RESEARCH PAPER | Medical Science | Volume : 4 | Issue : 9 | September 2014 | ISSN - 2249-555X

A Case of DVT Due to Protein C and Protein S Deficency - A Case Report

KEYWORDS: deep vein thrombosis, protein c and protein s,

Dr. P.S. Usha Rani M.D.	Dr. M.Abdur Rahim M.D.	Dr. G. Jameel Ahmed
Assistant Professor of Medicine, Kurnool Medical college,Government General Hospital Kurnool.	Associate Professor of Medicine, Kurnool Medical college,Government General Hospital Kurnool.	Resident in General Medicine, Kurnool Medical College, Kurnool. Government General Hospital Kurnool.

Dr.P.Prashanthi	Dr. K.Newton Issac	Dr.Sripada. Muvvagopal
Resident in General Medicine, Kurnool Medical College, Kurnool. Government General Hospital Kurnool.	Resident in General Medicine, Kurnool Medical College, Kurnool. Government General Hospital Kurnool.	Professor & HOD of General Medicine, Kurnool Medical College, Kurnool. Government General Hospital Kurnool.

ABSTRACT *The primary form of venous thrombosis is deep vein thrombosis. It usually involves the extremities and has risk of embolization. the causes of dvt are both inherited and acquired. among these the deficiency of naturally occurring vitamin k dependant anticoagulants namely protein c and protein s deficiency is very rare ,which can present as deep vein thrombosis.,*

we report a case of deep vein thrombosis in a 16 year old make with involvement of right external iliac to popliteal veins who on subsequent evaluation was found to have deficient levels of protein c and s. patient was started on long term oral anticoagulation therapy and is on regular follow up without new episodes.

INTRODUCTION:

Coagulation is the process by which thrombin is activated and soluble plasma fibrinogen is converted into insoluble fibrin.is a condition in which there is an increased tendency to form blood clots. There is a balance between the natural anticoagulants and clotting mechanisms many factors can disrupt this balance .The natural anticoagulants are required to help stop the clotting process, deficiencies of one of these substances can upset this balance and lead to thrombophilia. It may be hereditary or acquired(surgery, cancer, pregnancy, or medications. The two most common hereditary thrombophilia conditions are the factor V Leiden and prothrombin 20210 gene mutations. Inherited deficiencies of the natural coagulants[2] are uncommon. Protein C deficiency occurs in ≈1 of every 200 to 500 people, whereas protein S deficiency can be expected in ≈1 of every 500 individuals. Antithrombin deficiency is the least common of the 3 deficiencies, occurring in ≈1 of every 2000 to 5000 people. People with hereditary protein C or protein S deficiency have about a 2- to 11-fold increased risk for developing a DVT or PE in comparison with those without a deficiency.[3]

We report acase of dvt in a young male without any evident risk factors involving the right external iiac upto popliteal veins,who had reduced functional levels of protein c and s levels.the diagnosis was confirmed with colour doppler and was started on oral anticoagulant therapy and was discharged asymptomatic and put on regular follow up.

CASE REPORT:

A 15 year old male patient presented with complaints of swelling of right thigh from 7 days. There was history of pain abdomen and chest pain from 3 days. The rest of the examination was normal. There were no similar complaints in the past. His younger brother died at the age of 14, it was sudden death and cause of death not known. No other fraternal and maternal family members have similar illness. On physical examination there was swelling and tenderness of right lower limb from inguinal to popliteal region.

On investigation, he was mildly anemic with Hb 10g/dl and ESR 102mm/hr. CBP , Prothrombin time/INR, bleeding time, clotting time ,serum electrolytes, liver function tests and renal function tests were within normal limits. Chest x ray was normal. Ultrasonogram of abdomen showed mild hepatomegaly with increased bladder wall thickness. CT pulmonary angiogram was normal. Doppler study of lower limb revealed dilated right EIV,SFV,and popliteal vein with echogenic material and absence of colour flow (fig 1-3)

