

Congenital Syphilis as a Cause of Hydrops Fetalis in a Woman With Syphilis Incognito

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Abstract

The aim of this study is to report a case of congenital syphilis which was diagnosed at 28th gestational week with hydrops fetalis. A 20-year-old primigravid woman immigrant from Moldavia was referred to our perinatology unit at 28th gestational week because of fetal ascites. Her blood group was 0 Rh positive and her husband's was B Rh positive. She was diagnosed as non-immun hydrops fetalis. A day following the admission the fetus died. By the diagnostic work-up congenital syphilis was demonstrated, and the mother was treated conveniently. In Turkey although there is no supportive data for routine testing, targeted screening of patients who immigrate from high-risk countries, who had sexual intercourse with high-risk person, and whose partner is working in high-risk countries should be screened at the beginning of pregnancy, and rescreened at the beginning of the third trimester.

Keywords: congenital syphilis, perinatal outcome, screening

Özet

Hidrops Fetalis Nedeni Olarak Konjenital Sifiliz

Bu çalışmada hidrops fetalis olarak belirti veren bir konjenital sifiliz olgusu sunulmaktadır. Moldovya göçmeni, 20 yaşında primigravid bir kadın perinatoloji kliniğimize 28. gebelik haftasında fetal asit nedeni ile refer edildi. Annenin kan grubu 0 Rh pozitif ve kocasının B Rh pozitif idi. Hasta nonimmün hidrops fetalis olarak kabul edildi. Hospitalizasyonun ertesi günü intrauterin fetal exitus gerçekleşti. Diagnostik çalışmalar neticesinde fetüse konjenital sifiliz tanısı konuldu ve annenin tedavisine hemen başlandı. Ülkemizde rutin tarama için yeterli destekleyici data olmasına rağmen yüksek risk bulunan ülkelere göç eden hastalar, yüksek riskli kişilerle cinsel ilişkide bulunma hikâyesi olanlar, kendisi veya erkek arkadaşı/kocasını yüksek riskli ülkelere çalışan kadınlar gebelik tanısı konulduğunda ve üçüncü trimester başında sifiliz için taramadan geçirilmelidir.

Anahtar sözcükler: konjenital sifiliz, perinatal prognoz, tarama

Introduction

Syphilis is a chronic systemic infectious disease caused by the spirochete, *Treponema Pallidum*. Syphilis occurs throughout the world but it is most frequent in large urban area. By the extensive use of penicillin the incidence reached a nadir in the 1950s, but increased through the 1980s and peaked in 1990. Since then have declined to lowest rate in 1998. Importantly, over 80% of women with syphilis are in the reproductive age group and congenital syphilis may occur at any time during pregnancy via transplacental transmission. Rates of congenital syphilis closely parallel the rates of primary and secondary syphilis. In women with untreated early syphilis, 40% of pregnancies result in spontaneous abortion, stillbirths, or perinatal deaths (1). So, early diagnosis and treatment is important to prevent fetal complications. The purpose of this study is to report a case of congenital syphilis

which was diagnosed at 28th gestational week with hydrops fetalis.

Case Report

A 20 years old primigravid woman immigrant from Moldavia was referred to our perinatology unit at 28th gestational week with fetal hydrops. Her blood group was 0 Rh positive and her husband's was B Rh positive. She was diagnosed as non-immun hydrops fetalis. The ultrasonographic fetal biometry and anatomic survey revealed one active female fetus with hydrops fetalis, placentomegaly and hepatosplenomegaly. There was no anatomical defect causing hydrops, so blood tests were performed to detect etiology. A day after the admission the patient underwent ultrasonographic examination with a complaint of loss of fetal activity. There was no cardiac activity. By oxytocin induction 2200 gr, 47 cm, 0/0 APGAR female fetus was born. The fetus was edematous and hydropic (Figure 1). The placenta was paler, thicker, and larger than normal (Figure 2). In the laboratory examination Venereal Disease Research Laboratory (VDRL) slide test and the *Treponema Pallidum* Microhemagglutination Assay were positive. In the pathologic examination of fetus the li-

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Figure 1. The fetus with hydrops.



Figure 2. Placenta of the fetus.



Figure 3. Hepatomegaly and splenomegaly in autopsy.

ver and spleen were larger than normal (Figure 3), and diffuse inflammation and hemophagocytosis was detected, so the intrauterine infection was confirmed. Treponemal stains were positive in splenic tissue.

In the history of patient, none of the characteristic features of primary or secondary syphilis was reported. She has been married for 3 years with her husband and none of the partners had any sexual intercourse with another person. In the physical evaluation of both the mother and father, no lesion was detected. Father's testing for *T. pallidum* was negative. The mother was treated with weekly 2.4 million units intramuscular Benzathine penicilline G for 3 doses. The mother's VDRL titers were followed with 4 weeks interval until the negative results were obtained.

