INTRODUCTION

Upper-extremity deep vein thrombosis (UEDVT) commonly refers to thrombosis of the axillary and/or subclavian veins. It is classified as primary or secondary. Primary UEDVT is a rare disorder (2 per 1,00,000 persons per year). It refers to effort thrombosis or Paget-schroetter syndrome or idiopathic UEDVT. It usually occurs in young healthy persons. Secondary UEDVT develops in older persons with medical co-morbidities, malignancies, central venous catheters etc. and accounts for most cases of UEDVT.

CASE REPORT

A 76 years male right handed, diabetic and hypertensive since 20 years on regular treatment, presented with swelling all over left upper limb up to the neck since 15 days. There was no history of pain, fever or trauma. There was no history of similar swelling in the past. He was operated for inguinal hernia 20 years back.

The patient gave history of lifting heavy baggage on and off in the last 15 days during flight to and return from USA. On examination, his pulse rate was 84/min. All peripheral pulses were equally well felt; his blood pressure was 140/80 mmHg. He was afebrile. On local examination there was swelling over left upper limb, neck, axilla, infra-clavicular area. There was mild discoloration but no tenderness over left upper limb or neck; few distended veins were seen over left infraclavicular region. Rest of the systemic examination was within normal limits. On fundus examination both eyes showed grade II diabetic retinopathy.

Investigations revealed Hb of 13.8 g%, TLC -13500/ cmm, DLC – N72 L24 M2 E2, Platelet -3.38 lacs/ hpf. Urine routine and microscopy was normal. Renal and liver function test, serum electrolytes were within normal limits.

Ultrasoundography abdomen and pelvis showed a nonobstructive 13 mm calculus in lower pole of left kidney along with mild enlargement of prostate. 2D Echo showed early diastolic dysfunction with ejection fraction of 60%. Peripheral Doppler of left upper limb was suggestive of acute hypoechoic intraluminal thrombus in left subclavian vein in entire length and axillary vein with absent flow (Fig. 1). Veins were non compressible. Brachial, radial, ulnar, cephalic vein were patent. There was diffuse subcutaneous soft tissue edema. Chronic thrombus was seen in proximal segment of left internal jugular vein. Distal internal jugular vein was patent. Doppler of other limbs was normal. CT Thorax and Abdomen showed non opacification of left subclavian and innominate veins after intravenous contrast with evidence of intraluminal thrombus (Fig. 2). Nonopacification of left internal jugular vein was evident. Right innominate vein and superior vena cava was normal. Lungs were normal. Left renal calculus of 11 mm was seen. Rest of CT was normal. Pulmonary angiography showed small focal filling defect along lateral wall of aortic arch with adjacent calcific plaque

Abstract

A case of primary upper-extremity deep vein thrombosis (UEDVT) or Paget-Schroetter syndrome is reported. It is effort thrombosis usually affecting young healthy individuals and is a rare condition. Our patient was an elderly male with history of exertion using the left upper limb. He responded to limb elevation and anticoagulation. No other cause for thrombosis was found.

Fig. 1: Peripheral Doppler of left upper limb showing acute hypoechoic intraluminal thrombus in left subclavian vein.
measuring 15.8 x 5.7 mm. Rest of the pulmonary angiogram was normal.

To conclude patient had thrombosis of left axillary, left subclavian and left IJV, left innominate vein till its confluence into SVC. He was treated with low molecular weight heparin for 5 days, and then was put on Warfarin for 3 months with INR monitoring. Target INR was 1.5-2.0. Our patient’s INR was 1.6-2.0.

Further investigations were done to exclude other causes of venous thrombosis including coagulation profile, antithrombin III, protein C, protein S, homocysteine levels and APLA, which were normal. Factor V Leiden levels could not be done. D-dimer levels were raised to 2000 ng/L. Prostate specific antigen was 11 ng/mL. In the elderly thrombosis of the deep veins of the upper limb can be a precursor to a malignancy, so we investigated him thoroughly for the same. However, none of the investigations revealed anything. Patient later underwent prostatic biopsy also which was normal.

Diagnosis of Paget-Schroetter syndrome of effort related UEDVT was made. After 3 months repeat Doppler of left upper limb showed near complete recanalization of the veins. There was no swelling. He is still under regular follow up.

**DISCUSSION**

Paget-Schroetter syndrome is the spontaneous, primary UEDVT described by von-Schroetter in 1884 in Vienna and by Paget in 1875 in London. Hughes, in a review of 320 patients in 1948, originated the eponym “Paget-Schroetter syndrome”. It is either truly idiopathic or is related to physical activity or arm positioning with or without anatomic compression at the thoracic outlet. It is also called effort syndrome. It usually affects young healthy adult men. While the dominant arm is usually involved, one-third of the patients may have the non-dominant side affected.

Spontaneous thrombosis in the arms has been reported in athletes such as golfers, football, tennis and baseball players, weightlifters and cheerleaders; as well as painters and beauticians. In a subgroup of patients however no underlying cause is found. Some of these patients may have an occult malignancy, which typically remains elusive until sometime after the thrombotic manifestation. The most commonly associated malignancies are lymphoma and lung cancer.

