AMERICAN JOURNAL OF MEDICAL CASE REPORTS

ISSN 2374-216X

Antiphospholipid syndrome: A rare cause of multiple intra-abdominal infarcts

Bharat Khialani*; Christopher Sia

***Bharat Khialani**

Department of Medicine, The Alfred Hospital, Melbourne, Australia

Abstract

Antiphospholipid syndrome (APS) results in a hypercoagulable state, provoking arterial and venous thrombosis. Here we describe a case of catastrophic APS on a background of previously asymptomatic lupus anticoagulant, resulting in severe abdominal pain due to superior mesenteric artery thrombosis, and renal and splenic infarcts resulting in multiorgan failure and Intensive Care admission. Diagnosis was made on laboratory findings of prolonged activated partial thromboplastin time (APTT), positive lupus anticoagulant and non-arterial phase CT scans of the abdomen. Management involved urgent surgical thrombectomy and anticoagulation.

Keywords

antiphospholipid syndrome; multiorgan failure; intensive care; abdomen

Clinical Case

A 62-year-old male presented with acute onset abdominal pain. His past medical history was significant for hypertension and paroxysmal atrial fibrillation, maintained in sinus rhythm on sotalol. He was on aspirin 100mg daily for systemic embolism prophylaxis (CHADSVASC = 1) [1]. Four years prior to this admission, he was incidentally found to have a positive lupus anticoagulant after a series of "screening" blood tests ordered by his General Practitioner, without prior thromboembolism. However, he had been lost to follow up for this, and was never reviewed by a specialist practitioner.

On examination in the Emergency Department, he was febrile at 38.2 degree Celsius, in sinus rhythm and hypertensive with a blood pressure of 190/85. Abdominal examination revealed upper abdominal tenderness and guarding. His venous lactate was mildly elevated at 3.0 mmol/L (N=0.6-2.2) and his white cell count was elevated at 16.8 x 10^9/L. His renal function was deranged, suggesting acute kidney injury.

Computed Tomography (CT) scan of his abdomen and pelvis revealed bilateral wedge shaped renal infarcts and a splenic infract (Figure 1). There was poor opacification of the superior mesenteric artery (SMA) however there was no evidence of bowel wall thickening to suggest bowel ischaemia. This was further assessed with a CT abdominal angiogram which demonstrated a 3.3cm occlusive SMA thrombosis.

Review of blood tests from previous admissions revealed an activated partial thromboplastin time (APTT) of 96 (N=26.0-38.0) and a positive lupus anticoagulant. Repeat coagulation testing demonstrated a prolonged APTT that failed to correct with mixing studies, confirming the diagnosis of antiphospholipid antibody syndrome. The results of investigations are summarised in Table 1. Transthoracic and transoesophageal echocardiogram found no evidence of endocarditis, atrial or ventricular thrombus.

The patient was commenced on an unfractionated heparin infusion and had an emergent surgical thrombectomy, before transfer to the Intensive Care Unit (ICU) for supportive management. Immunosuppresants, particularly glucocorticosteroids, were considered but not utilized during his admission. He was discharged home on warfarin after 2 weeks in hospital.

Discussion

APS is a systemic autoimmune disorder characterized by arterial or venous thrombosis and/or pregnancy morbidity in the presence of persistent laboratory evidence of antiphospholipid antibodies. The most common site of arterial thrombosis is in the cerebral vasculature [2], usually resulting in a stroke or transient ischemic attack. Occlusions of the retinal, coronary, renal, and mesenteric arteries can also sometimes occur. Catastrophic APS is used to describe APS cases presenting with a life-threatening condition due to multi-organ involvement, in particular resulting in renal or hepatic failure.

The diagnosis of APS is based on a combination of clinical features and laboratory findings. The Sapporo or Sydney Classification Criteria [3] may be useful for clinicians in diagnosing some patients and documenting key disease features.

In patients presenting with severe acute abdominal pain, intestinal ischaemia should be considered. Although embolism due to cardiac arrhythmias such as atrial fibrillation, infective endocarditis, recent myocardial infarction and aortic atheroma are more common, prothrombotic conditions such as APS should be considered in younger patients. A normal lactate does not exclude the diagnosis of mesenteric ischaemia (pooled sensitivity 86%, 95% CI 73-94%) [4]. The diagnosis may also be suspected on CT scans of the abdomen and pelvis, even on non-arterial phase contrast scans as demonstrated in this case. Management of arterial thrombosis usually involves urgent revascularisation either via surgical thrombectomy or endovascular techniques, supplemented by systemic anticoagulation. Glucocorticosteroids, plasma exchange, intravenous immunoglobulin and Rituximab have been utilized in catastrophic APS.

Our patient raises the controversial issue of management of the asymptomatic patients with positive antiphospholipid antibodies without evidence of prior thrombolembolism or underlying connective tissue disease. The estimated incidence rate of thrombosis in unselected cases is approximately 1 per 100 patient-years [2], however this estimate varies widely. Warfarin is the standard of care medication for the chronic treatment of patients with APS. The INR should be maintained between 2.0 and 3.0 to prevent recurrent events. In retrospective series, aspirin alone has been of little or no benefit for the prevention of thrombotic APAS manifestations in patients who have experienced previous events [5].

Lessons from Practice

- Catastrophic APS is a rare but serious cause of widespread arterial thrombosis.
- The diagnosis is made on clinical and laboratory criteria.
- Management consists of anticoagulation and immunotherapy with glucocorticoisteroids, plasma exchange and intravenous immunoglobulin.
- The role of chemoprophylaxis for primary prevention in patients with positive antiphospholipid antibodies remains controversial; while low dose aspirin has limited benefit.

Table

Table 1: Results of coagulation studies.

Test	Value	Normal Range
International normalised ratio (sec)	1.1	0.9-1.3
Prothrombin Time (sec)	14.4	10.6-15.3s
Activated Partial Thromboplastin Time (sec)	100.3 (failed to correct with mixing)	26.0-38.0s
Reptilase Time (sec)	15.8	14.0-24.0s
DRVVT Screen Ratio	1.49	0.80-1.20
DRVVT Screen Mix Ratio	1.26	0.80-1.20
DRVVT Confirm Ratio	1.06	0.80-1.20
Anticardiolipin IgG (IU/ml)	1	0-10
Beta-2 Glycoprotein IgG (IU/ml)	1	0-7
Prothrombin gene mutation	Not detected	
Factor V Leiden	Not detected	
Antithrombin (%)	88	86-134
Protein C function (%)	91	72-153
Protein S (%)	135	65-146
ANCA	Not detected	
ANA	Not detected	
Anti-dsDNA	Not detected	
ENA	Not detected	
JAK 2 mutation	Not detected	