

CLINICAL CASE

Dent. Med. Probl. 2012, 49, 3, 439–442
ISSN 1644-387X

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Congenital Syphilis in a Multiple Children Family – Own Case

Kiła wrodzona w wielodzietnej rodzinie – przypadek własny

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A – koncepcja i projekt badania; B – gromadzenie i/lub zestawianie danych; C – opracowanie statystyczne;
D – interpretacja danych; E – przygotowanie tekstu; F – zebranie piśmiennictwa

Abstract

Syphilis is an infectious disease that in recent years has been affecting a fast growing number of people – globally and in Poland. The objective of the study was to present a case of occurrence of congenital syphilis in one child raised in a multiple children family. The study presents the case of a 12-year-old boy. The examining doctor noticed the symptomatic appearance of his incisors and facial profile. The consequent analysis of the patient's medical history and physical examination gave grounds to confirm late congenital syphilis. The analysis of the patient's dental history, community interview and an examination of medical files provided by the family's general practitioner confirmed the case of congenital syphilis. It was discovered that the mother was diagnosed with secondary syphilis as late as in the second trimester. Despite administering adequate treatment, it was impossible to protect the child from being infected with *Treponema pallidum*. Tests performed on the infant showed a heightened VDRL titre (higher than in the mother's case). Therefore, the boy underwent antibiotic treatment (penicillin). The other children by the same mother show no symptoms of *Treponema pallidum* infection. During the clinical trial, the following was identified: Hutchinson's teeth with pitted enamel hypoplasia, gothic palate, saddle nose, Olympian brow as well as unclear speech which may suggest prior congenital *syphilitic coryza*. Diagnosing syphilis at an early stage of pregnancy and the commencement of specialist treatment does not preclude infecting the foetus or the occurrence of congenital syphilis symptoms. Running tests even before the conception would not only have decreased the risk of further spreading of the disease but also it would have allowed giving birth to a healthy child (Dent. Med. Probl. 2012, 49, 3, 439–442).

Key words: congenital syphilis, acquired syphilis, Hutchinson's teeth.

Streszczenie

Kiła jest chorobą zakaźną, która w ostatnich latach charakteryzuje się zwiększoną liczbą zachorowań na świecie i w Polsce. Celem pracy jest opis przypadku występowania kiły wrodzonej u dziecka z rodziny wielodzietnej. Opisano przypadek 12-letniego chłopca. Lekarz dentysta zauważył zmiany na zębach siecznych oraz w profilu twarzy. Dokładne badanie anamnestyczne i kliniczne potwierdziło przypadek późnej kiły wrodzonej. Zostało to także potwierdzone w historii pacjenta u lekarza rodzinnego. U matki chłopca stwierdzono kiłę wtórną pod koniec drugiego trymestru ciąży. Mimo podjęcia właściwego leczenia, nie była możliwa ochrona dziecka przed zakażeniem *Treponema pallidum*. W badaniu przesiewowym dziecka w kierunku kiły (VDRL) wykazano podwyższone miano swoistych przeciwciał, większe niż u matki. Chłopca leczono penicyliną. U innych dzieci tej samej matki nie potwierdzono zakażenia kiłą. W badaniu stomatologicznym wykazano następujące objawy kliniczne kiły: zęby Hutchinsona z hipoplazją szkliwa, podniebienie gotyckie, nos siodełkowaty, czoło olimpijczyka oraz niewyraźną mowę, która mogła sugerować objaw Saplikizowej (*Coryza syphilitica*). Rozpoznanie kiły we wczesnym okresie ciąży, a następnie właściwe leczenie specjalistyczne nie zapobiegają zakażeniu płodu lub wystąpieniu objawów kiły wrodzonej. Wykonanie odpowiednich testów nawet przed poczęciem dziecka nie tylko zmniejsza ryzyko późniejszego rozprzestrzenienia się choroby, lecz także umożliwia urodzenie zdrowego dziecka (Dent. Med. Probl. 2012, 49, 3, 439–442).

Słowa kluczowe: kiła wrodzona, kiła nabyta, zęby Hutchinsona.

