

Anasarca sparing one limb

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Abstract

Upper extremity deep vein thrombosis commonly refers to thrombosis of the axillary and/or subclavian veins. This condition may lead to some serious complications like pulmonary embolism if the diagnosis and treatment are delayed. We introduce here one such case where the diagnosis of this rare condition was difficult because of the atypical clinical manifestation but the chylous ascites provided a valuable clue which led to early diagnosis and treatment of this patient.

Keywords: Chylous ascites, Paget-Schroetter syndrome, upper extremity deep vein thrombosis

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Introduction

Upper extremity deep vein thrombosis (UEDVT) most commonly refers to thrombosis of the axillary and/or subclavian veins. It is a serious medical emergency which if left untreated may lead to severe complications like pulmonary embolism and superior vena cava (SVC) syndrome. In this report, we highlight the difficulty in diagnosing this serious condition when it presents atypically.^[1,2]

Case Report

A 24-year lady presented to medical accident and emergency with chief complaints of swelling of body for 1 month. She had associated breathlessness and decreased urinary output for 7 days. There was past history of pulmonary tuberculosis around 10 years back for which she took complete treatment and declared cure. On general examination, pulse was 106/min feeble, respiratory rate was 30/min, BP was 100/60 mmHg, and neck veins were full. She had edema involving whole of the lower extremities. Left upper extremity was disproportionately swollen as compared to right

upper limb which was relatively normal [Figure 1]. Crepitations were heard on her right infraclavicular area of chest which fixed and static throughout course of disease course a sequelae to old healed tuberculosis. A most probable diagnosis of anasarca due to chronic renal failure was considered. Blood gas analysis revealed a pH of 7.35, PaO₂ 80%, PaCO₂ 67%, and bicarbonate 21.4 mmol/L. Kidney function tests, liver function tests, serum proteins, albumin, Na, K, Ca, phosphate, urine routine, and microscopy were done, but none was conclusive for the cause of edema. An ultrasound abdomen done to evaluate the size of kidney revealed minimal free fluid. A cremish white fluid was aspirated. This presentation of anasarca sparing right upper limb with milky white ascites suggested obstruction of long thoracic duct. On re-evaluation of the patient few dilated veins were seen in left supraclavicular region. There was no history of any missed abortions, coagulopathies, venous thrombosis, or pulmonary embolism in patients as well as in other family members. There was no history of any trauma to the extremity, intravenous catheterization, or needle insertion for any purpose in past few months. In view of distribution of edema, dilated veins in neck and cremish white ascites, a clinical diagnosis of UEDVT involving the junction of jugular vein and subclavian vein was thought. Upper extremity Doppler ultrasound confirmed so revealing a thrombus in the left subclavian vein extending up to the proximal part of left internal jugular vein. The CT scan of the

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chest further confirmed the presence of thrombus in deep vein of upper extremity and ruled out any other cause of compression of a long thoracic duct. A note of few lymph nodes adjacent to common carotid and left subclavian artery along with bronchiectatic changes in both the lungs. The CT scan of the abdomen showed the presence of ascites. Ascitic fluid analysis confirmed it to be chylous ascites. Her prothrombin time, INR, and aPTT were normal. D-dimer was elevated. Rheumatoid factor and anti-nuclear antibody were negative, while CRP (C-reactive protein) was raised in patient. The patient was started on anticoagulation therapy with heparin followed by warfarin. Her condition started improving in the next 2 days and her pain and swelling of the left upper limb gradually decreased and also the swelling of both the lower extremity.

Discussion

This was a case UEDVT leading to obstruction of the long thoracic duct which in turn led to obstruction of lymph and anasarca involving three extremity and chylous ascites. As would have been expected in obstruction of the long thoracic duct at the junction of internal jugular and subclavian vein was easily missed but the presence of ascites gave important clue to reach to the diagnosis. Application of important known principles of lymphatic drainage of body [Figure 2] leads to the diagnosis.

