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Spontaneous Internal Jugular Vein Thrombosis as Initial Presentation of Systemic Lupus Erythematosus that is Associated with Antiphospholipid Syndrome: Case Report

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Abstract: ***Spontaneous internal jugular vein thrombosis is an uncommon entity. A 24-year-old Saudi male presented with painful neck mass. Doppler ultrasonographic examination revealed evidence of thrombosis involving the right internal jugular vein. The patient had no central venous catheter, oropharyngeal infection, or cancer Internal jugular vein thrombophlebitis is rare but can be life-threatening if not recognized and treated early. Primary care physicians should have a high index of suspicion for internal jugular vein thrombosis (IJVT) in any patient presenting with painful neck swelling.***

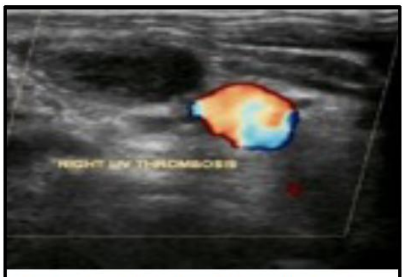
1. Introduction

Venous thrombosis is one of the most important manifestations of antiphospholipid syndrome (APS)(1). Unusual sites of venous thrombosis such as jugular vein, inferior vena cava and mesenteric veins were reported secondary to antiphospholipid syndrome (2).Internal jugular vein (IJV) thrombophlebitis, a rare entity, usually occurred after venous catheter, malignancy or an oropharyngeal infection and often led to septicemia and pulmonary embolism (1).This article present a case of spontaneous IJV thrombophlebitis associated with systemic lupus erythematosus.

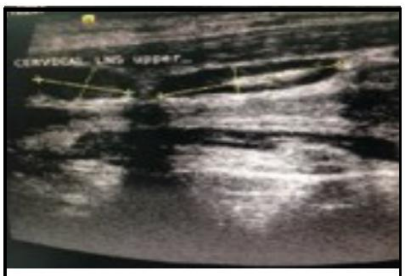
To the best of our knowledge, only three cases in the literature had been previously reported IJV thrombosis as a primary presentation of APS (13, 14, 15).

2. Case Presentation

A 24-year-old Saudi male hadfever and a painful mass on the right side of his neck for 5 days but no recent trauma, venous-catheter insertion in the neck, or oropharyngeal infection. No history of contact with TB patients. No history of joint pain or skin rash. Physical examination showed T 37.7C°, pulse rate 89/min, blood pressure121/ 74 mm/Hg, respiratory rate 20/min. Ear and oral cavity examination were normal. Examination of his neck showed painful, soft, immobile right sided neck mass along the anterior border of the sternocleidomastoid muscle without hyperemia or local hotness. There were cervical, bilateral apical axillary and inguinal lymphadenopathy, non-tender, firm, regular surface. On laboratory examination, CBC showed Hb 12.5 gm/dl, WBC count 8.5/ mm. Renal and liver function tests were normal. Prothrombin and activated partial thromboplastin times were prolonged and not correct with mixing studies. Chest X-ray at presentation was normal. Neck Doppler ultrasound showed evidence of thrombosis involving right internal jugular vein that extends to the right subclavian vein, lymph nodes in supraclavicular and cervical areas, with intact fatty hilum (Figs. 1 and 2).



figure(1) right internal jugular vein thrombosis extending to right subclavian vein.



figure(2) cervical lymph node.

CT neck & chest with IV contrast showed filling defects in the right internal jugular vein consistent with thrombosis, supraclavicular lymph node seen in the right side, no pulmonary nodularity, no evidence of mediastinal lymphadenopathy.Evectional biopsy showed a reactive lymph node (non-specific lymphoid hyperplasia) No evidence of lymphoid neoplasm as demonstrated by CD3, CD5, CD20, CD23 & CD30.

