Is Syphilis a Potential New Factor of the POEMS Syndrome?

Jun-Jie Yin, Ai-Min Wu, Zhi-Feng Mao, Zheng-Qi Lu, Xue-Qiang Hu

Department of Neurology, Third Affiliated Hospital of Sun-Yat-Sen University, Guangzhou, Guangdong 510630, China

To the Editor: POEMS syndrome is a rare paraneoplastic syndrome due to an underlying plasma cell disorder.^[1] Herein, we report a rare case of POEMS syndrome, which initially manifested as syphilis and stroke during the course.

A 56-year-old man with a 9-year history of progressive polyneuropathy was admitted to hospital, for feeling worse accompanied with chest discomfort and dyspnea in recent 1 month. In 2005, the patient started complaining of his limbs weakness and numbness then progressed to unable to walk. In 2006, the patient was diagnosed as chronic inflammatory demyelinating polyneuropathy (CIDP) by the local hospital and given methylprednisolone therapy, then long-term oral course of prednisone and azathioprine. However, the effect was unsatisfied. In 2010, he was referred to our hospital due to worsen symptoms.

He reported a history of unclear sexual behavior when he was young. Blood routine test show white blood cell $17 \times 10^9/L$, platelet count (PLT) $363 \times 10^9/L$, and cerebrospinal fluid (CSF) protein 0.69 g/L were slightly elevated. Thyroid hormones untested. Serum treponema pallidum particle agglutination test positive, other syphilis relative tests such as serum toluidine red unheated serum test, rapid plasma reagin (RPR), Venereal Disease Research Laboratory (VDRL), and CSF RPR, VDRL tests were all negative. Nerve conduction study results demonstrated diffuse and markedly reduced sensory and motor nerve conduction velocities, and prolonged F-wave responses. Other infectious correlation test such as Epstein–Barr virus, cytomegalovirus, varicella zoster virus, hepatitis B virus, hepatitis C virus, HIV were negative. Antinuclear antibodies, anti-dsDNA anti-body etc., showed normal. We

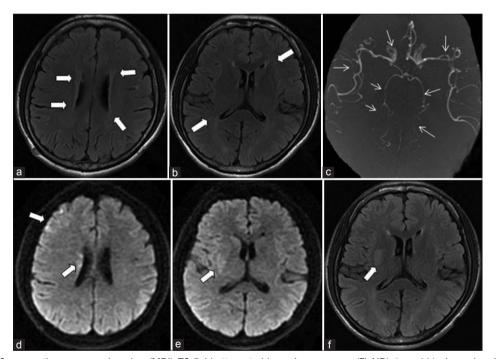


Figure 1: In 2010, magnetic resonance imaging (MRI) T2-fluid attenuated inversion recovery (FLAIR) (a and b) showed multi-focal ischemia in bilateral cerebral, the peripheral of anterior and posterior horns of the lateral ventricles (arrows). Magnetic resonance angiogram (c) showed intracranial artery multi-segment stenosis (arrows). In 2013, MRI diffusion-weighted imaging (d and e) showed acute infarction in right-side frontal lobe, area of corona radiata and basal ganglion (arrows). Compared with that in 2010, MRI T2-FLAIR (f) showed new infarction focus in the right-side basal ganglion area (arrow).

Access this article online	
Quick Response Code:	Website: www.cmj.org
	DOI: 10.4103/0366-6999.159364

Address for correspondence: Dr. Xue-Qiang Hu,
Department of Neurology, Third Affiliated Hospital of
Sun-Yat-Sen University, Guangzhou, Guangdong 510630, China
E-Mail: huxueqiangzssy@qq.com

re-administered methylprednisolone with penicillin additionally, following which his symptoms substantially improved, consistent with the probable diagnosis of neurosyphilis. [2] In 2013, he was hospitalized because of left paralysis. Brain magnetic resonance imaging (MRI) indicated acute cerebral infarction [Figure 1d-f] compared with brain MRI and magnetic resonance angiogram in 2010 [Figure 1a-c]. In 2014, his symptom deteriorated with deeply chest discomfort, chest pain, and short of breath. Our examination revealed hypermyotonia accompanied with hyperpigmentation of the limbs. Laboratory results showed elevated thyroid-stimulating hormone and reduced FT3, FT4, accompanied with intractable hypoproteinemia and proteinuria. Heart ultrasonic examination (serious pulmonary hypertension), abdominal ultrasonic (hepatosplenomegaly and slight ascites), chest computed tomography scan revealed (1) bilateral pulmonary emphysema, (2) slight bilateral pleural and pericardial effusion, and enlarged mediastinal lymph nodes, (3) bilateral gynecomastia; serum IgG λ chain level was increased, those, together highly indicated that he got POEMS syndrome.

Case reports and retrospective studies suggested that infections may alter the clinical presentation and natural history of POEMS syndrome. [3,4] However, few studies on POEMS syndrome overlap with syphilis infection had been reported before. The current case indicated the syphilis and POEMS syndrome may be related. We speculated that syphilis infection in the pathogenesis of POEMS syndrome most likely activated a series postinfectious immune-mediated condition and ultimately led to hematologic malignancy.

This case also demonstrated the need to consider the possibility of some syphilis and POEMS syndrome associated disorders (such as neurosyphilis/CIDP, and syphilis associated stroke/POEMS syndrome associated stroke) may overlap. Failure to recognize this potential relationship may result in either inappropriate therapy or omission of further therapy. Future immunological and pathological studies of overlapping syphilis and POEMS syndrome may help clarify the mechanisms underlying these potentially overlapping and related disorders.

REFERENCES

- Shi X, Hu S, Yu X, Zhuang Q, Luo M, Jiang Q, et al. Clinicopathologic analysis of POEMS syndrome and related diseases. Clin Lymphoma Myeloma Leuk 2015;15:e15-21.
- 2. Berger JR, Dean D. Neurosyphilis. Handb Clin Neurol 2014;121:1461-72.
- Kim DE, Kim HJ, Kim YA, Lee KW. Kaposi's sarcoma herpesvirus-associated Castleman's disease with POEMS syndrome. Muscle Nerve 2000;23:436-9.
- Yamac K, Senol E, Ataoglu O, Fen T. A case report of multicentric Castleman's disease with simultaneous Epstein-Barr virus infection. Tumori 2001;87:343-5.

Received: 11-03-2015 Edited by: Yuan-Yuan Ji

How to cite this article: Yin JJ, Wu AM, Mao ZF, Lu ZQ, Hu XQ. Is Syphilis a Potential New Factor of the POEMS Syndrome?. Chin Med J 2015;128:1834-5.

Source of Support: Nil. Conflict of Interest: None declared.