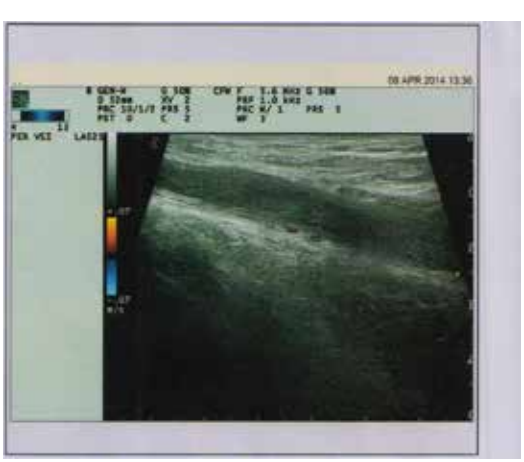

FIGURE.1.THROMBOSIS IN THE RIGHT EXTERNAL ILIAC VEIN

Volume : 4 | Issue : 9 | September 2014 | ISSN - 2249-555X

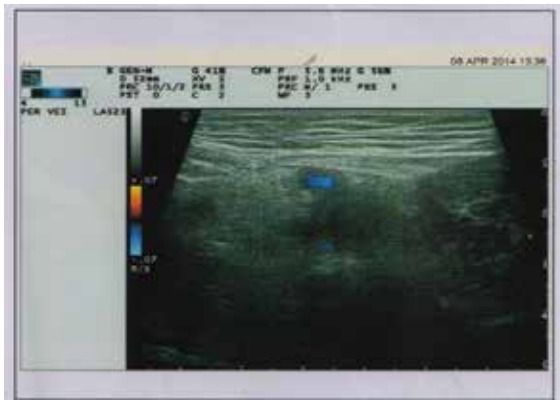

FIGURE.2.THROMBOSIS IN THE RIGHT COMMON FEMORAL VEIN

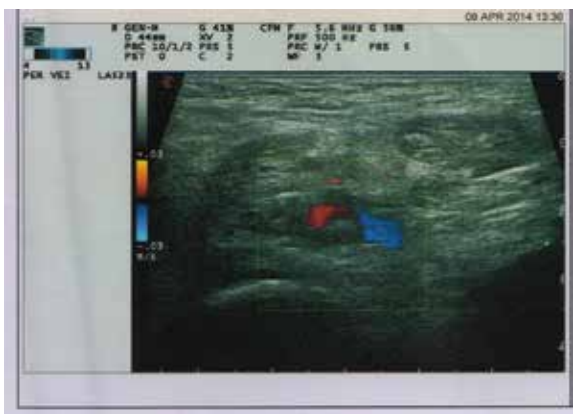

FIGURE.3. THROMBOSIS IN THE RIGHT POPLITEAL VEIN

Suggestive of deep vein thrombosis.In view of the unprovoked nature of the disease,throombophilia was suspected and investigations for thrombophilia profile was sent before starting the patient on low molecular weight heparins, which revealed protein c (functional) 22% of normal(70-140), protein s (functional) 22% of normal(60-140), antithrombin 3(functional) was 110% of normal(80- 120), Patient was then overlapped with oral acenocoumarol with discontinuation of LMWH. After achieving the therapeutic INR the patient was discharged on oral anticoagulant therapy with acenocoumarol 4 mg. Since then patient was on regular followup with close monitoring of PT/INR. Patient symptomatically improved as pain and swelling subsided.

DISCUSSSION

Venous thrombi are intravascular deposits composed of fibrin and red cells with a variable platelet and leukocyte component[4].Deep vein thrombosis (DVT) is a common illness that can result in suffering and death if not recognized and treated effectively. Death can occur when the pulmonary emboli, which pass to and obstruct the arteries of the lungs. DVT and pulmonary embolism (PE) most often complicate the course of sick, hospitalized patients but may also affect ambulatory and otherwise healthy persons. Deep vein thrombosis is a major complication in orthopedic surgical patients and patients with cancer and other chronic illnesses . The factors traditionally implicated in the pathogenesis of venous thrombosis are activation of blood coagulation, venous stasis, and vascular injury

