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# Patient with antiphospholipid syndrome presenting with testicular torsion-like symptoms

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ABSTRACT

Testicular torsion, a urological emergency, occurs due to absence of testicular blood supply secondary to a mechanical twist of the spermatic cord. The authors describe a 28-year-old male who presented with torsion symptoms, first in the left testicle and four months later in the right testicle. Doppler ultrasound and surgical exploration revealed disruption of blood flow but no evidence of spermatic cord twisting. Additionally, physical examination findings at the time of presentation were inconsistent with testicular torsion. Hematologic workup revealed triple positive antiphospholipid syndrome as the cause of testicular ischemia. The patient was successfully treated with aspirin and therapeutic heparin.

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## 1. Introduction

Testicular torsion, a urological emergency, occurs due to twisting of the spermatic cord. Consequently, testicular blood supply is compromised, resulting in testicular infarction. The authors present a patient who experienced metachronous bilateral episodes of testicular pain and diminished arterial flow, suspicious for torsion. The patient exhibited classic testicular torsion symptoms, but scrotal exploration revealed no spermatic cord twisting after both events. After hematologic workup, the patient's presentation was attributed to microthrombi in testicular circulation secondary to antiphospholipid syndrome (APLS). Testicular infarction due to hypercoagulable states are rarely described in the literature; only two previous reports attributed the infarction to APLS.[1–5] This is the first reported case of APLS presenting with testicular infarction as its only manifestation.

## 2. Case presentation

A 28-year-old man with no past medical history presented to an outside hospital with a 3-day history of acute abdominal pain, left testicular pain, and vomiting. The patient's episode began after a weight-lifting session. An ultrasound showed no flow to the left testicle with concern for testicular torsion. The patient underwent surgical exploration and bilateral orchidopexy at that institution where no evidence of torsion was found.

Two days later, the patient presented at the authors' institution with worsening left testicular pain despite orchidopexy. Notably, the patient denied any difficulty voiding, changes in urine color, fever, flank pain, penile discharge, recent sexual activity, or testicular trauma. Physical exam revealed a tender left testicle, but no swelling or erythema of the scrotum. The patient had a preserved cremasteric reflex. Laboratory values showed a white blood cell count of 12.9 $\times$ $10^3$/cubic mL and platelet count of 259 $\times$ $10^3$/cubic mL.

Scrotal duplex ultrasound revealed normal blood flow and homogenous echotexture in the right testicle. The left testicle had no arterial flow, some preserved venous flow, and minimal heterogeneous echotexture. Scrotal exploration revealed no twisting of the left spermatic cord, but the left testicle appeared non-viable with a blue mottled appearance. After left orchiectomy, the patient's pain subsided, and he was discharged.

Four months later, the patient returned with a one-day history of acute right testicular pain. Similar to his previous presentation, his review of systems was otherwise negative and his physical exam was normal except for testicular tenderness.

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**Table 1**
Laboratory values for presentation of right testicular pain.

| Item | Value | Normal Range | Units |
|---|---|---|---|
| Platelets | 65 | 150–350 | 1000/cubic mL |
| INR | 1.4 | 0.9–1.1 | – |
| WBC | 5.23 | 4.5–11.0 | 1000/cubic mL |
| PTT Baseline | 42.5 | 22.9–30.6 | seconds |
| PTT (1hr 1:1 mixing study) | 34.5 | 22.9–30.6 | seconds |
| PTT (4hr 1:1 mixing study) | 43.1 | 22.9–30.6 | seconds |
| RVVT Confirm Ratio | 2.4 | 1.0–1.4 | – |
| Anti-Cardiolipin IgG | 34 | 0–20 | phospholipid units |
| Anti-B2 glycoprotein antibodies | 22 | 0–20 | phospholipid units |

INR = international normalized ratio; WBC = white blood cell count; PTT = partial thromboplastin time; RVVT = Russell Viper Venom Test.

