scoring system was 35%.

Within the first week of treatment, the patient's heart failure improved dramatically as did the pain in her arms. She became mobile and active with improvement of NYHA to class II. She was discharged on warfarin as anticoagulant and medical therapy for her heart failure. There was no further pain in her upper limbs or neck. There were no neurological symptoms.

On follow-up after 15 days, she was clinically assessed and her baseline biochemistry was repeated. Her INR was done which came out to be 2.5. Repeat uric acid was 6.8. There was no further pain in her upper limbs or neck. There were no neurological symptoms. On 30 days follow-up, her clinical status was stable and her biochemistry including uric acid was now normal. Her BNP after one month of optimal medical therapy showed more than 50% reduction with a value of 790 pg/ml. Seattle heart failure scoring system based on her clinical condition and biochemistry showed a marked reduction in 1 year mortality from 35% to just 18%. Her cardio-pulmonary exercise test (VO2 max) was done to grade the severity of failure and the functional capacity of the patient and it came out to be 10 ml/min/kg at anaerobic threshold of 1.3. This is a significantly low value and is likely to improve further with continued medical treatment.

DISCUSSION

Superior vena cava syndrome (SVCS) is an unusual condition with external compression due to malignancies as the most common cause. SVC thrombosis also occurs secondary to indwelling central venous devices in 25% of cases as well as in different hypercoagulable states.2 A study conducted in a hospital in Turkey to determine the prevalence thrombosis of brachio-
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cephalic veins and SVC in the hospital setting, showed thrombosis of the brachiocephalic veins and SVC to be 0.03% over a period of 3 years.

Although obese women older than 35, who have a family or personal history of thromboembolism are at higher risk and the risk of developing deep vein thrombosis or pulmonary embolism rises by 5 times after delivery especially after caesarean section.4 But this is only true for deep vein thrombosis affecting pelvic veins and veins of lower extremities. Little has been written about superior vena cava thrombosis in pregnancy associated with peripartum cardiomyopathy; with only few case reports with either Behcets disease or after in vitro fertilization techniques.5

This patient was not obese with BMI of 23 kg/m2 and she had no personal or family history of thromboembolism. She had conceived spontaneously and there was no past history of use of oral contraceptive pills. Although she had an emergency caesarean section which is a well known risk factor for thromboembolism in postpartum period but the Doppler ultrasound of lower extremities as well as pelvic veins was absolutely normal.

The obstruction of superior vena cava leads to increased venous pressure in the upper body resulting in oedema of the head, neck, and arms. Most of studies showed swelling of the arm, head, and neck as major symptom present in 97% of patients with cough, oedema and swelling which is visually striking but generally of little clinical consequence. This oedema can cause a functional compromise of the larynx or pharynx, causing dyspnoea, stridor, cough, hoarseness, and dysphagia. Cerebral oedema may lead to cerebral ischaemia, haemorrhage, confusion, coma, and possibly death.6

In this patient, the major symptoms affecting the patient were due to heart failure whereas the SVC obstruction led to just painful neck and arms without swelling. The patient was also found to have pulmonary embolism. The association of clinically recognized PE with SVC thrombosis is limited to case reports and is life threatening. Screening for coagulation disorders like for protein C, protein S, and antithrombin-III deficiencies factor V Leiden, the prothrombin gene mutation, hyper homocysteinemia, and antiphospholipid antibodies is recommended for idiopathic SVC thrombosis with a family history of deep vein thrombosis (DVT), a history of recurrent, unexplained pregnancy loss, or a personal history of a prior DVT.7

Duplex ultrasound is the initial imaging test of choice for diagnosing SVC obstruction because this technique is noninvasive and has high sensitivity and specificity. Venography provides excellent characterization of venous anatomy but has several drawbacks. There may be technical difficulty in cannulating the vein in an edematous arm. The test requires an iodinated contrast agent, which may cause an allergic reaction, nephrotoxicity, or a chemical phlebitis that can worsen the pre-existing thrombosis.8 Multi-detector row CT (MDCT) with multiplanar and 3D imaging is an effective tool in evaluation of the SVCS and has a greater advantage than the other imaging techniques. 3D volume rendering is a useful technique in determining and describing collateral circulations in addition to the primary disease process.8

Magnetic resonance angiography (MRA) is an accurate, noninvasive method for detecting thrombus in the central thoracic veins, such as the SVC and brachiocephalic veins. MRA correlates extremely well with venography and provides more complete evaluation of central collaterals. But it is expensive and not universally available.9

Anticoagulation is the cornerstone of therapy. Anticoagulation helps maintain patency of venous collaterals and reduces thrombus progression with organization of the thrombus and possible formation of micro-channels through the organized thrombus. Typically, overlapping unfractionated heparin is used as a “bridge” during introduction of warfarin, which if introduced in isolation can actually increase thrombosis with skin necrosis. Most studies recommend warfarin to be continued for a minimum of 6 months, with a goal INR of 2.0 - 3.0. Thrombolysis/thrombectomy and surgical decompression are often successful, but less frequently used.10

REFERENCES


