Case Report

Catastrophic antiphospholipid syndrome in leprosy

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ABSTRACT: Catastrophic antiphospholipid syndrome is an acute and life threatening variant of antiphospholipid syndrome with a high mortality rate. Many infections are known to be accompanied by the thrombotic manifestations of this syndrome. We came across a patient of leprosy who developed bowel ischaemia secondary to mesenteric venous thrombosis as a part of catastrophic antiphospholipid syndrome and later on succumbed. We thereby wish to highlight the need for early diagnosis and aggressive treatment of this potentially fatal condition in patients with infections.

KEY WORDS: Antiphospholipid; Antibody; Catastrophic; Leprosy; Syndrome

INTRODUCTION

Antiphospholipid syndrome (APS) is the association between antiphospholipid antibodies and hypercoagulable state. An acute and devastating variant of this is termed catastrophic antiphospholipid syndrome. Many infections appear to be accompanied by clinical manifestations of the APS, particularly catastrophic APS. The mortality rate is high and death is usually due to multiorgan failure. We present a case of catastrophic antiphospholipid syndrome in a patient with leprosy who succumbed to bowel ischaemia thereby highlighting the need for a high degree of suspicion and early aggressive treatment.

CASE DETAILS

A 35 year old male presented with non-healing ulcers on anterior aspect of both legs since 2 weeks. He was a recently diagnosed case of leprosy and was on treatment for the same. On clinical examination he had madarosis and thickened ulnar, radial and greater auricular nerves. All peripheral pulses were well felt. There was no cardiac murmur or carotid bruit. Investigations showed platelet count 80000/mm³. Chest radiograph and urine examination were normal. Lipid profile and tests for other hypercoagulable states (protein C, protein S, Antithrombin III, homocysteine, factor V Leiden) were normal. Anticardiolipin antibodies (ACLA) as determined by ELISA method were positive (IgG 35.0 GPLU/ML <8.0, IgM 154 MPLU/ML <8.0). After six weeks, prothrombin time and aPTT were normal, but, anti neutrophil cytoplasmic antibodies were negative. Eight weeks later, he developed abdominal pain and distension with fluid filled blisters over both legs (Figure 1) and developed ulcerations (Figure 2).

Figure 1: Blisters on the legs
Figure 2: Ulcers on the leg

Abdominal examination showed free fluid with well heard bowel sounds. Ultrasound of the abdomen showed diffuse oedematous bowel loops and ascites. Doppler of the lower limb arteries did not reveal any abnormality. On the next day, he developed bilious vomiting and blackish stools. Bowel sounds were absent. Continuous nasogastric aspiration with antibiotics and parenteral nutrition was started. Serum electrolytes at this stage were normal. CT scan with CT angiography of abdomen showed diffuse thickening of small and large bowel loops with superior mesenteric vein thrombosis. Patient was put on heparin. Colonoscopy showed multiple submucosal hemorrhages in the ascending and transverse colon. The ileoceleal valve was erythematous and edematous. The terminal ileum and rest of the visualized colon were normal. The patient deteriorated and later, succumbed. An autopsy showed ischaemic colitis with pulmonary haemorrhages. A final diagnosis of Leprosy with catastrophic antiphospholipid syndrome was made.

DISCUSSION

The term “antiphospholipid syndrome” was used to denote the clinical association between antiphospholipid antibodies and a syndrome of hypercoagulability. The most commonly detected subgroups of antiphospholipid antibodies (aPL) are lupus anticoagulant, anticardiolipin antibodies, and anti–β₂-glycoprotein I (β₂-GPI) antibodies. The syndrome can occur in its primary form or secondarily in association with other autoimmune disorders. A minority of patients with the antiphospholipid syndrome present with an acute and devastating syndrome termed “catastrophic antiphospholipid syndrome” characterized by multiple simultaneous vascular occlusions throughout the body. The organs most commonly affected are the kidneys and the lungs. Manifestations include renal thrombotic microangiopathy, respiratory distress syndrome (ARDS) and disseminated intravascular coagulation. Mandal et al described a patient with catastrophic APS who presented with dilated cardiomyopathy and bilateral retinal artery thrombosis. Al-Ansari reported a case of a 51 year old woman with left renal infarction, thrombotic superior mesenteric artery and an infarcted left adrenal gland diagnosed to have APS. The mortality rate is high and death is usually due to multiorgan failure.

Many infections have been found to be associated with aPL positivity. Specific triggering infections encountered to date include malaria, dengue, typhoid fever, viral infections of upper respiratory tract, urinary infections and sepsis. Canpolat et al have described catastrophic APS in an adolescent girl with parovirus B19 infection. Ehrenfeld et al described two patients of severe falciparum malaria with ARDS in one of them, and pulmonary embolism and splenic infarction, in the second. Both were found to have high levels of anticardiolipin antibodies. It has been emphasized and reported that infections may not only trigger the production of these antibodies but also appear to be accompanied by clinical manifestations of the APS itself. This has been seen particularly in patients with catastrophic APS. The pathophysiology of this disorder is poorly understood. Thrombosis can be self-perpetuating in patients with an underlying hypercoagulable state. Molecular mimicry has been proposed for the development of catastrophic APS following infections. Aberrant neutrophil activity has been implicated in the organ failure of severe sepsis, and it is likely that a similar situation exists in patients with catastrophic APS. Examination of autopsy specimens from patients with multiple organ failure and severe sepsis shows sequestration and aggregation of neutrophils in renal blood vessels and large-scale infiltration in the lungs, resulting in ARDS. In patients with leprosy (particularly in the multibacillary type of leprosy) the anticardiolipin antibodies may be β₂-GPI dependent, as is found in autoimmune diseases. Lucio’s phenomenon is a rare manifestation of leprosy in which the histopathological findings are related to microvascular thromboses in the absence of inflammatory infiltration of the vessel walls. Treatment of catastrophic antiphospholipid syndrome includes either plasmapheresis or intravenous immune globulin. The rationale for plasmapheresis derives from its documented effectiveness in treating the hemolytic–uremic syndrome and thrombotic thrombocytopenic purpura. The fibrinolytic
agents streptokinase and urokinase have also been used to treat acute thrombotic microangiopathy with varying success. Since thrombosis tends to be a self-perpetuating process, an aggressive therapeutic approach is warranted in these patients.

CONCLUSION

Catastrophic antiphospholipid syndrome is an uncommon and potentially fatal condition. A wide variety of infections including leprosy can be associated with thrombotic events in patients with APS. More patients develop catastrophic APS following infection, as opposed to simple APS. This emphasizes the need for early diagnosis and aggressive treatment.

REFERENCES