

Diffuse Systemic Arterial Calcification and Renal Infarction in a Young Female with Primary Antiphospholipid Syndrome Presenting as Accelerated Hypertension: A Case Report

Abstract

Antiphospholipid syndrome (APS) is a prothrombotic autoimmune disorder that can involve both arterial and venous systems, with renal manifestations commonly resulting from vascular injury. Diffuse premature systemic arterial calcification in young patients, however, is an unusual and underrecognized presentation.

We report the case of a 28-year-old female who presented with accelerated hypertension. Imaging with CT aortic angiography revealed multifocal renal infarctions along with extensive calcification involving multiple medium-sized arteries, including renal, coronary, internal mammary, intercostal, and iliac vessels. Renal function was preserved. Laboratory evaluation demonstrated elevated anti- β 2 glycoprotein I IgG antibodies, with persistent positivity on repeat testing after 12 weeks, while other autoimmune markers were negative.

Based on the presence of arterial thrombosis and persistent antiphospholipid antibody positivity, a diagnosis of primary antiphospholipid syndrome was established. The diffuse arterial calcification was considered secondary to chronic thrombotic vasculopathy.

This case highlights an unusual imaging presentation of antiphospholipid syndrome and emphasizes the importance of considering a prothrombotic etiology in young patients presenting with unexplained systemic arterial calcification and renal infarction.

Introduction

APS is a systemic autoimmune disorder characterized by the persistent presence of antiphospholipid antibodies and a clinical predisposition to arterial and venous thrombosis, as well as pregnancy-related morbidity [1]. The syndrome is most commonly associated with antibodies directed against β 2-glycoprotein I and cardiolipin, which promote a prothrombotic state through endothelial dysfunction, platelet activation, and complement-mediated injury [2].

Renal involvement in APS is well recognized and encompasses a spectrum of vascular lesions, including renal artery thrombosis, thrombotic microangiopathy, and cortical infarctions, collectively referred to as APS nephropathy [3]. These vascular insults may lead to secondary hypertension through activation of the renin-angiotensin-aldosterone system, even in patients with preserved renal function [4]. Imaging plays a crucial role in detecting these manifestations, with computed tomography (CT) angiography demonstrating characteristic findings such as wedge-shaped cortical defects and vascular occlusions [5].

Although arterial thrombosis is a known feature of APS, diffuse premature systemic arterial calcification in young individuals is exceedingly rare and not well described in the literature. Such findings may mimic accelerated atherosclerosis or metabolic vascular disease, posing a diagnostic challenge, particularly in the absence of conventional cardiovascular risk factors [6]. Chronic thrombotic vasculopathy resulting from recurrent endothelial injury and vascular remodeling has been proposed as a potential mechanism underlying this unusual presentation [7].

In this report, we describe a young female with primary APS presenting with accelerated hypertension, multifocal renal infarctions, and extensive systemic arterial calcification. This case highlights an uncommon imaging phenotype of APS and underscores the importance of considering prothrombotic disorders in young patients with atypical vascular calcification.

Case Presentation

A 28-year-old female presented with accelerated hypertension. There is no prior history of chronic kidney disease, diabetes mellitus, dyslipidemia, or known connective tissue disorder.

Laboratory evaluation revealed:

- Normal renal function parameters
- Positive ANA
- Elevated anti- β 2 glycoprotein I IgG (25 units)

- Negative anticardiolipin antibodies (IgG and IgM)
- Negative anti-dsDNA antibodies

CT aortic angiography was performed for evaluation of secondary hypertension.

Imaging Findings

Findings included:

- Multifocal wedge-shaped non-enhancing cortical areas in both kidneys consistent with chronic renal infarcts

Extensive calcification of:

- Renal arteries
- Right coronary artery
- Internal mammary arteries
- Inferior epigastric arteries
- Bilateral intercostal arteries
- Common and internal iliac arteries
- Splenic artery
- No aortic wall thickening or branch vessel mural enhancement
- No imaging features of large-vessel vasculitis
- Mild right lower lobe ground-glass/consolidation, non-enhancing

Discussion

APS is characterized by a prothrombotic state due to antibodies directed against phospholipid-binding proteins, most commonly β 2-glycoprotein I. While venous thrombosis is common, arterial involvement can occur and may affect coronary, cerebral, and renal arteries.

Renal manifestations of APS include: renal artery thrombosis, segmental renal infarction, thrombotic microangiopathy, APS nephropathy.

In this patient, multifocal renal infarcts likely resulted from recurrent arterial thrombosis. Chronic ischemia of renal parenchyma would activate the renin-angiotensin-aldosterone system, leading to renovascular hypertension.

The extensive systemic arterial calcification observed in this case is notable given the patient's young age. The pattern was not consistent with: Takayasu arteritis -absence of mural thickening or enhancement, CKD-medial vessel wall calcification -normal renal function, fibromuscular dysplasia - absence of beading pattern and classic premature atherosclerosis - lack of conventional risk factors. Instead, the findings are most consistent with chronic thrombotic vasculopathy. Recurrent endothelial injury, thrombosis, vascular remodelling, and intimal fibrosis may result in premature arterial aging and calcification. The diffuse arterial calcification was considered secondary to chronic thrombotic vasculopathy [8].

Diagnosis: Based on the combination of clinical presentation, imaging findings, and serological evaluation, a diagnosis of primary APS with chronic thrombotic vasculopathy was established.

Clinical criteria: The patient fulfilled the clinical criterion for APS by the presence of arterial thrombosis, evidenced by multifocal renal infarctions on CT angiography. These findings represent end-organ ischemic injury secondary to arterial thrombotic events.

Laboratory criteria: Laboratory evaluation demonstrated elevated anti- β 2 glycoprotein I IgG antibodies. Importantly, repeat testing performed after 12 weeks also remained positive for anti- β 2 glycoprotein I IgG antibodies, thereby fulfilling the requirement for persistent antiphospholipid antibody positivity as per classification criteria. Anticardiolipin antibodies and anti-dsDNA antibodies were negative.

Thus, the patient fulfilled both clinical and laboratory criteria according to the revised Sydney (Sapporo) classification criteria for APS.

In the absence of clinical or serological evidence of systemic lupus erythematosus or other autoimmune diseases, the diagnosis was consistent with primary APS.

The diffuse arterial calcification was considered secondary to chronic thrombotic vasculopathy [9].

This case highlights an under-recognized imaging phenotype of APS: diffuse medium-sized arterial calcification associated with renal infarction and secondary hypertension.

The absence of anti-dsDNA antibodies and lack of clinical lupus features suggest primary APS rather than secondary APS associated with systemic lupus erythematosus. This case highlights an unusual imaging presentation of antiphospholipid syndrome and emphasizes the importance of considering a prothrombotic etiology in young patients presenting with unexplained systemic arterial calcification and renal infarction [10].

Conclusion

Primary APS should be considered in young patients presenting with accelerated hypertension, renal infarctions, unexplained diffuse arterial calcification and positive anti- β 2 glycoprotein I antibodies.

Recognition of this imaging pattern is crucial for early diagnosis and prevention of progressive vascular injury.

References

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