CASE REPORT

# UNUSUAL INITIAL PRESENTATION OF PRIMARY ANTIPHOSPHOLIPID SYNDROME AS POSTPARTUM SYMMETRICAL PERIPHERAL GANGRENE

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**Background** Antiphospholipid syndrome (APS) is an acquired prothrombic state with recurring thromboembolic and obstetric complications in the presence of antiphospholipid antibodies. Isolated skin manifestation especially symmetrical peripheral gangrene (SPG) in postpartum phase is reported rarely. To highlight this unusual presentation of APS with SPG we present a case of young female who developed SPG on her third postpartum day. Postpartum period runs a high risk of sepsis but development of such extensive and rapid ischemic changes in APS is seen uncommonly.

**Keywords:** Antiphospholipid syndrome; Postpartum Symmetrical peripheral gangrene; thrombophilia

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## INTRODUCTION

Antiphospholipid syndrome (APS) is a well-established cause of acquired prothrombic state and is an autoimmune disorder with diverse clinical presentations ranging from recurring thrombosis to pregnancy morbidity in the existence of antiphospholipid antibodies (aPL).[1] The classification criteria of APS was updated in 2006.[2] According to the criteria APS is present if at least one of the clinical and laboratory criteria are present. Clinical criteria include; vascular thrombosis (one or more clinical episodes of arterial, venous or small vessel thrombosis in any tissue or organ) or pregnancy morbidity. Pregnancy morbidity includes; unexplained deaths of one or more morphological normal foetus at or beyond 10$^{th}$ week of gestation, premature births of one or more morphologically normal neonates before 34th week of gestation or unexplained consecutive three or more spontaneous abortions before the 10$^{th}$ week of gestation. Laboratory criteria includes presence of lupus anticoagulant (LA) / anticardiolipin (aCL) antibodies/ beta 2 glycoprotein-I antibodies in plasma on two or more occasions at least 12 weeks apart.[2] APS can be primary or secondary depending on association with other autoimmune diseases. When no evidence of identifiable cause is found it is termed as primary while secondary APS is most seen with Systemic lupus erythematosus (SLE).[3] The most common thromboembolic event outlined in APS is deep venous thrombosis (DVT) of the leg veins. Pulmonary embolism (PE) and both transient and ischemic stroke, are other thrombotic manifestations of APS. Obstetric features are the hallmark of APS in which recurrent early miscarriage, late pregnancy loss, placental insufficiency and preeclampsia are the most common presentations.[4] Symmetrical peripheral gangrene (SPG) is gangrene of peripheral extremities resulting from symmetrical digital ischemic damage without vasculitis or large vessel obstruction. Hypercoagulable state remains the chief associated disorder.[5] In SPG larger vessels are mostly spared so the peripheral pulses are usually palpable. Disseminated intravascular coagulation (DIC) has been reported in literature to be associated with 85–100% cases of SPG.[6]

We report a unique case in which a young female in her early postpartum period presented with DIC and SPG which initially mimicked as vasculitis but turned out to be primary APS.

## CASE PRESENTATION

A 25-year -old female Gravida 3 Para 3 presented to the emergency department on her third postpartum day with purple discoloration and painful superimposed watery blisters on left foot and black discoloration and purpuric rash on right foot. She had a normal vaginal delivery in hospital. There was no prior history of tobacco smoking, intermittent claudication, recurrent miscarriages, collagen vascular disease or similar family history. She was vitally unstable, running a high-grade fever (103–104 F) and was hypotensive with rapid pulse. All peripheral pulses were palpable except posterior tibial artery and dorsalis pedis artery bilaterally. Feet were cold and swollen with intact sensation. She had moderate anaemia and jaundice. Examination of the chest and abdomen were unremarkable.

On investigation her Hb was 8.7 g/dl, total leukocyte count (TLC) was 23,000/mm$^3$ and platelet count was 55,000/mm$^3$. Liver function tests (LFT) were serum bilirubin 9 mg/dl, alanine aminotransferase (ALT) and aspartate aminotransferase (AST) were 530 U/L and 343 U/L

