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Case Report

Jugular Vein Thrombosis: The Initial Manifestation of Systemic Lupus Erythematosus with Hyperthyroidism: A Case Report

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Abstract

Background: Systemic Lupus Erythematosus (SLE) is an autoimmune disease that typically presents with arthritis, rash, mouth ulcers, hair loss, and kidney damage. Although rare, the formation of Deep Vein Thrombosis (DVT) can also be a manifestation of SLE, which can lead to misdiagnosis and mistreatment.

Case summary: A 21-year-old female patient presented with persistent swelling and pain that had begun 20 d prior on the right side of her neck. An ultrasound revealed thrombosis of the right internal jugular vein. Despite treatment, her symptoms did not improve but instead worsened. Three days prior, the patient developed chest tightness and dyspnea, prompting her to seek medical attention at our hospital. Upon physical examination, she presented bilateral proptosis, bilateral neck swelling and tenderness, particularly on the right side, thickened breath sounds in both lungs, and scattered wet rales in both lower lungs. Following admission, the patient's condition rapidly deteriorated, with the onset of fever, panic attacks, and diarrhea. The patient was diagnosed with SLE, pulmonary artery embolism, jugular vein thrombosis, and hyperthyroidism based on laboratory tests, imaging tests, and genetic test results, as well as her symptoms and signs.

Conclusion: We present the case of a patient who was diagnosed with SLE after presenting with jugular vein thrombosis. We provide a summary of the patient's diagnostic and treatment history and analyze the risk factors associated with DVT in SLE patients to enhance the understanding of DVT occurrence in this population.

Keywords: Systemic lupus erythematosus; Jugular vein thrombosis; Deep vein thrombosis; Hyperthyroidism

Introduction

Systemic Lupus Erythematosus (SLE) is an autoimmune disease that affects multiple systems and organs and is most common in young women. Patients with SLE produce a significant number of autoantibodies that attack their own cells, tissues, blood components, and vascular tissues. This results in decreased blood counts, arterial stenosis, and Deep Vein Thrombosis (DVT). Here, we present the case of a patient with jugular vein thrombosis as the initial manifestation of a disease that was ultimately diagnosed as SLE. We provide a summary of the patient's diagnostic and treatment history and analyze the risk factors associated with DVT in SLE patients to enhance the understanding of DVT occurrence in this population.

In conclusion, when encountering patients, especially young women, who exhibit DVT without obvious causes, it is necessary to investigate whether they have autoimmune diseases such as SLE to avoid delaying diagnosis of the disease.

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Case Presentation

Chief complaints

A 21-year-old female patient presented with a chief complaint of "swelling and pain on the right side of the neck for 20 d, aggravated by chest tightness and shortness of breath for 3 d".

History of present illness

A 21-year-old female patient presented with persistent swelling and pain that had begun 20 d prior on the right side of her neck. Ultrasound revealed thrombosis of her right internal jugular vein, which was unsuccessfully treated with oral rivaroxaban. Three days prior, her symptoms had worsened, and she had experienced chest tightness and dyspnea, which prompted her consultation at our hospital.

History of past illness

The patient had no history of specific diseases and was in good health.

Personal and family history

The patient's personal and family history was unremarkable.

Physical examination

On physical examination, the vital signs were as follows: Body temperature, 36.3°C; blood pressure 140/93 mmHg; heart rate 132 beats/min; respiratory rate 25 breaths/min. The patient presented with bilateral protruding eyeballs, bilateral neck swelling, and localized pressure pain, predominantly on the right side. Her bilateral lung respiratory sounds were thickened, and scattered wet rales could be heard in both lower lungs.

Laboratory examinations

Blood parameters were as follows: Total leukocyte count, 13.45 × 10^9/L; neutrophil percentage, 0.89; erythrocyte count, 3.37 × 10^{12}/L; hemoglobin 76 g/L; platelet count, 54 × 10^9/L; and high-sensitivity C-reactive protein 37.596 mg/L. The coagulation parameters included a prothrombin time of 15.9 s, a prothrombin time activity of 56.8%, a prothrombin time ratio of 1.36. 1.36, 2.02 μg/mL D-D polymer, and 5.79 μg/mL fibrinogen degradation product. Anti-cardiolipin antibody, protein C and protein S were negative. Immunologic tests were positive for antinuclear antibodies. Immunoglobulin: Complement C3 (0.34 g/L) and complement C4 (0.03 g/L). The genetic test results suggested positivity for lupus anticoagulant substances.

Imaging examinations

Thyroid ultrasound revealed diffuse thyroid lesions. computed tomography of the neck veins and pulmonary arteries suggested bilateral internal jugular vein thrombosis and bilateral pulmonary artery branch thrombosis.

Final diagnosis

The patient was eventually diagnosed with SLE.

Treatment

Methylprednisolone sodium succinate 80 mg IV infusion Q12h was administered, and the patient was discharged after 2 wk of treatment with improvement of symptoms. Outside the hospital, oral methylprednisolone tablets 32 mg qd, oral farfarin sodium tablets 3.75 mg qd and methimazole tablets 10 mg qd were continued.

Outcome and follow-up

The patient's internal jugular vein thrombosis and bilateral pulmonary artery branch thrombosis resolved within 3 months. She was followed up by telephone for one year and remained in stable condition with no complaints of particular discomfort.

