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Case Report

An Unusual Thrombotic Event in Systemic Sclerosis: A Case Report

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Abstract

Introduction: The Authors described a clinical case characterized by an unusual thrombotic event in patient with Systemic Sclerosis.

Clinical findings: We described a clinical case of a younger male with SSc who had a deep vein thrombosis of subclavian vein, axillary vein, humeral vein of the right upper limb in absence of risk factors that could justify this very difficult event.

Diagnosis, therapeutic interventions, outcomes: CT angiography confirmed succlavian vein thrombosis and echo doppler ultrasound study of the right arm which showed thrombosis of the right succlavian vein, thrombosis of axillary vein and thrombosis of humeral vein. The patient underwent therapy with edoxaban, after on month of therapy, he has not yet a complete resolution.

Conclusion: We presented a clinical case of SSc complicated by a venous thrombosis in patient whitout comorbidities treated with NAO (new oral anticoagulant).

Introduction

Systemic Sclerosis (SSc) is an autoimmune, systemic, chronic disease characterized by an immune dysregulation, vascular damage that occurs with Raynaud phenomenon, pulmonary hypertension and renal crisis; it also important is the fibrosis of the skin and internal organs (1). Patients with SSc may be at an increased risk for venous thromboembolism (VTE) including pulmonary embolism (PE) and deep vein thrombosis (DVT). Generally, the reported frequency of various thrombotic events (THEs) is low in SSc, however the same risk for THEs is still higher than in non-SSc individuals (2).

We reported a clinical case of a younger male with SSc who had a deep vein thrombosis of subclavian vein, axillary vein, humeral vein of the right upper limb in absence of risk factors that could justify this very difficult event.
This clinical case is really unique! In first, no comorbidity was found to justify the thrombotic event, secondly no coagulation deficts were found and third the patient was poorly responsive to therapy.

Below we will explain the detailed description of the case and the results that have emerged.

Patient presentation

A 33-year-old male patient with SSC was admitted to our Rheumatology Department for massive edema on the right arm, which, according to the patient, occurred in good health.
Diagnosis of Systemic Sclerosis was made at the age of 17 years according to the American College of Rheumatology criteria. At the onset of the disease, the patient had edema of both hands, accompained by pain and ulcers in the digits. Patient presented, also, arthritis of wrists, hands, shoulders, knees. Diagnosis of Systemic Sclerosis with Rheumatoid Arthritis (RA) in overlap was made and was prescribed therapy with aspirin 75 mg/day, amlodipine 5 mg/day and methotrexate 7.5 mg/week. Patient discontinued the therapy with methotrexate from severe gastrointestinal effects and he received therapy with tocilizumab at dosage of 162 mg/week; this drug led to an immediate benefit characterized by a significant improvement of joint involvement still well controlled.

Clinical Findings

If we consider the recent clinical event represented by edema of right arm, on inspection, findings were diffuse skin thickening and edema of right arm in presence of normal arterial radial pulse.

Diagnostic Assessment

The patient performed Doppler ultrasound diagnostic arterial and venous of the right arm that did not show deep vein thrombosis and arterial thromboembolism. Nonetheless, the patient had right arm pain and he started therapy with mild anti-inflammatory drugs without improving symptoms. Few days later, he started therapy with enoxaparin at dosage of 6000 U.I/die for fifteen days and CT angiography of lung and right arm was planned.

The laboratory evaluation: white blood cells 7.49 K/ml (micro) (normal 4.00- 10.00) with 74.3 %of neutrophils (normal: 55-70%). Haemoglobin value was 14.5 g/dl (normal: 13-17 g/dl), platelet 287 K/ ml (micro) (normal 150-400 10*3/mm*3), alanine transferase (ALT) 29 U/l (normal 0-40), aspartate transferase (AST) 26, C - reactive protein at 0,2 mg/l, Von Willebrand factor Antigen was 141, 20% (normal value 100-170%); Lupus Anticoagulant (LAC) 1,40; IL-6 75,28 pg/ml (1.5-7), Protein C anticoagulant 82% (70-140); normal values for IgM and IgG anticardiolipin and B2 GPI IgM and IgG.

The immune profile, including rheumatoid factor, anti-cyclic citrullinated antibody, anti-La, anti-Ro, perinuclear antineutrophil cytoplasmic antibody and antineutrophil cytoplasmic antibody were negative. The antinuclear antibody (ANA) was positive at 1/1280/ with a speckled pattern, and antitopoisomerase 1 (anti-Scl 70) was positive by immunoblotting assay. No coagulation factors deficits were found.

After ten days, symptoms were long lasting with ongoing heparin treatment; the patient performed a second echo doppler ultrasound study of the right arm which showed thrombosis of the right succlavian vein, thrombosis of axillary vein and thrombosis of humeral vein. The patient complained of chest pain and he performed CT angiography of the lung with negative result for pulmonary embolism.

However, CT angiography confirmed succlavian vein thrombosis (fig 1, fig. 2).

Therapeutic Intervention

The patient started therapy with edoxaban at dosage of 60 mg/day, ongoing therapy.

