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BRIEF REPORT

Fatal thrombotic thrombocytopenic purpura in discoid lupus

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ABSTRACT

The association between TTP and systemic lupus erythematosus (SLE) has been infrequently reported, however an association with discoid lupus was not reported in the literature. Usually, patients with TTP have increased SLE activity and frequent renal involvement. Here we present a case of a 65 year old man who presented to the emergency department with malaise, fever and purpura of several days duration. His past medical history was significant for discoid lupus diagnosed by biopsy 34 years ago as well as hypertension. On laboratory investigation, he had low complement levels and ADAMTS13 level of < 5 %. A peripheral blood smear showed the presence of schistocytes. On the basis of clinical and biochemical findings, he was diagnosed with TTP.

Key words: thrombotic thrombocytopenic purpura, discoid lupus, systemic lupus erythematosus, plasma exchange

Thrombotic thrombocytopenic purpura (TTP) is an acute and uncommon disorder resulting from occlusion of small arterioles and capillaries by microthrombi. It is defined as a pentad consisting of microangiopathic hemolytic anemia, thrombocytopenia, nonfocal neurologic abnormalities, renal function impairment and fever. Discoid lupus is a variant of cutaneous lupus characterized by well-defined inflammatory plaques that may occur independently or as manifestation of systemic lupus erythematosus. There are few reports about the association between TTP and SLE but the association of TTP with discoid lupus was not reported in the literature.[1]

A 65-year-old man who presented to the Emergency Department (ED) due to lightheadedness , purpuric rash, subjective fever, and dark urine for 4 days, he also had one episode of confusion with difficulty expressing his thoughts. The patient was diagnosed with discoid lupus by biopsy in 1980 and was not on any immunosuppressants as he showed no clinical signs of lupus in the last 34 years. Physical examination revealed pale conjunctivae, petechiae on his extremities and lower trunk area (Fig. 1A), and discoid discoloration on scalp (Fig. 1B).

The blood tests on admission showed: hemoglobin 11.6 g/dl, platelets 11,000 mm3, LDH 1641 U/L, haptoglobin <10mg/dl, total bilirubin 4.5 mg/dl, direct bilirubin 1 mg/dl, peripheral schistocytes (+), serum creatinine 1.4 mg/dl, troponin 0.34ng/dl, ANA (+) <1:80, C3: 86 (Normal 88), C4: 6(Normal 10),ds-DNA (-),ADAMTS13 <5%, blood smear showed schistocytes. The diagnosis of TTP was made on the basis of biochemical and clinical findings. He was immediately treated with high dose steroids and plasma exchange. On day 3 of plasma exchange, patient suffered from shortness of breath with lung congestion due to cardiac ischemia and atrial fibrillation leading to intubation and subsequently fatal asystole. TTP is a rare and usually proceeding complication in the context of SLE.2,3 It is seen in 2% of patients who have SLE and specially in those with severe lupus activity and renal involvement. Diagnosis in these patients can be challenging as the two diseases have considerable overlapping clinical features however the exact diagnosis is important as it may carry substantial prognostic and therapeutic implications with a majority of patients having both SLE and TTP dying with TTP.[4]

Anemia, thrombocytopenia, fever, neurological abnormalities, and renal disease occur in both, however, a microangiopathic peripheral blood smear with an initial schistocyte count of >1% without any other cause helps distinguish TTP from SLE flare.5,6 Although now the diagnostic criteria has been revised from the pentad to the dyad of thrombocytopenia and microangiopathic hemolytic anemia.[3]

The occurrence of concomitant TTP and SLE has a "slower tempo" of development probably because patients diagnosed with SLE are on immunosuppressive therapy and corticosteroids, which may suppress the immune mechanisms involved in the pathogenesis of TTP. The association of SLE and TTP although

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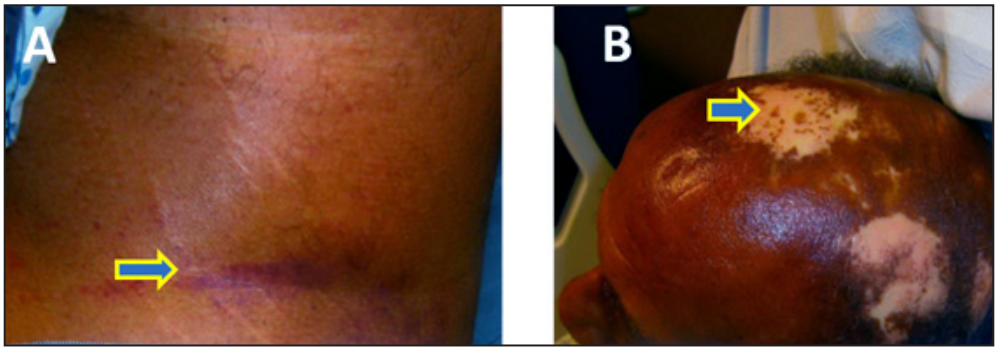

Figure1, (A) Petechiae, purpura, and ecchymoses on extremities;
(B) Discoid discolor on scalp (Arrow)

rare, can be fatal, which warrants early diagnosis and aggressive management. Immediate treatment with plasma exchange and corticosteroids should be initiated as soon as the diagnosis of TTP is suspected. When seen together, these two etiologies have a higher mortality than either disease alone however plasma exchange transfusion has decreased the mortality of TTP from 90% to 8%-25%. Asymptomatic myocardial infarction is an important cause of death in TTP due to microthrombi formation that affects the conduction system. Therefore, autopsy of patients with TTP who died suddenly should involve a detailed examination of the heart tissue.[7]

Knowledge of this syndrome and the high risk of sudden death justifies the need for quick diagnosis and treatment.[6] Patients with SLE who develop TTP, do significantly worse than those with idiopathic TTP, with mortality of 34%-62.5%.

The case we presented is noteworthy because it shows for the first time that low activity discoid lupus not only can cause TTP but also can be fatal and that such cases need more aggressive treatment with immunosuppressants such as rituximab and cyclophosphamide.

Physicians should be alert of the possibility of TTP when treating a patient with discoid lupus. Even in cases of discoid lupus with low disease activity, TTP can be life-threatening. Hence patients with history of discoid lupus who present with TTP are recommended to undergo aggressive treatment with plasma exchange and immunosuppressants.

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