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Thrombotic Microangiopathy in a Patient with Systemic Lupus Erythematosus

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Abstract

The term thrombotic microangiopathy (TMA) includes several medical conditions characterized by the association of mechanical hemolytic anemia and peripheral thrombocytopenia. There are few studies in the literature focusing on this pathology, and its causes are dominated by bacterial and viral infections and systemic disease. Indeed, TMA is a serious complication that can occur in patients with systemic lupus erythematosus (SLE), adversely affecting prognosis and increasing mortality. The etiology of TMA in these patients may be multifactorial and may overlap among different entities. We present the case of a 22-year-old female who was previously diagnosed with SLE. She was hospitalized with acute kidney injury, severe bicytopenia, and other features consistent with her flare of lupus.

Keywords: *Thrombotic microangiopathy; Lupus erythematosus*

Introduction

The term thrombotic microangiopathy (TMA) includes several medical conditions characterized by the association of mechanical hemolytic anemia and peripheral thrombocytopenia [1]. Few studies in the literature have focused on this condition, and its etiology is dominated by bacterial and viral infections and systemic disease [2]. Systemic lupus erythematosus (SLE) is a chronic autoimmune disease of unknown cause. Renal involvement, usually due to immune complex-mediated glomerular disease, is common, but vascular disease also occurs and generally has a negative prognosis and increases mortality [3]. We report his case of a patient diagnosed with class IV LN presenting with TMA.

Patient and Observation

A 22-year-old Moroccan women with LN was admitted with acute kidney injury [serum creatinine (SCr): 6.1 mg/dl], bicytopenia (haemoglobin=7.9 g/dl, platelets=64G/L), positive anti-dsDNA. The anemia (hemoglobin = 7.9g/dl) is normocytic (VGM=77.6fl), normochromic (CCMH=35.1g/dl and TCMH=27.2pg), regenerative (reticulocyte rate=143G/L). The rate of schistocytes was 7% (Figure 1). Thrombocytopenia is severe at 64G/L. Hyperleukocytosis was found at 16.1G/L with a predominance of neutrophils (15.5G/L) [4,5].

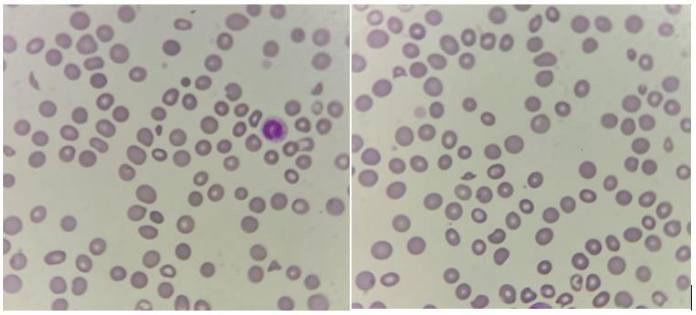

Figure 1: *Microscopic pictures of the schizocytes found in the patient.*

Discussion

TMA represents a group of syndromes with multiple etiologies, both inherited and acquired [6]. Globally, TMA classification can be complex and confusing. One way to classify them is to classify them into primary and secondary causes. Primary TMAs include TPP (thrombotic thrombocytopenic purpura) and atypical HUS (hemolytic uremic syndrome). Secondary TMAs include infectious diseases such as Shiga-toxin producing E. coli and

other conditions such as malignancies or autoimmune diseases as SLE [7]. Clinically detectable TMA is rare in SLE and mostly has a histopathological nature. It may magnify the renal destruction caused by lupus by causing and increasing local inflammation with damage to the diseased kidney. Patients with LN have more severe and active renal disease. The etiology of TMA in LN remains unclear and may be multifactorial. Since SLE is an immune complex-mediated disease, activation of the classical pathway has been suggested to play an important role in the development of TMA (Table 1).

Table 1: *Analytical parameters of the patient.*

Analytical parameters	Value	Reference value
Hemoglobin	7.9 g/dl	11.5-17.5 g/dl
VGM	77.6 fl	76-96 fl
CCMH	35.1 g/dl	31-36 g/dl
TCMH	27.2 pg	24.4-34 G/L
Leukocytes	16.1 G/L	3.8-11 G/L
Platelets	64 G/L	150-445 G/L

Several studies have shown that dysregulation of the alternative complement pathway may also be involved, which is consistent with low C3 and C4 levels in LN [4,5]. This case reports TMA in an SLE patient. The patient presented with bicytopenia and other features consistent with lupus flares. However, the severity of this case was explained by her diagnosis of TMA, one of the most serious complications in SLE patients. TMA is a complex process involving imbalances between immunity, coagulation and complement caused by a variety of factors, in this case a severe lupus flare-up. Local or systemic complement activation induces endothelial damage that is present in both primary and secondary causes of TMA. Whatever the cause, TMA is a devastating condition that results in systemic and renal damage and compromises patient prognosis and survival [6,7].

Conclusion

Thrombotic microangiopathy (TMA) is a severe renal vascular injury presenting with progressive life-threatening thrombocytopenia, microangiopathic hemolytic anemia, and progressive renal failure. Although rare, TMA is commonly found in patients with Class IV lupus nephritis (LN) and plays an important role in the progression and exacerbation of LN.

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