UEDVT is a condition characterized by thrombosis of deep veins of upper extremity. UEDVT is classified as primary or secondary depending on the predisposing risk factors like use of central venous catheter. Primary UEDVT is a rare disorder (2 per 1,00,000 persons per year)^[3] that refers either to effort thrombosis or idiopathic UEDVT. The term Paget-Schroetter syndrome was first used by Hughes in 1949 to recognize the cases of upper

extremity thrombosis secondary to effort reported by Paget in 1858 and Schroetter in 1899.^[4] Patients with Paget-Schroetter syndrome develop spontaneous UEDVT, usually in their dominant arm, after strenuous activity such as weight lifting, rowing, and wrestling. It has been suggested that the heavy exertion causes microtrauma to the vessel intima and leads to activation of the coagulation cascade.^[4] In contrast to Paget-Schroetter syndrome, patients with idiopathic UEDVT have no known trigger or obvious underlying disease. However, idiopathic UEDVT may be associated with occult cancer, factor V Leiden mutation, protein C deficiency, protein S deficiency, antithrombin deficiency, and prothrombin 20210A mutation.^[5] UEDVT (secondary UEDVT) may be the complication of subclavian vein catheterization for administration of cancer chemotherapy, total parenteral nutrition, and prolonged administration of antibiotics therapy. The present case did not have any obvious predisposing cause of thrombosis. The index case had chylous ascites. Liang *et al* reviewed 22 cases of chylous ascites and they observed that main causes were malignant tumor (6/22), hepatic cirrhosis (5/22), tubercular (4/22), and rarely traumatic and congenital lesion. None of these had UEDVT as a cause for chylous ascites.^[6] Although our patient had past history of pulmonary tuberculosis for which she took treatment, complete treatment was declared cure then. At present she did not had any feature of active tuberculosis on both examination and investigations. Blalock, *et al* observed in their experiments on animals concluded



Figure 1: Photograph of the patient

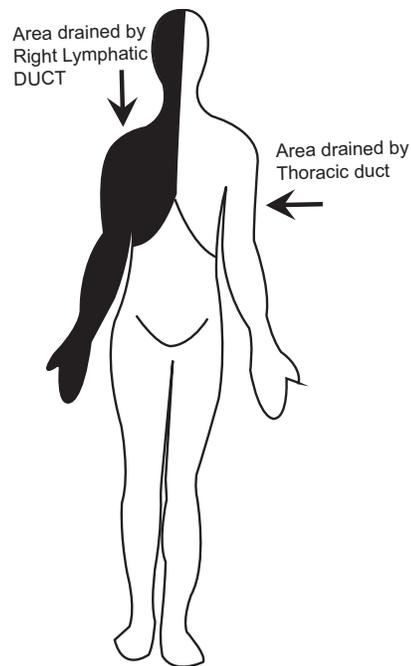


Figure 2: Lymphatic drainage of body

that the tying of the thoracic duct did not lead to chylous ascites.^[7] However, contrary to their, Baram *et al* presented a case of chylothorax and chylous ascites secondary to idiopathic subclavian vein thrombosis. Two years later, the same patient was diagnosed to have advanced hepatocellular carcinoma.^[8] The knowledge of lymphatic drainage of body made the diagnosis possible. Left upper and both lower extremities drain in the long thoracic duct while right upper extremity drain into the right lymphatic duct which ultimately drain at the junction of jugular and subclavian vein on their respective side.^[9] This report highlights the difficulty of diagnosing subclavian vein thrombosis in patients having anasarca involving three limbs, i.e. left upper and both the lower extremities. The swelling resulting from thrombosis in the deep vein of upper extremity may easily be misinterpreted as generalized edema and the treatment could thus be delayed leading to serious complication. The knowledge of such a presentation is very important for those looking after critically care unit especially when UEDVT is on rise. To our knowledge there are multiple manuscripts of incidence, risk factors, incidence, case reports of UEDVT, but none have described such a presentation of UEDVT having anasarca sparing one limb with chylous ascites.

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