Case Report

Atypical Presentation of Anti-phospholipid Syndrome with Triple Antibody Positive (ATAP) Syndrome Presented as Non-healing Foot Ulcer in a Middle-aged Adult Female

Amishi Nitin Rathod[1], Vishal Nitin Rathod[2], Sunil M Shahane[3]

Middle aged women presented with non-healing ulcer over foot following trivial injury. She was not having any co-morbid condition. Investigations revealed APLA Syndrome with triple antibodies positive (Lupus anticoagulant, Anticardiolipin antibody and anti B2-glycoprotein antibodies). Patient responded well with anti-platelet and skin grafting at local site.

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Key words **: Triple antibody positive.**

Case Report

39-year-old female presenting with complain of non-healing wound on the lateral aspect of the right foot for 2 months. Patient was alright 2 months ago, when she sustained minor blunt trauma to right foot resulting in mild swelling followed by small Ulcer. It increased in size over 2 months period. There was no history of motor or sensory system disturbance in lower extremities, no history of bleeding or discharge from the wound. There is no Hyperpigmentation/ Varicose Vein or raised temperature of the skin around the wound There was no history of skin rash, oral ulceration, photosensitivity, or bleeding from any other site. Patient did not give any history of co-morbid condition or Collagen Vascular Disorder /Inflammatory Bowel Disease. She does not give history of significant weight loss. She refused any history of medication taken for illness. she is non-smoker and non-alcoholic. Patient underwent debridement of the wound 1 month ago and wound did not heal even after 2 months of proper dressing, antibiotics and anti-inflammatory drugs (Figs 1&2).

Examinations — On examination, patient hemodynamically stable. All the peripheral pulses were palpable. One large Ulcer over lateral malleolus and another small Ulcer adjacent to it were present without any changes of the surrounding skin. The Ulcers were oval in shape with size of 9x4 cms and 2x1 cms respectively. The ulcer had an indurated margin with Floor of the wound covered with slough. There was no active discharge. Abdominal examination revealed no splenomegaly or hepatomegaly. Central nervous system did not reveal any sensory or motor deficit or Peripheral Nerve thickening or tenderness.

Editor's Comment :

- Middle age female presenting as a non healing ulcer after ruling out common causes like Vasculitis and Pyoderma Gangrenosa, Antiphospholipid Antibody Syndrome should be considered.

Investigations — The hemogram showed Hemoglobin of 9.4g/dL, a Leucocyte Count of 9030/L, Platelet count: 169X 10^9/L. The Random Blood Sugar was 111mg/dL. The Blood Urea Nitrogen was 38mg/dL and serum Creatinine 1.24mg/dL. The serum Sodium-138.0 mEq/L, Potassium - 3.85 mEq/L, Chloride-101.8 mEq/L. The Prothrombin time 14seconds and INR were 1.35 seconds. The HBsAg, HCV and HIV were negative. The CRP- 3.74 mg/L and ESR-75 mm respectively. Pus culture grown pseudomonas aeruginosa of non-significance. No fungal elements or acid-fast bacilli seen.

The Anti CCP Antibodies were negative (2.00 U/ML). The sickling test was also negative The Anti-Nuclear Antibody (ANA) with titer (1:1000) was positive with a homogeneous pattern (Immunofluorescence method) cytoplasmic positivity seen. THE C-ANCA was Negative but P-ANCA was Positive. Based on the clinical profile of the patient with investigations, diagnosis of Vasculitis Ulcer was made. Color Doppler of bilateral lower limbs and abdomen was normal with no evidence of arterial blockage or Varicose Vein. The lupus anticoagulant, anti cardiolipin antibodies IgG 97U/ml (positive >40), anti-beta-2 glycoprotein of IgG 124.46 RU/ml (positive >20) was positive. Biopsy from the edge of the wound revealed granulation tissue with infiltration of Lymphocytes, Plasma cells and few Neutrophils, no granulomas or fungal elements seen. Histopathological diagnosis was Necrotic Foot Ulcer.