Follow-up and Outcomes

After one month of therapy there is complete resolution of axillary and humeral vein but there is still a thrombosis of succlavian vein.

Discussion

Systemic Sclerosis includes inflammation followed by severe vascular lesions of small arteries; mainly venous thrombosis occurs less in patients with scleroderma than in healthy population.

Thrombotic and vascular events may occur in patients with underlying various comorbidity as well as Antiphospholipid antibody syndrome associated, cardiovascular disease and /or coagulation factors deficits (3). However, and this mechanism is clearer in pulmonary arterial hypertension, we think that in this clinical case a thrombosis in situ has occurred (4).

The choice of edoxaban 60 mg once a day was based on a decision to block a Xa factor who is a pivotal component of coagulation cascade. Edoxaban was preferred for antiplatelet and endothelial effects, is very safer, more effective, more convenient than warfarin Systemic

Sclerosis patients (5). This drug is very safer for our young patient, is particularly advantageous in our patient who has difficult venous access due to skin fibrosis and subcutaneous joint contractures and do not require routine international normalised ratio (INR) monitoring. Finally, the patient has gut hypomotility which may affect warfarin and vitamin K absorption, resulting in unstable INRs (6).

Patient Perspective

The patient has well tolerated therapy but is still dissatisfied for the poor results achieved.

Consent Section

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Highlights case

We described a clinical case of a younger male with SSc who had a deep vein thrombosis of subclavian vein, axillary vein, humeral vein of the right upper limb in absence of risk factors that could justify this very difficult event.
The patient tolerated therapy with edoxaban, after on month of therapy, he has not yet a complete resolution.
We presented a clinical case of SSc complicated by a venous thrombosis in patient whitout comorbidities treated with NAO (new oral anticoagulant).

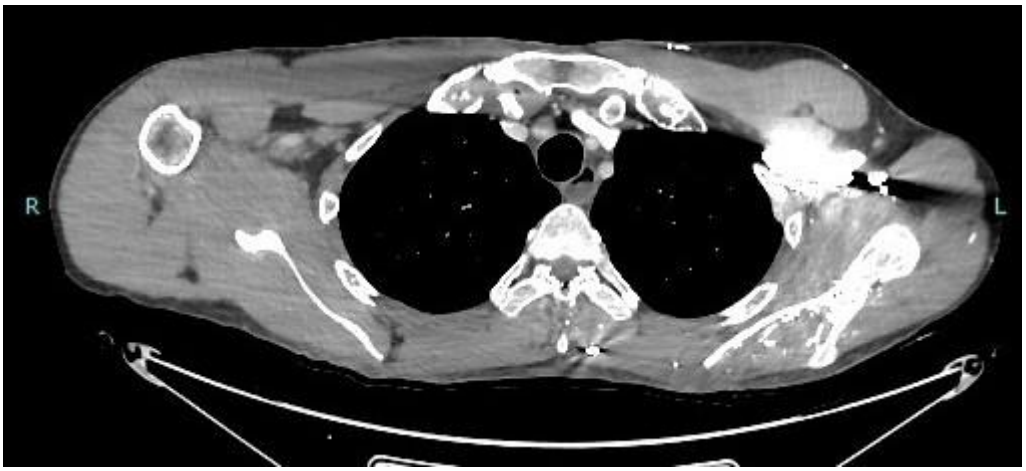

FIG 1 shows chest angio-CT. CT axial.
MPR volumetric reconstruction on the coronal plane

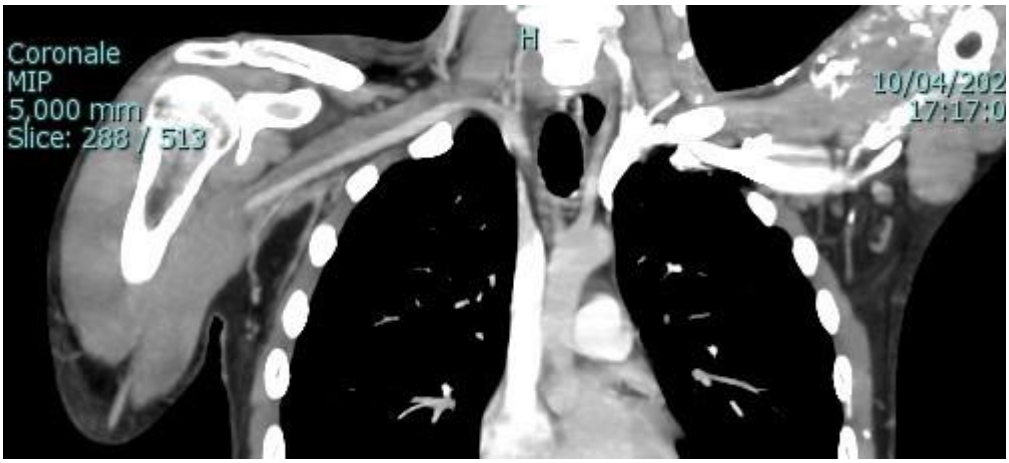

FIG 2
Failure to display the axillary vein and right subclavian until the confluence with the jugular vein.

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