respectively. Serum creatinine was 0.5 mg/dl and blood urea were 40 mg/dl. Total protein was 6.9 g/dL, albumin was 2.1 g/dL, and CRP was 59 mg/dL. On the work-up of sepsis, her blood and urine cultures were sterile. On coagulation screen her Prothrombin Time (PT) was 25 seconds, INR was 1.78 with Activated Partial Thromboplastin Time (APTT) 52 sec. Lipid profile was as follows; serum cholesterol 120 mg/dL, serum triglyceride 103 mg/dL, high-density lipoprotein cholesterol (HDL) 40 mg/dL, and low-density lipoprotein cholesterol (LDL) 80 mg/dL. Alkaline phosphatase (ALP) was 224 U/L, lactate dehydrogenase (LDH) was 489 U/L and rheumatoid factor (RF) was negative. The aCL and beta 2 glycoprotein 1 antibodies were negative however a LA was positive on two different occasions 12 weeks apart. Screening for lupus, rheumatoid arthritis and primary vasculitis was negative. The chest X-ray was normal. Urinalysis was also normal. The ultrasonography of the pelvis showed postpartum state of uterus without any fluid collection or retained product of conception. Colour doppler ultrasound of the lower extremity vessels revealed monophasic pattern with moderate arterial insufficiency in right dorsalis pedis artery and left dorsalis pedis artery showed no flow. The diagnosis of primary APS was established on grounds of sustained elevated levels of LA on two occasions and exclusion of other connective tissue disorders and all usual causes of distal symmetrical gangrene in postpartum period.

Medical treatment with broad spectrum antibiotics, low molecular weight heparin and cilostazol was started. By day 7, improvement was noted with resolution of swelling and appearance of sharp demarcation of margins. She was discharged on cilostazol. After 2 weeks she developed wet gangrene of left foot and left foot amputation was done while the right toes were autoamputated.

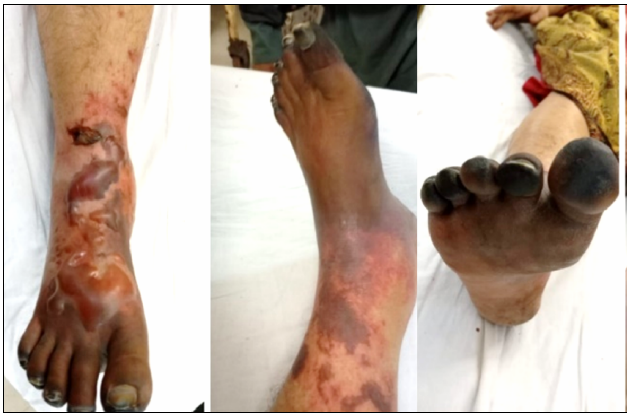

**Figure-1: Postpartum 3rd day of extremity appearance**

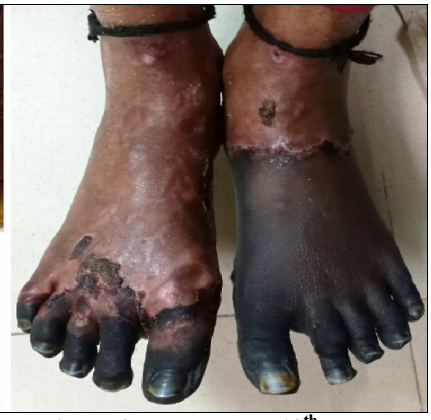

**Figure-2: Postpartum 10th day extremity appearance; dry gangrene with established line of demarcation**

## DISCUSSION

In the present case, negative workup for peripheral vascular disease, major occlusive disease, systemic vasculitis, connective tissue disorder or any other distinguishable cause for gangrene led us to explore the uncommon aetiologies like APS. APS rarely presents with skin manifestations only and when it does it presents with livedo reticularis, palmar erythema, acrocyanosis, large cutaneous necrosis, digital gangrene or ischemic ulcers. SPG as the initial presentation of primary APS has been reported very rarely in literature. The most common underlying associated disease with SPG is SLE.

A very similar case to ours was seen in a 25-year-old woman who presented one week after delivering a preterm baby with dark discoloration of the feet, hands and the tip of the nose which progressed bilaterally in a symmetrical fashion up to the wrists and just above the ankles.[7] In literature we find another case where APS was diagnosed in a 75-year-old woman who presented with SPG. She had a history of recurrent abortions and smoking and now presented with SPG in all four extremities. Workup led to the diagnosis of APS.[8]

While reviewing literature we came across two other cases where SPG was the initial presentation of APS. In one of the cases SPG was seen in a 12-year-old girl who presented to the emergency department with discoloration of arms and legs which later developed gangrene. She tested positive for APS.[9] In the other one APS was diagnosed in a 45-year-old male smoker who presented with SPG.[10]

## CONCLUSION

Although SPG can be seen in DIC (postpartum phase) but primary APS in postpartum period presenting as SPG is rarely reported in literature. The purpose of reporting this rare presentation is to highlight the importance of investigating such patients for APS so that timely intervention can be done, and patients may be prevented from catastrophic complications.

**Informed consent:** For publication of the case and pictures written informed consent was taken from the patient

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