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CASE REPORT

A Case of Latent Syphilis in Pregnancy Masked by Beçhet's Disease

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Abstract

Syphilis in pregnancy is a worrisome topic due to its fetal consequences. The diagnosis and management of such a case might be intriguing when overlapped with Behçet's disease, an inflammatory disease involving multiple organ systems. This case presents a 35-year-old pregnant female, a known case of Behçet's disease and hypothyroidism, presenting at 33 weeks to the emergency department at Bahrain Defense Force (BDF) hospital with vaginal leaking and a solitary oral ulcer. As per the guidelines of the ministry of health, she underwent perinatal screening and was found to be positive for the venereal disease research laboratory (VDRL) test and was diagnosed with latent syphilis. The diagnosis of latent syphilis in a known case of Behçet's disease presenting with a solitary oral ulcer is unpredictable. The importance of perinatal screening is emphasized in this case report.

Keywords: Latent Syphilis, Behçet's disease, Pregnancy

Introduction

Syphilis is an infection caused by the spirochete bacteria *Treponema pallidum*. It causes concern during pregnancy due to its risk of vertical transmission to the fetus and the health consequences that it has on the baby. Early detection and management of infected individuals during pregnancy is important to prevent its serious consequences.[1]

Behçet's disease (BD) is an inflammatory disease with multisystem involvement, characterized by oral and genital ulcers along with skin and ocular lesions. The disease was first described in 1937 by the dermatologist Hulusi Behçet and was designated its name after him.[2]

The following case report discusses both BD and latent syphilis in pregnancy while signifying the importance of prenatal screening for infectious diseases that can significantly affect the outcome of the pregnancy and the effects of both diseases on the mother and fetus.

Case presentation

A 35-year-old female, gravida 6, para 5, presented to the labor room at BDF hospital with vaginal leakage and dyspareunia at 33 weeks of gestation to be assessed for possible premature rupture of membranes and early onset of labor. The patient has a history of five spontaneous pregnancies, which were all vaginal deliveries without complications.

She has been suffering from BD since the age of 23, initially presenting with recurrent episodes of poly-arthralgia, bilateral flank pain, chronic constipation, and oral ulcers, denying any history of genital ulcers, uveitis, deep venous thrombosis, and recurrent miscarriages. In 2017, her diagnosis was

confirmed by serological testing, which was positive for the human leukocyte antigen (HLA) B5, and she was started on treatment using colchicine, an alkaloid agent extracted from the plant colchicum, which caused her symptoms to drastically subside. She has no family history of any auto-immune or rheumatological diseases.

Upon admission, she was afebrile and had stable vital signs. Moreover, she was assessed for her vaginal leaking, and premature rupture of membranes (PROM) was excluded since no vaginal pooling was found. This was supported by a negative amnicator test, and a high vaginal swab had no significant growth. Upon systemic examination, she had a single oral ulcer, no genital ulcers, and no other significant signs were present. A cardiotocography (CTG) was used on multiple occasions to assess the fetal heart rate and uterine contractions, and it was reassuring throughout. An ultrasound scan was performed by the senior resident and no abnormalities were noted. As part of the prenatal screening, she was screened serologically for infectious diseases as part of the routine medical workup since the patient had not been attending her outpatient antenatal visits due to the pandemic of the corona virus disease of 2019 (COVID-19). The patient was found to be serologically VDRL positive with a titer of 1:2, with her serum also testing positive for Treponema antibodies, classifying her as having late latent syphilis.

When asked for further details, she had a single sexual partner to date, her husband, and she denied a history of blood transfusions. With regards to her surgical history, she only had surgical ablation and reduction of hypertrophied inferior turbinates. An immediate treatment plan was initiated in coordination between the infectious disease and, obstetric and gynecological departments. The patient received three doses of benzathine penicillin at a dosage of 2.4 million units once weekly for three consecutive weeks.

A detailed fetal scan was done to exclude any congenital abnormalities in the fetus, including signs of congenital syphilis. The scan showed a single viable fetus in a cephalic presentation with present fetal movements and fetal heart sounds with an expected weight of 2800 grams, with the estimated weight and age corresponding to the gestational age at the date of the scan.

The patient delivered a healthy baby boy at the 39th week of gestation by a spontaneous vaginal delivery. The baby was alive and well, with a birth weight of 3.96 kg. The baby was screened negative for congenital syphilis or other diseases.

After the 2nd week postpartum, the patient suffered from severe flares of BD symptoms, experiencing her first episode of uveitis ever since her initial symptoms appeared 13 years ago. She also suffered from severe arthralgia with the eruption of some oral ulcers. She resumed her medical management and her symptoms have flared intermittently since.

Discussion

BD is characterized by recurrent episodes of vasculitis with multi-organ involvement. Also known as the Silk Road Disease, it is more common in countries along the "silk road," which includes Turkey, Japan, and Iran. It commonly affects individuals between 20 and 40 years of age. Its etiology is not yet determined, but it has been associated with HLA and non-HLA mechanisms. Despite the data available about the disease, there is usually a delay between the onset of symptoms and the time of diagnosis due to the variations in clinical presentation and the vast differential diagnosis.[3]

The clinical manifestations of BD can be categorized systematically. Mucocutaneous manifestations include orogenital ulcers, with oral ulcers predominating and being the initial complaint in 80% of the patients. Other non-specific cutaneous manifestations include variable vasculitic lesions, affecting 48–88% of the cases. Ocular manifestations usually involve the retina and uvea, prevailing in 30–70% of the patients, with a possible consequence of blindness in another 25%. Arthropathies are found in 45–60% of patients with BD. Gastrointestinal manifestations are reported in 3–26% of patients, varying in different populations and being more frequent in Japan than in the Middle East and Mediterranean region. In males, neurological involvement affects 5–10% of the patients.[4]

The diagnosis of BD is based on clinical instincts. Biopsy and histological studies are insignificant in the diagnosis of BD, illustrating neutrophilic infiltrates, lymphocytic aggregates, and vascular proliferation, all of which are unspecific.[5]

Pregnancy usually decreases the symptoms of BD, and theories suggest that the attenuation of the cell-mediated immune response in pregnancy plays a role in the improvement of the clinical manifestations of BD. Another mechanism involved is the decrease in the chemotaxis and adherence of neutrophils. The hormonal changes during pregnancy aid in immunosuppression, and this supports that in the postpartum period, as the hormones normalize, the symptoms flare, as what happened to our patient.