Discussion

SLE is an autoimmune disease that affects multiple systems and organs and is most common in young women. SLE patients produce a large number of autoantibodies in their body, which attack their own cells and tissues, and blood components; consequently, vascular tissues often become the target organs for attack by autoantibodies, which cause damage, resulting in hematopoiesis, arterial stenosis, and DVT [1-3]. It has been reported in the literature that thrombotic events, mainly lower-extremity DVT, occur in 10% to 20% of SLE patients; the present patient had a rare case of jugular vein thrombosis as the first manifestation, and similar cases have only been reported in a small number of the available studies [4,5]. SLE is gradually becoming one of the common risk factors for DVT, and the mechanisms may be as follows:

Vascular endothelial damage, the secretion of procoagulant factors by inflammatory cells, hypoproteinemia, hyperglobulinemia, and organ damage due to the disease activity itself in patients with SLE are all risk factors for DVT. These risk factors are closely related to the triad of thrombosis proposed by Virchow [6], including stagnant blood flow, damaged vascular endothelia, and a hypercoagulable state of the blood, which contribute to DVT in patients with SLE.

As one of the most common clinical manifestations of SLE, lupus nephritis is also one of the major risk factors for DVT in patients with SLE. The hypercoagulable state of the blood results from the loss of large amounts of anticoagulant-related substances (including coagulation regulators such as antithrombin III, protein C, protein S, and fibrinogen) through the kidneys; in addition, excessive activation of the renin-angiotensin-aldosterone system secondary to hypertension resulting in endothelial injury and tissue and organ edema and causing impaired venous return can lead to an increased risk of DVT; and last, patients with SLE who have significant renal pathology tend to have a greater degree of disease activity and a significantly increased risk of DVT [7].

Antiphospholipid syndrome, often secondary to SLE, is the most common cause of acquired thrombosis, accounting for 15%-20% of all DVT episodes. Antiphospholipid Antibodies (APAs), the hallmark antibodies of antiphospholipid syndrome, are a group of autoantibodies with phospholipids and/or phospholipid-binding proteins as antigens, including anti-beta2-GPI antibodies (aβ2-GPI), anticardiolipin antibodies (aCLa), and Lupus Anticoagulant (LA), among others [2]. APAs can both directly damage vascular endothelial cells and inhibit the release of fibrinogen activators, as well as prolong the phospholipid-dependent clotting time, contributing to a hypercoagulable state in the blood [8]. aCL binds to phospholipids *via* vascular endothelial cells, leading to impaired vasodilation and allowing platelet aggregation and thrombus formation, while exposed vascular collagen triggers the endogenous coagulation system. LA is an important predictor of thrombosis risk, and lupus anticoagulant-positive patients have a greater risk of thrombosis when accompanied by aβ2-GPI positivity. The thrombosis of the bilateral internal jugular vein and bilateral pulmonary artery branches in our patient was closely related to endothelial damage to blood vessels and the hypercoagulable state of blood due to SLE (Figure 1).

Autoantibodies produced during SLE can affect various organs, including the thyroid. Liu et al. [9] reported that the incidence of hyperthyroidism in 2796 SLE patients who were followed for an average of 10.5 years was 6.4%. Coagulation and fibrinolytic dysfunction in patients with hyperthyroidism have been recognized, and cases of hyperthyroidism with DVT have been reported, which may be related to the increased concentration of procoagulant factors caused by excess serum free thyroxine in patients with hyperthyroidism, which leads to hypercoagulation and a weakened fibrinolytic system, increasing the risk of DVT [10,11]. In this case, SLE combined with hyperthyroidism and multiple branch thrombosis of the bilateral jugular veins and bilateral pulmonary arteries were closely related to SLE disease activity and APAs, but the hypercoagulable state of the blood due to hyperthyroidism should not be ignored.

Conclusion

In conclusion, SLE patients are prone to misdiagnosis and underdiagnosis due to their autoimmune response, which can lead to the involvement of different organ systems, with wide variations in symptomatic manifestations, especially in the early stages of the disease. In clinical practice, when encountering patients, especially young female patients, presenting DVT without an obvious cause, it is necessary to exclude autoimmune diseases such as SLE to avoid delaying diagnosis of the disease.

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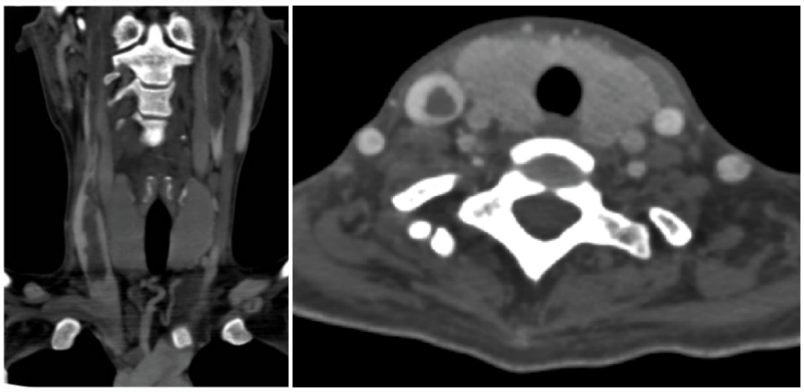

Figure 1: Neck CTA showed right jugular vein thickening and bilateral jugular vein thrombosis.

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