The most important natural anticoagulants are protein C, protein S, and antithrombin .The normal balance between clotting and bleeding is disrupted when there is a deficiency of one of the natural anticoagulants. Low levels of the natural anticoagulants or natural anticoagulants that do not work properly can either be inherited or may occur during certain life event. People with hereditary protein C or protein S deficiency have about a 2- to 11-fold increased risk for developing a DVT or PE in comparison with those without a deficiency.Deficiencies of the natural anticoagulants are associated primarily with an increased risk for blood clots in veins, and seem to play little or no role in development of blood clots in arteries, eg, heart attack and stroke. However, a recent study suggests that protein C and protein S (but not antithrombin) deficiencies may be associated with an increased risk for forming arterial blood clots in people younger than 55 years.

Warfarin is currently the most commonly prescribed anticoagulant for long-term treatment after a DVT or PE, but it must be given initially with an additional injectable anticoagulant (usually heparin, low-molecular-weight heparin, or a similar drug) until the warfarin is fully effective. If there is protein C or protein S deficiency, patient should not receive warfarin without first receiving another anticoagulant at the same time. Initial treatment with warfarin alone in people with protein C or protein S deficiency may temporarily make clotting worse or precipitate a new clot or a severe skin rash known as skin necrosis. After a first DVT or PE, the risk for developing a second clot is probably higher for individuals with a deficiency of one of the natural anticoagulants than for those without this deficiency. If there is deficiency of one of the natural anticoagulants but have never a blood clot, then routine be treatment with an anticoagulant. Another major risk factor for blood clots is obesity, which poses a more potent risk than some of the hereditary thrombophilias.

Routine screening for inherited thrombophilias is not indicated in patients with VTE provoked by immobility, surgery, and malignancy, or in those with arterial thrombosis with arteriosclerosis risk factors. The finding of a strong thrombophilia has several clinical consequences: it decreases the threshold to recommend long-term anticoagulation in a patient with unprovoked VTE[5]; Complete spontaneous lysis of large venous thrombi is uncommon, and even when patients with venous thrombosis are treated with heparin, complete lysis occurs in fewer than 10% of cases[6] Bleeding is by far the most common complication of oral anticoagulant therapy. Randomized studies have shown that the risk of bleeding is influenced by the intensity of anticoagulation, and several studies have shown that the risk of clinically important bleeding is reduced by lowering the therapeutic range for the INR from 3.0 to 4.5 to 2.0 to 3.0 [7]

CONCLUSION: in every case of DVT presenting in a young patient underlying coagulation defects should be considered. We recommend doing protein C and S in a young patient of DVT as a initial workup.

No conflict of interest
Funding:no source of funding

Volume : 4 | Issue : 9 | September 2014 | ISSN - 2249-555X

REFERENCE

1.Combined Protein C and Protein S Deficiency Presenting as Deep Vein Thrombosis Sharma Bishnu Hari Kattel, Sudhir Regmi, Arpana Neopane and Damber Badhur Karki | 2.Deficiencies of Natural Anticoagulants, Protein C, Protein S, and Antithrombin ,Brea Lipe, MD,Deborah L. Ornstein, MD | 3.Deficiencies of natural anticoagulants.circ.ahajournals.org/content/124/14/e365.by B Lipe ſ 2011 | 4.Freiman DG. The structure of thrombi. In: Colman RW, Hirsh J, Marder VJ, Salzman EW, eds. Thrombosis and Hemostasis: Basic Principles and Clinical Practice. Philadelphia, Pa: JB Lippincott; 1987:1123-1135 | 5.Curr Treat Options Cardiovasc Med. 2009 Apr;11(2):114-28.Thrombophilia: 2009 update.Foy P, Moll S. | 6.Gallus AS, Hirsh J. Treatment of venous thromboembolic disease. Semin Thromb Hemost. 1976;2:291-331 | 7.Landefeld CS, Goldman L. Major bleeding in outpatients treated with warfarin: incidence and prediction by factors known at the start of outpatient therapy. Am J Med. 1989;87:144-152 |