Though occasionally, patients may be asymptomatic, initial complaints usually include heaviness in the affected arm, mild dull ache and pain in the neck, shoulder and/or axilla of the involved limb. More dramatic signs may include ecchymosis and swelling, discoloration and mottled skin, distention of the cutaneous veins of the involved extremity.

The subclavian vein may be compressed during lateral abduction of the arm, causing turbulence or obstruction to the flow. Microscopic intimal injury may occur stimulating the coagulation cascade. Repetitive motion may also contribute to anatomical narrowing of the thoracic outlet. Lifting of heavy weights may cause further narrowing of the costocalvicular space through depression of the shoulder.

Traditionally treatment has consisted of limb elevation and anticoagulation. More recently, catheter directed thrombolytic therapy followed by surgical decompression of the thoracic outlet has been advocated. Preliminary studies suggest that ultrasound may accelerate thrombolysis by enhancing enzymatic fibrinolysis and mechanically disrupting the thrombus.

The majority of patients improve overtime. However, a significant proportion of patients with either primary or secondary UEDVT suffer from a post-thrombotic syndrome due to chronic venous insufficiency.

There are some special features in our case. He was an elderly man of 76 years. So far, we have some across only one case report in an elderly male aged 53 years. Initially we could not find any predisposing cause of the thrombosis. Then on questioning he mentioned carrying a shoulder bag on his left shoulder and intermittent lifting of a heavy suitcase on and off during the recent air travel. There also appeared to be a link between air travel and venous thrombosis; but the link is likely to be weak as it mainly affects passengers with additional risk factors for venous thrombosis and mostly calf veins.

The patient continues to follow up with us. We are aware that considering his age an occult malignancy which may be elusive now can present at a later date.

We have presented this case to raise the awareness of the rare condition of Paget-Schroetter syndrome. Through there are reports in literature, an individual
physician’s experience with this condition is usually limited, so the diagnosis requires a high index of suspicion.

REFERENCES

API Announcement

Award Sessions
1. Dr. DP Basu Young Award in Cardiology.
2. E Merck Award.
3. Dr. JN Berry Memorial Award and
4. Dr. MJ Shah Memorial Award in Tropical Medicine

There will be four award sessions at the 2008 Annual Conference of API at Kochi. The rules and regulations of these awards are as under.

1. Papers that are accepted for presentation in the Award Session at the Annual Conference will be divided subject - wise into four groups.
   Group I   Cardiology   DP Basu Young Award
   Group II  Chest Diseases  E Merck Award
   Group III Other Specialties  JN Berry Memorial Award
   Group IV  Tropical Medicine  MJ Shah Memorial Award

   The Award of Dr. JN Berry Memorial Award and E Merck Award are given in alternate years in Group II and III papers. At the 2008 Annual Conference at Kochi, Dr. JN Berry Memorial Award will be for "Other Specialties" and E Merck Award for "Chest Diseases". Dr. DP Basu Young Award will be for "Cardiology" and Dr. MJ Shah Memorial Award for "Tropical Medicine".

2. The competitor must be the first author of the paper submitted for presentation at the API sessions of the Annual Conference. A testimonial must be submitted from the head of the institution that the major work has been done by the competitor. Papers which are previously presented or published will not be considered. The competitor should also give a written pledge stating that the work has not been presented or published before. He should be a member of API.

3. Dr. JN Berry Memorial and DP Basu Young Awards are worth Rs. 1000/- each. E Merck Award Rs. 2000/- and Dr. MJ Shah Memorial Award is worth Rs. 2500/-

4. The upper age limit of the competitor is 40 years.

5. The decision will be taken by a panel of judges appointed by the Governing Body of API.

6. The candidate must apply for the award and full manuscript of the paper will have to be submitted. The paper will be presented in separate award session.

7. Eight copies of full manuscript will have to be submitted to Dr. SK Bichile, President - Elect and Chairman Scientific Committee, APICON 2008, "107, Modi Chambers, 1st Floor, French Bridge Corner, Opp. Opera House, Mumbai 400 004; e-mail : skbichile@vsnl.net, medicaloncology@mtnl.net.in; mobile : 09324556411 by 31st July, 2007. One copy of the paper should be sent to Dr. Sandhya Kamath, Hon. General Secretary of API Unit No. 6 and 7, Turf Estate, Opp. Shakti Mill Compound, Off. Dr. E Moses Road, Near Mahalaxmi Station West, Mumbai 400 011. Tel. 022-6666 3224, 2491 2218; Fax : 2492 0263.

8. The decision of the panel of judges will be final and binding to all concerned.

Prestigious Awards of API
2. Distinguished Member (2007)

Suggestions should be accompanied with 8 copies of brief bio-data of the suggested name, so as to reach the API office not later than 31st July2007

Dr. Sandhya Kamath
Hon. General Secretary