Lupus anticoagulant, anti-cardiolipin IgM (ELISA), anti-beta -2 glycoprotein 1 antibodies IgM (ELISA) were positive on two occasion 12 weeks apart. Patient was started on LMWH and warfarin for thrombosis.

One month later during follow up, patient developed thrombocytopenia and symmetrical arthritis; involving

small joints of the hands and knees, and he was evaluated for autoimmune disease, ANA, anti dsDNA was sent and came back positive. Patient was diagnosed as antiphospholipid syndrome secondary to SLE based on American college of rheumatology diagnostic criteria (4).

3. Discussion

In 1986, antiphospholipid syndrome was initially reported by Hughes, Harris and Gharavi (5). APS is still one of the most common identifiable acquired thrombophilicstates that is characterized by elevated titers of antiphospholipid antibodies, namely, the lupus anticoagulant, anticardiolipin antibodies and/or Anti beta -2 glycoprotein 1 antibodies at least in two occasions 12 weeks apart. (6) The literature revealed the presence of lupus anticoagulant and anticardiolipin antibodies in about 1% to 5 % of young healthy control subjects (7). Venous and arterial thrombosis in APS range from 0.5% to 30% (7). The prevalence of APS in women is five times greater than men as reported by the multicenter Euro-Phospholipid Project (7).

The antiphospholipid syndrome in SLE patients associated with the evidence of an antiphospholipid antibody in the serum that has been reported as a triad of thrombosis either venous or arterial, recurrent fetal loss and thrombocytopenia (8). Increased titer levels of antiphospholipid antibodies in SLE patient usually correlate with clinical manifestations. Primary antiphospholipid syndrome is defined as absence of clinical or serological evidence of SLE (9). The prevalence of thrombosis associated with antiphospholipid antibodies in patients with systemic lupus erythematosus is higher with lupus anticoagulants (15% - 34%) compared with anticardiolipin antibodies (12%-30%) as reported in previous studies (10).

In 1912, Internal jugular vein thrombosis was initially reported as a complication of peritonsillar abscess. The classic triad behind intravascular thrombosis formation includes blood vessel trauma, stasis of blood flow, and a hypercoagulable state.

Idiopathic IJVT requires more in-depth investigations to look for thrombophilia disorders or malignancy. IJV thrombosis most commonly presents with painful swelling of the neck (11). The clinical symptoms in some occasions were misleading or absent. Reported complications of IJVT include pulmonary embolism, intracranial venous thrombosis, elevated intracranial pressure, septic emboli, facial edema and loss of vision (11).

The recommended Radiological studies to diagnose IJVT include ultrasound, contrasted computed tomography (CT) scan, magnetic resonance imaging (MRI), nuclear medicine scan and contrast venogram.

Treatment of most patients with thrombotic complications is similar to that of patients with non–APS-related thrombosis, with heparin therapy followed by oral anticoagulation (7). Treatment of the underlying cause or autoimmune disease is also required. APS with history of previous thrombosis requires long term anticoagulation with warfarin. Recent evidence suggests that the target INR 2.5-3.5 is required for secondary prevention of thrombosis in this condition (12).

4. Conclusion

Internal jugular thrombosis is an uncommon presentation of systemic lupus erythematosus that is associated with APS (2). Doppler ultrasonography has been advised to be the initial diagnostic test for any patient with neck swelling as it is non-invasive and readily available in an emergency setting. Primary care physicians should have a high index of suspicion for internal jugular vein thrombosis in any patient presented with painful neck swelling.

List of Abbreviations

APS: antiphospholipid syndrome
IJVT: Internal Jugular Vein Thrombosis
SLE: Systemic Lupus Erythromatosis
LMWH: Low Molecular Weight Heparin
CT: Computed Tomography
MRI: Magnetic Resonance Imaging
VTE: Venous Thromboembolism
TB: Tuberculosis

Competing Interests

The authors declare that they have no competing interests.

Authors' Contributions

All authors read and approved the final manuscript.

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