Vasculitic Neuropathy in a Third Trimester Pregnant Patient with Systemic Lupus Erythematosus

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ABSTRACT

Systemic lupus erythematosus (SLE) is an inflammatory, autoimmune disease that predominantly occurs in women of childbearing age. A 20-year-old, 35 week pregnant patient was admitted to our clinic with a 20 day history of leg pain, numbness in the feet, and left foot drop. On further examination, the patient reported oral ulcers once a month, occasional joint swelling, increasing malar rash under sunlight. Sensory-predominant sensorimotor polyneuropathy was detected on EMG, and a diagnosis of vasculitic neuropathy-related SLE was made. SLE patients with vague signs and symptoms may have new neurologic signs during pregnancy due to relapse of systemic disease. Moreover, there can be an increase in both fetal and maternal mortality. Therefore, clinicians should be wary of possible aforementioned complications during prenatal examinations of their pregnant patients with SLE.

Key words: Systemic lupus erythematosus, pregnancy, peripheral neuropathy

Sistemik Lupus Eritematosuslu Bayan Hastada Üçüncü Tremesterde Gelişen Vaskülitik Nöropati

ÖZET

Sistemik lupus eritematosus (SLE) daha çok doğurganlık çağındaki kadınlarda görülen inflamatuar otoimmun bir hastalıktır. 20 yaşındaki 35 haftalık gebe olan bayan hasta kliniğimize 20 gündür olan bacak ağrısı, bacaklarda kuvvetsizlik ve sol düşük ayak şikayetleri ile başvurdu. Özgeçmişinde ayda bir defa olan oral ülserler, zaman zaman eklem ağrıları ve gün ışığında belirginleşen malar rash vardı. yapılan EMG de duyusal ağırlıklı sensorimotor polinöropati tespit edildi ve hastaya SLE' ye bağlı vaskülitik nöropati tanısı konuldu. silik semptom ve bulguları olan SLE hastaları gebelik dönemindeki hastalığın sistemik alevlenmeleri neticisinde kötüleşebilir, dahası bu durum fetal ve maternal mortalitenin artmasına da neden olabilir. Dolayısıyla klinisyenler SLE li hastalarının gebelik dönemindeki alevlenmelere bağlı gelişebiliecek muhtemel komplikasyonlara karşı uyanık olmalılardır.

Anahtar kelimeler: Sistemik lupus eritematosus, gebelik, periferal nöropati

INTRODUCTION

Systemic Lupus Erythematosus (SLE) is an important systemic disease with high morbidity and mortality(1). As one of the causes of secondary vasculitis, this autoimmune inflammatory disease can show a wide variety of systemic manifestations. Major organ involvement of the renal system (lupus nephritis) or of the central nervous system worsens the overall prognosis. Both the central and peripheral nervous system can be affected

Received: 20.01.2013, Accepted: 12.03.2013

with a wide variety of neuropsychiatric complications such as mood disorders, cognitive disorders, myelopathy, movement disorders, peripheral neuropathies. SLErelated neurological involvement can be seen nearly 19-75% of patients (1,2). Neuropathy develops in approximately 5% of all cases of SLE. The most common forms of peripheral nervous system involvement in SLE are mononeuritis multiplex and distal sensory or sensorymotor polyneuropathy (3). The effects of pregnancy on

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lupus are controversial, but the reports of lupus flares during pregnancy are greater. SLE patients may develop complications such as gestational hypertension, preeclampsia, eclampsia and HELLP syndrome during pregnancy. Moreover, the fetus may experience intrauterine growth restriction, preterm delivery, low birth weight or fetal demise (1). The incidence of life-threatening exacerbations during pregnancy have been reported to be approximately 5-41% (4).