Considering clinical presentation along with specialized investigations, non-healing ulcer with APLA Syndrome diagnosed. Patient was given anti-platelet

[1]MBBS, Department of Medicine, Dr D Y Patil Medical College, Mumbai, Maharashtra 411018 and Corresponding Author

[2]MS, Resident, Department of Orthopaedics, Bombay Hospital & Medical Research Centre, Mumbai, Maharashtra 400020

[3]MS, Head, Department of Orthopaedics, Nanavati Max Super Specality Hospital, Mumbai, Maharashtra 400056

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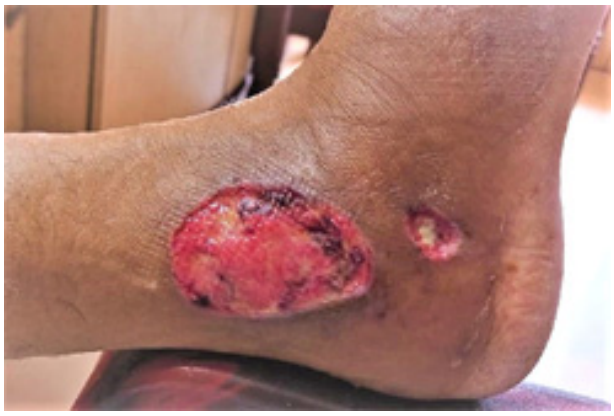

Fig 1 — Non healing ulcer over Right foot.

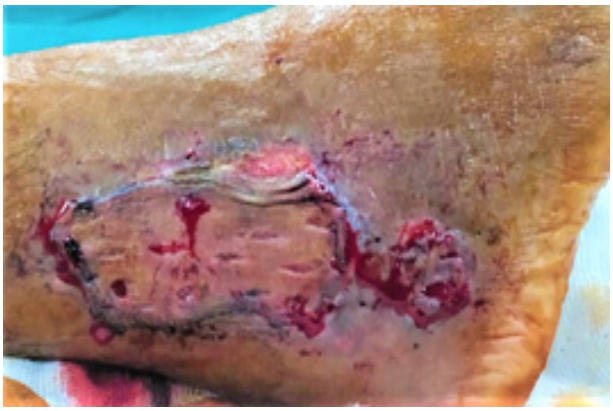

Fig 2 — Non healing ulcer after skin grafting

and skin grafting done. Patient improved and discharged from hospital.

Discussion

Antiphospholipid Syndrome is Autoimmune Disorder, resulting due to autoantibodies against anticardiolipin and lupus anticoagulant present on the Plasma membrane causing a hypercoagulable state[1]. Although the exact etiology of APS is still not clear, genetics is believed to play a key role in the development of the disease, Genetic Markers: HLA-DR4, HLA-DR7 and HLA-DRw53 It is more common in women than in men[2]. Clinically important anti-phospholipid antibodies are associated with Thrombosis and Vascular disease. In pregnant women affected by APS, there is an increased risk of miscarriage and intrauterine growth retardation[3]. The Anti-phospholipid Syndrome responsible for most of the miscarriages in later trimesters. It is estimated that the incidence of APS is approximately 5 cases per 100,000 persons per year and the prevalence is approximately 40-50 cases per 100,000 persons.

It is very rare to have APLA Syndrome with non-healing ulcer as present in our case report. Treatment includes wound care, pain management, Anti-platelet agents, Blood thinner if major artery or venous blockage, warfarin is used, the INR is kept between 2.0 and 3.0[4] in case of triple positive instead of warfarin directly acting oral anticoagulant are used[5] and skin grafting for non-healing ulcer. In refractory cases Plasmapheresis may be considered.

Conclusion

Middle aged women present with evidence of arterial, venous blockage or repeated miscarriage and occasionally present with non-healing ulcer then Antiphospholipid antibody (APLA) Syndrome should be suspected.

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