BD does not dictate the mode of delivery; a normal vaginal delivery imposes no maternofetal risks. It however increases the rate of obstetric complications, including miscarriages, gestational diabetes mellitus, PROM, and preterm delivery, amongst others. Fetal complications from BD are rare, and neonatal BD is transient, lasting up to 8 weeks. Topical use of steroid creams or topical sucralfate is efficient in managing orogenital ulcers of BD in pregnancy. Tropicamide and steroid drops are used for ocular manifestations. Oral medications including prednisolone, azathioprine, cyclosporine, and tacrolimus are all safe to be used in pregnancy. Colchicine, methotrexate, and infliximab are not approved by the federal drug agency (FDA) to be used in pregnancy. Breastfeeding is recommended in patients with BD, with the data showing little to no evidence of the harm of the medications on the infants breastfed by their affected mothers.[6]

Treponema pallidum, a spirochete bacterium, is the causative organism causing syphilis. For diagnosing syphilis, both the history and clinical presentation are important. The diagnosis is confirmed by identifying the bacterium by dark-field examination, serologic tests, or by immunofluorescent staining and polymerase chain reaction assays. Clinical suspicion is highly required for the diagnosis since the manifestations of syphilis, "the great imitator," coincide with multiple diseases, including Behçet's disease.[7,8]

There has been a yearly incidence of up to 6 million new cases of syphilis, implementing a high level of morbidity and mortality in the precious infants affected by the congenital disease.[9]

Syphilis in pregnancy, if unmanaged, may cause serious complications for the fetus. Parenteral benzathine penicillin G is the treatment of choice to prevent vertical transmission of the infection. A study revealed that screening for syphilis during pregnancy increased from 89.8% to 97.2%, causing a reduction in the incidence of congenital syphilis from 11.7% to 3.2% and a reduction in the incidence of stillbirths or fetal loss from 19.0% to 3.3%.[10]

Congenital syphilis is mostly thought to be transmitted transplacentally, though labor also imposes a possible risk of transmission. Mothers with untreated early latent syphilis had a 20% prematurity rate, 4% neonatal deaths, 10% stillbirths, 40% suffered from congenital syphilis, and 20% of infants were born full-term unaffected by syphilis. With untreated late-latent syphilis, the numbers were reduced; about 10% of infants were born with congenital syphilis and another 10% were stillborn.[11]

A study conducted in Brazil showed that gestational syphilis was more common in multiparous women and women who had not received adequate prenatal care, with 78.23% of pregnant women having the diagnosis. According to the study, 39.26% of the diagnosed cases of gestational syphilis occurred during the first trimester, 31.11% during the second trimester, and the remainder during the third trimester.[12]

A study of a female in her first trimester was diagnosed with latent syphilis and treated with benzathine penicillin as per the CDC guidelines. During her second trimester, she had an initial ultrasound scan that revealed no abnormalities. A month later, her scan revealed features of congenital syphilis. A fetal autopsy revealed cutaneous erythematous papular rash, hepatomegaly and ascites, hydrothorax, obstructive hydrocephalus, ischemic and hemorrhagic brain injuries, productive meningitis with dystrophic intracranial calcifications, and thymic involution.[13]

The use of ultrasonography is of great importance in detecting congenital syphilis. The commonest ultrasonographic findings are hepatosplenomegaly, placentomegaly, and intra-uterine growth restriction, intrahepatic calcifications, ascites, fetal hydrops, and intrauterine fetal death.[14]

A few cases have been reported in literature regarding both syphilis and BD. This study reported two cases presenting with panuveitis who were given an initial diagnosis of BD. They had a history of recurrent orogenital ulcers that persisted despite their treatment as BD. Further investigations revealed that both cases had a coinfection of syphilis and human immunodeficiency virus (HIV) infection, and when treated, the symptoms were alleviated. This indicates that syphilis and HIV coinfection may mimic the clinical presentation of BD.[15]

A case was reported of a 22-year-old man with a history of recurrent orogenital ulcers and hyperpigmented tender nodules distributed along the shins. He was initially diagnosed with BD and treated with steroid therapy, causing an improvement in the nodular lesions but an aggravation of the orogenital erosions. Upon further investigations, he was diagnosed with secondary syphilis and was accordingly treated, leading to a gradual improvement in his symptoms clinically.[8]

Conclusion

BD and syphilis are rare diseases in pregnant women, but both can cause serious implications for both the mother and the fetus. Though Behçet's disease usually subsides clinically during pregnancy, it may present with rare complications such as abortion, and its post-natal flares can be very debilitating. On the other hand, syphilis infection is consequential since it can cause serious complications and the possible loss of the fetus. Prenatal care and screening play a significant role in preventing and identifying such unfortunate events early enough to help counsel the family. Luckily, for our patient and her baby, there were no complications, nor were any anomalies identified.

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