Discussion

Syphilis was first described in the medical literature in 16th century. Following the discovery of penicillin the incidence of syphilis reached a nadir in the 1950s (2). A dramatic resurgence was seen in the late 1980s and early 1990s due to the increased incidence of intravenous drug abuse, HIV infection, transcountries and transatlantic travel, liberalism in sexual life and relaxation and changes in screen and treatment policies in several countries (1). Since then, by strict screening programmes and treatment protocols rates have declined significantly. Still in some developing countries, syphilis is an endemic disease. World Health Organization reports that a big part of 12 million new syphilis cases per year is guessed to live in Southeast Asia and African countries. Concordant with the incidence of syphilis, congenital syphilis cases also increases. In some African countries congenital syphilis rates are as high as 8-10% (3,4). In Turkey, there is no sufficient data about incidence of syphilis in pregnancy. In the study of our clinic having limited number of pregnant patients, no sero-positive woman was detected (5).

If syphilis occurs in pregnancy, fetal infection is possible in all trimester and all stages of the disease. The fetus does not generally manifest clinical disease before 18 weeks since the relative incompetence of immune system (6). The rate of fetal infection is principally related with the stage and duration of maternal infection. It is highest at early syphilis and lowest in late syphilis. Fetal infection at early latent stage is somewhat higher than the late syphilis. The transmission rates in untreated patients are 100% during the second stage, 80% during the early latent stage, and 30 % during the late latent syphilis. Infants born to treated mothers have only a 1-2% risk of infection (7).

“Syphilis incognito” is a special form of syphilis in which the infected person has no clinical symptom but the diagnosis is made with serological tests during the routine screening. The person does not know when, where and from whom the infection transmitted. In a venereal diseases hospital in Greece, 480 of 711 cases (67%) were syphilis incognito (8). Also our patient was considered as syphilis incognito. Such cases are important in expansion of disease and congenital infection.

The clinical manifestation of congenital syphilis may be intrauterine growth restriction, non-immune hydrops fetalis, stillbirth and preterm delivery. Two-thirds of infants with

congenital syphilis will be asymptomatic at birth. Two weeks later maculopapular lesions, rhinitis, hepatosplenomegaly, chorioretinitis, petechias and pseudoparalyse occur. The physiopathology of stillbirth is explained with placental infection or fetal hydrops (9). In our patient both the placental infection and hydrops were detected.

Benzathine penicillin is effective for preventing maternal transmission to the fetus and for treating fetal-established infection (1). In pregnancy, the other alternatives doxycycline and tetracycline are contraindicated because of their teratogenic effects. Another alternative erythromycin is ineffective to prevent fetal infection. So, pregnant women who have a history of penicillin allergy should be desensitized and treated with penicillin. A second dose of benzathine penicillin may be administered 1 week after the initial dose for women who have primary, secondary, or early latent syphilis. Ultrasonographic signs of fetal syphilis (i.e., hepatomegaly and hydrops) indicate a greater risk for fetal treatment failure. Response to treatment must be monitored with the non-treponemal antibody tests. In 6-month period titers should decrease 4-fold and become negative by 12-24 months.

One of the unique feature of treatment is Jarisch-Herxheimer reaction which is a systemic inflammatory response caused by massive release of treponemal lipopolysaccharides and characterized by shaking chills, fever, myalgia, tachycardia and hypotension. In pregnant patients, uterine contractions, premature labor, decelerations, decreased fetal movements and fetal demise may occur. So the patients should be monitored closely. In our patient such an allergic reaction did not observed.

Prenatal screening remains the most important factor in identification of infants at risk for development of congenital syphilis and ideally results in diagnosis and treatment during gestation. Routine maternal serologic testing for syphilis is mandatory especially in high-risk population. Also second testing at the beginning of the third trimester (28 weeks) should be performed for these patients. It is also indicated for

women with suspect lesions or a history of exposure to an infected sexual partner. Additional testing at delivery is recommended for mothers who live in areas with a high prevalence of syphilis. In the USA, women who are at high risk of acquiring syphilis, or who belong to communities with a high prevalence of syphilis, are re-screened at 28th gestational week and again at delivery (1). In our country although there is no supportive data for routine testing, targeting screening of patients who are immigrant from high-risk countries, who had sexual intercourse with high-risk person, and who or her partner/husband have job in high-risk countries should be screened at the beginning of pregnancy, and rescreened at 28th gestational week. If the diagnosis of syphilis is established, coordinated prenatal care and follow-up are important in the management of pregnant women.

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