CASE

A 20-year-old, 35 weeks, gravida 1 parita 0 pregnant patient was admitted to our clinic with a history of leg pain lasting for 20 days, numbress in the feet and left foot drop. In another Obstetrics and Gynecology Clinic Center, she had been started on magnesium therapy for her pain. On her physical and neurological examination, a bilateral malar rash was detected. Cranial nerve examination was normal. The motor examination showed left foot dorsiflexion 2/5, plantar flexion -5/5 and no motor deficit was detected on other muscle examinations. On sensory examination, there was bilateral hypoesthesia under the ankles. Deep tendon reflexes were not detected. Bilateral flexor plantar responses and cerebellar tests were also normal. On further examination, the patient reported the occurence of oral ulcers once a month, occasional joint swelling and photosensitivity, with a worsening malar rash under the sunlight. There was no history of prior recurrent miscarriage, genital ulcer, a known disease diagnosis or previous medication use. Magnesium therapy was interrupted by Obstetrics. The cranial and spinal magnetic resonance imaging was normal. Complete blood count, biochemistry tests, vitamin B12, folate, brucellosis, thyroid function tests were normal. Lab studies revealed ANA positivity, ESR: 82mm/hr, CRP: 102, platelets 325000, urea 8.3, creatinine 0.45, potassium 3.8 and sodium 140 respectively, however, antiphospholipid antibodies, such as lupus anticoagulant, could not be tested due to the lack of availability of these tests at our hospital. Thrombocytopenia was not found at follow-up visits. Two weeks after discontinuation of magnesium, bilateral achilles reflexes were not detected and bilateral upper extremity reflexes were normoactive. EMG was consistent with sensory-predominant sensorimotor polyneuropathy. As a result of rheumatology consultation, a definitive diagnosis of SLE was made. SLE-related sensorimotor polyneuropathy was diagnosed due to hypoesthesia of both feet, muscle weakness in distal lower extremity and areflexia, supported by EMG findings. She was decided to start on steroid therapy but at the 37th week of gestation, and during the second week of hospitalization, the patient had a preterm vaginal delivery giving birth to a healthy male baby. The patient's complaints subsided postpartum but some symptoms remained. The patient was then referred to rheumatology for long-term disease management.

DISCUSSION

Systemic Lupus Erythematosus (SLE) is an inflammatory, autoimmune rheumatologic disease with frequent neurological complications occuring in upto 50% of patients. The most common neurological symptom is headache, and less frequently mood changes, cerebrovascular disease, cognitive disorders and epileptic seizures are seen. The symptoms of neuropathy, myasthenia gravis, myelopathy, and movement disorders are very rare (5,6). The most common rheumatologic autoimmune disease in the reproductive age is SLE with an incidence of approximately 1/1000 (7). The most important presentation in pregnant patients with SLE is exacerbation of the disease (4). Previous studies have reported increased frequency of complications such as preeclampsia, eclampsia, renal disease, preterm delivery, non-elective cesarean section, postpartum hemorrhage, low birth weight and delivery-related deep vein thrombosis. Hypertensive complications of pregnancy occur in approximately 10% to 20% of all pregnant women with lupus (8,9). Our patient did not develop eclampsia or preeclampsia, however, she had a preterm delivery at 37th week of gestation, which is consistent with previously reported literature.

During pregnancy, while the frequency of attacks is still debated, the general opinion for the SLE exacerbation during pregnancy has recently moved in the direction of increased attack frequency. Three prospective studies have confirmed this situation and have shown that flare-ups generally occur in the first and second trimesters (10-12). Unlike the works mentioned, neuropathy occurred in third trimester in our patient. In another study, the incidence of nephritis was reported to increase in pregnant patients with SLE (13). Our patient did not have previously known kidney disease nor was she found to have kidney disease during her regular prenatal visits. Some authors suggest that exacerbations during pregnancy may be due to discontinuation of medication (14). In addition, an increased risk of preeclampsia and eclampsia during pregnancy in patients with SLE has been reported (15). Some complications such as spontaneous abortion, intrauterine growth retardation, preterm labour, fetal heart blocks, and systemic venous thromboses can be considered in pregnant SLE patients and in the fetus (16). Due to these reasons, an increase in the frequency of cesarean sections in pregnant SLE patients and prolonged length of stay with delayed hospital discharge can occur (17). Another study has suggested that major systemic and non-renal involvement are rarely seen (18). Tandon A et al mentioned that the frequency of exacerbations during pregnancy do not increase (19). Central nervous system (CNS) involvement is seen more commonly than peripheral nervous system (PNS) involvement in SLE. In 30% of cases, PNS involvement occurs electrophysiologically without the presence of symptoms (20), and is present usually in the form of sensory or sensorimotor polyneuropathy. In accordance with the literature, sensory-predominant sensorimotor polyneuropathy has occured in our case. Our patient's disease was not diagnosed before pregnancy due to minor clinical signs. The disease exacerbation during pregnancy and the development of a rare neurological involvement, such as polyneuropathy, led to a definitive diagnosis of SLE. We think our case is interesting in this respect. In the literature, vasculitic neuropathy due to SLE has not been previously reported during pregnancy.

In conclusion, the frequency of exacerbations of rheumatic diseases can vary during pregnancy. As in our case, there may be flares of SLE during this period. Thus, some patients may be diagnosed with SLE due to worsening of latent systemic disease or due to the development of complications during pregnancy. Cinicians must be aware about the presence of neurological and other complications during the follow-up of pregnant patients with SLE.

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