Systemic Lupus Erythematosus with Deep Vein Thrombosis and Cutaneous Ulcer

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Abstract
We are reporting a case of systemic lupus erythematosus (SLE) with left upper limb and lower limb deep vein thrombosis (DVT) due to protein S deficiency which was aggravated by anticoagulants. Oral anticoagulant-induced skin necrosis also developed in this patient. This patient was negative for anti-phospholipid antibodies (APLA). Such a case is rarity where SLE patient without APLA has protein S deficiency.

Introduction
Protein S deficiency is rare in SLE and is usually seen in association of anti-phospholipid antibody syndrome. In the present case, protein S deficiency was not associated with anti-phospholipid antibodies (APLA) and such a case is rarity. Protein C deficiency in our patient is secondary to oral anticoagulant therapy or due to deep vein thrombosis (DVT) itself. Oral anticoagulant-induced skin necrosis in patient of congenital protein S deficiency is also very rare. We describe a patient of SLE with protein S deficiency who presented with DVT and developed oral anticoagulant-induced skin necrosis.

Case Report
A 52 year old post-menopausal female who was a known case of SLE for the last seven years and was on treatment for SLE (steroids and azathioprine) presented with complaints of acute onset pain and swelling in left upper limb for five days. On examination diffuse swelling of left upper limb was present which was tender and pitting and one axillary lymph node was also enlarged on the left side to the size of 3 cms. Breast examination was normal. Color Doppler study diagnosed deep vein thrombosis (DVT) of left upper limb. Axillary lymph node biopsy was suggestive of reactive hyperplasia. Antiphospholipid antibody profile (anti-cardiolipin antibody IgG and IgM, β2 glycoprotein1 IgG and IgM and lupus anticoagulant) was negative.

Treatment with enoxaparin 1 mg/kg subcutaneous (SC) twice a day and oral anticoagulant (acenocoumarin 2 mg/day) was started. Enoxaparin was stopped after five days while oral anticoagulant was continued. After five days of stopping enoxaparin, swelling in the left upper limb increased and also involved the left lower limb. Repeat color Doppler examination of venous system was done which suggested DVT in left upper limb and lower limb.

Meanwhile patient also developed a necrotizing ulcer in left axillary area (Figure 1a). Biopsy from ulcer edge was taken which showed acute inflammatory exudate. Estimation of protein C and S functional activity was suggestive of very low activity of both (protein C activity- 19%, [normal 70-130%]; protein S activity- 22%, [normal 77-143%]).

Oral anticoagulant was stopped and enoxaparin 1 mg/kg SC BID was restarted. Patient improved, swelling subsided in upper and lower limb and ulcer also healed over a period of one month (Figure 1b).

Repeat estimation of protein C and S activity was done after three months which showed normal protein C activity (126%) and decreased protein S activity (49%).

Discussion
Here is a case of SLE with left upper limb and lower limb DVT due to protein S deficiency which was aggravated by oral anticoagulants. Protein S has an established role in the protein C anticoagulant pathway, where it enhances the factor Va and factor VIIIa inactivating property of activated protein C (APC).1 Protein S deficiency is rare in SLE and is usually seen in association of APS.2 In the present case, protein S deficiency was not associated with Anti-phospholipid antibodies (APLA) and such a case is rarity where SLE patient without APLA has protein S deficiency. Protein C deficiency in our patient is secondary to either oral anticoagulant or DVT itself, protein C normalized after stopping oral anticoagulants showing its deficiency may have been due to oral anticoagulant. Oral

Fig. 1: (a) Cutaneous ulcer, (b) Healed ulcer

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anticoagulant-induced skin necrosis is a rare complication of anticoagulant treatment. Warfarin-induced skin necrosis occurs almost exclusively in patients with venous thrombosis between the 3rd and 10th day after starting oral anticoagulation. Although protein C deficiency is the most common underlying hypercoagulable state reportedly associated with warfarin skin necrosis, very few cases have been linked to congenital protein S deficiency. Protein S deficiency should be considered when a patient of SLE presents with DVT and develops a necrotizing ulcer after oral anticoagulant therapy.

References