**Table 2**
12 week confirmatory antibody testing.

| Item | Value | Normal Range | Units |
|---|---|---|---|
| RVVT Confirm Ratio | 1.8 | 1.0–1.4 | – |
| Anti-Cardiolipin IgG | 50 | 0–20 | phospholipid units |
| Anti-B2 glycoprotein antibodies | 24 | 0–20 | phospholipid units |

RVVT = Russell Viper Venom Test.

Ultrasound revealed minimal arterial flow to the right testicle, suggestive of testicular torsion. Upon surgical exploration, orchidopexy sutures were seen, securely attaching the right testis to the scrotum in the normal orthotopic position, without cord twisting. A weak Doppler signal was detected in the posterior and inferior aspects of the testicle. The solitary right testicle was again fixed to the dartos fascia and the patient was admitted for hematologic evaluation, with concern for a thromboembolic cause. The patient's hematologic workup is detailed in Table 1.

Intravenous heparin and aspirin was started empirically to treat for possible arterial thrombosis, and the patient experienced decreased pain. After three days, a duplex ultrasound revealed markedly improved arterial flow to the right testicle. The following day, the patient was discharged on warfarin (INR goal = 2–3) and an enoxaparin bridge. After discharge the patient has continued on anticoagulation therapy without symptom relapse. The above mentioned antibody tests in Table 1 were again positive on testing at 12 weeks (Table 2).

Lastly, final pathology from his left orchiectomy demonstrated ischemic necrosis, lymphocytic vasculitis in small arteries, and associated organizing thrombosis causing arterial occlusion.

## 3. Discussion

Antiphospholipid syndrome (APLS) is an autoimmune disorder characterized by arterial or venous thromboses and at least one positive test for an antiphospholipid antibody. APLS involves disruption of the coagulation pathway, fibrinolysis, and vascular tone, making these patients more susceptible to thrombotic events. The patient's diagnosis of APLS involved several key diagnostic steps. The patient's elevated PTT and thrombocytopenia in the setting of arterial thrombosis was suggestive of a coagulation disorder. Additionally, the patient's mixing study did not correct his PTT back to normal levels – indicating an inhibitor in the coagulation pathway. Lastly, the patient had positive testing for APLS antibodies, a highly specific metric. The patient's Russell Viper Venom Test (RVVT) – which tests for lupus anticoagulant – was greater than 1.4 (2.4). Additionally, the patient had positive testing for anti-cardiolipin and anti-β2 glycoprotein antibodies. Thus, the patient has triple-positive antibody testing for APLS.

Most commonly, thrombotic events secondary to APLS manifest as deep vein thrombosis, thrombocytopenia, or livedo reticularis. Existing literature rarely describes testicular ischemia from arterial clotting. Two reports of testicular ischemia due to coagulation have attributed the pathophysiology to protein S deficiency. These reports describe young patients who have sudden onset of testicular pain, loss of blood flow, and no twisting of the spermatic cord – similar to this current report.[2,3]

Two previous reports describe testicular ischemia secondary to an organizing thrombosis from underlying APLS. In both cases the patients had manifestations of hypercoagulability, with symptoms such as ischemic colitis, deep vein thrombosis, and necrotic skin changes, prior to their testicular pain symptoms. This present case differs in several ways. Firstly, in this report the patient experienced bilateral torsion-like symptoms. Secondly, this is the first case that describes the first presentation of APLS as a torsion-like event in the absence of other manifestations of APLS.[1,4]

This case highlights a rare, but critical complication of APLS. Patients presenting with acute testicular pain and suspected testicular torsion should be evaluated and treated promptly with scrotal exploration. In the event twisting of the spermatic cord is not found, anticoagulation therapy and work-up for thrombosis should be considered, particularly in those with a history of hypercoagulability. Chronic anticoagulation should be considered under the direction of a hematologist as patients may experience future systemic manifestations of APLS.

## 4. Conclusion

Testicular ischemia due to thrombosis is a rare, but important complication of antiphospholipid syndrome. This may present as testicular torsion-like symptoms without evidence of spermatic cord twisting on surgical exploration. Urgent anticoagulation therapy is crucial as it may help salvage an ischemic testicle and prevent future manifestations of APLS. Clinicians should consider a thrombotic event as a rare etiology for the acute scrotum.

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