Antiphospholipid Syndrome may Lead to Soft tissue Defect Requiring Reconstruction

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Antiphospholipid syndrome (APS) manifests as systemic thrombotic disorders including recurrent deep vein thrombosis, pulmonary thromboembolism, brain stroke and fetal loss in the presence of antiphospholipid antibodies. The syndrome can be secondary to many causes including systemic lupus erythematosus or pulmonary antiphospholipid syndrome. The many etio pathogenic mechanisms involved generally act together so that it is difficult to say which is the main cause, especially in elderly patients.

Systemic lupus erythematosus (SLE) is an auto immune disease of unknown origin affecting virtually all organ systems. Beyond genetic and environmental factors, cytokine imbalances contribute to immune dysfunction, trigger inflammation, and induce organ damage. Patients with the APS who do not have SLE are considered to have the primary antiphospholipid antibody syndrome; patients with SLE who also have both antiphospholipid antibodies and relevant clinical events are considered to have the secondary antiphospholipid antibody syndrome. The evolution depends mainly on the multi-organ involvement and how early immune suppressive and/or anticoagulant treatment is started. We present a case with SLE who had soft tissue defect requiring reconstruction in this report.

Case report

The patient was 39 years-old female and had a recent high dose corticosteroid treatment for her active disease. Her past medical history showed that she was diagnosed as having SLE 10 years earlier. SLE was diagnosed on the basis of its clinical manifestations and the demonstration of characteristic anti-nuclear antibodies. The first manifestation of the disease was started with 2 episodes of skin rash that was erythematous, non blanching presented at an interval of 10 months. Symmetrical weakness of proximal muscle and elevation of serum muscle enzymes (serum aldolase) were detected at that time. During both episodes, the treating consultant found thrombocytopenia and the first episode was treated within travenous gamma globulin, and during these condepiode, the patient was started on oral prednisolone 2 mg/(kg day), which continued till the age of 34 years. After this, the patient had an uneventful period of 1 year; again after 1 year, th epatient presented with similar complaints described above. Further investigations were done that showed thrombocytopenia, muscle creatine kinase maximally raised, normal electromyogram, normal liver function test, and serum aldolase within the normal range.

Last active disease period was begun 5 months earlier. She had fatigue, fever, weightloss, skinrash, arthritis or arthralgias. Treatment was initiated with injection methyl prednisolone (MP) 1gm IV 3 days followed by oral prednisolone 1mg/kg body weight. The patient responded promptly with gradual amelioration of all clinical findings within the ensuing 10 days. Investigation at this time revealed Hb 9.9 gm/dl, normochromic normocytic RBCs, reticulocyte count 1.5%, total leucocyte count 5700/mm3 (polymorphs 77%, lymphocyte 25%), MCV 90.2, MCH 29.6, MCHC 30.8, platelet count of 90,000/dl, urea 21mg/dl, Cr 0.7 mg/dl, uricacid 3.6mg/dl, Na 137 meq/l, and K 4.4 meq/l. Liver and thyroid function tests, serum urea, creatinine were normal. Laboratory criteria were positive titers of lupus anticoagulant antibody moderate to high titers of anti cardiolipin Ig M(45MPL UI/ml) and moderate titers of Beta2-glycoprotein. The 24-h urinary protein excretion rate was 178 mg. Ultra sonography of the abdomen was within normal limits without any apparent pelvic/ovarian mass. Gynecological examination and Papanicolaou smear of the cervical region was normal. Mammography was within normal limits. Activsated partial thromboplastin time, anti-phospholipid antibody IgM (aPL) were within normal limits. At the end of treatment process, there was a skin gangrene over her left pretilbiaarea. Previous skin biopsy of the same lower limb skin showed the thrombotic vasculopathy with no evidence of vasculitis. The cyanotic areas progressed to gangrene. Then, she was referred to Plastic Surgery for her pre tibial wound. The wound exposing tibia was 7x10 cm in diameter. The defect was reconstructed with a hemo gastronemius flap and full thickness skin graft and a unevenfull healing was achieved (Figure 1a-c). During the wound healing period there was no wound healing delay or systemic activation of disease.

Discussion

APS is the most common acquired thrombophilia, characteristic clinical manifestations are heterogenous with venous and arterial thrombosis. Diagnosis is made through the association of clinical and laboratory criteria. The clinical criteria are thrombosis or history of obstetrical morbidity. Acceptable tests for lupus anticoagulant include the dilutated partial thromboplastin time, the kaolin clotting time test, or the Russell viper venom time test. Positive laboratory

Figure 1:

a) A pre Tibial defect occurred during the management of the disease.
b) Medial based hemi gastrocnemius flap transfer to the defect.
c) Complete healing of the wound.

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tests in clinically well persons do not justify either diagnosis of the APS or prophylactic treatment, but they do justify re examination and monitoring of the patient. There are striking inconsistencies among commercial laboratories doing antiphospholipid antibody assays [1]. Because antinuclear and anti-DNA antibodies occasionally occur in patients with the primary APS, clinical criteria for both the primary APS and SLE must be present for both diagnoses to be made [2,3]. In the secondary APS, manifestations of the primary antiphospholipid antibody syndrome adversely affect survival [4]. Some patients with SLE have transiently present (usually low-titer) antiphospholipid antibody that varies with disease activity. These patients, in contrast to those with sustained high-titer antibody, generally do not have complications of the APS [5]. In our case there was also high titers of antiphospholipid antibody and there was no internal organ involvment. Patients with positive “lupus anticoagulant” tests but normal results on activated partial thromboplastin time or Russellvervenom time tests generally have normal ELISA results for anticardiolipin antibody. The isolated “lupus anticoagulant” is thus of unknown significance. Catastrophic APS occurs in 1% of APS patients and associated with a mortality rate upto 50% [6]. In these sub group there are usually evidences of disseminated intravascular coagulopathy and micro angiopathichemolyticanemia. 

Although may affect any organ, orthopedic and cerebral involvement is a relatively low and under-recognized feature of APS [7,8].Our search yielded no reports concerning softtissue necrosis which required reconstruction in a case of APS. This patient had an unexpected evolution with improvements of tissue and skin gangren after highdose steroids. Our search for septic screen at admission to plastic surgery clinic was negative. There was no prolongation of PT and a PTT . Platelet counts at surgery time were normal and she was taking acetyl salicylic acid (100 mg/day). Under anti coagulant therapy there was no bleeding complication post operatively. So, this presentation highlights that reconstructive needs can occur in this group of patients as a complication of disease progress.

References