Syphilis is an infectious disease that in recent years has been affecting a fast growing number of people – globally and in Poland [1]. It is a sexually transmitted disease causing multi-organ infection that proceeds in three stages: primary syphilis (after 3 weeks from infection), secondary syphilis (after approx. 9 weeks from infection), and tertiary syphilis (that starts three years after the intrusion of the pathogen). The World Health Organisation reports that globally nearly a million pregnant women are affected by *Treponema pallidum*, which leads to miscarriage, giving birth to children with congenital syphilis, premature births with low birth weight [2]. If a woman infected with syphilis is pregnant, the infection is transmitted to the developing foetus. Most often it gets infected via the placenta around the 16th gestation week, though there have been cases of such infections already in the 9th or 10th gestation week [3]. The pathogen may also infiltrate amniotic fluid and thus infect the foetus. Another method of transmission from the mother onto the foetus is a direct contact of the child with an early-stage lesion during delivery. The probability of an intrauterine infection of the child is strictly related to the severity of the mother's infection: the stronger the disease expression is, the higher the risk of infecting the foetus. In the case of early syphilis it is nearly 100%, in the case of early latent syphilis – approx. 40%, and in the case of late latent syphilis – approx. 10% [4].

The presence of *Treponema pallidum* in the placenta initiates its rapid growth which may lead to premature deliveries (7th-8th month of gestation) as well as intrauterine fetal death or early miscarriages [5].

In Poland, in 2004 as many as 19 cases of congenital syphilis were reported; in 2005 there were 8 of them, and in 2006 – 6 [6].

According to Polish law, the performance of syphilis screen tests is obligatory in the first and third trimester of pregnancy. VDRL, a reactive nontreponemal serologic test, is routinely applied, which can confirm the presence of antibodies resulting from the current infection or passed syphilis. If the result is positive, it is necessary to verify it using the reactive treponemal tests, e.g. FTA or FTA-ABS. One can also use the method of direct detection of *Treponema pallidum* by a dark-field microscopic examination, collecting biological material from the placenta or umbilical cord. A positive VDRL result (titre greater than the mother's titre), confirmed with reactive treponemal tests FTA-ABS or by detecting IgM antibodies in the child's blood (IgM cannot pass via the umbilical cord), substantiates a congenital syphilis diagnosis [3, 4].

The aim of the study was to present a case of congenital syphilis from the perspective of dentistry.

Case Report

Patient R.A., aged 12, came to a dental clinic for a periodic check-up. The doctor was alerted by the symptomatic appearance of his facial profile and incisors. An analysis of the patient's medical history was carried out as well as a dental examination.

The medical interview, community interview and examination of medical files provided by the family's general practitioner confirmed the case of congenital syphilis. It was established that the boy's mother had been diagnosed with secondary syphilis as late as in the second trimester. It was also established that the child's father died of the very same disease before he was born. Despite administering adequate treatment, it was impossible to protect the child from being infected with *Treponema pallidum*. Tests performed on the infant showed a heightened VDRL titre (greater than the mother's titre), so antibiotic treatment (penicillin) was applied to the boy. The other children by the same mother show no symptoms of the *Treponema pallidum* infection. Even so, the two youngest children underwent preventive treatment soon after they were born.

During the extraoral dental clinic examination, saddle nose and Olympic brow were detected. The intraoral examination established the presence of gothic palate and developmental abnormalities of incisors, whose deformations were caused by pitted enamel hypoplasia (Fig. 1). In addition, primary and secondary dental caries of numerous teeth were discovered, marginal periodontitis caused by the presence of abundant calculus and very bad hygienic condition of the oral cavity. Also, unclear speech may suggest that the boy had congenital syphilitic coryza in his infancy.



Fig. 1. Pitted enamel hypoglasia on incisors

Ryc. 1. Dołkowata hipoplazja szkliwa na zębach siecznych

The treatment plan assumes conservative treatment of dental caries and removal of calculus. The boy and his guardian were instructed to have regular dentist appointments as well as on the proper brushing of teeth. Additional hygienic measures were implemented as well as fluoride prophylaxis. Other suggestions included elimination of aesthetic deficiencies of incisors in the future.

Discussion

Congenital syphilis has two stages: early and late. Symptoms of early congenital syphilis develop in children up to two years old. The first symptoms can appear immediately after delivery or shortly afterwards. Usually the child after intrauterine syphilis infection is delivered prematurely, with low birth weight and in poor condition in general. One of the first pathognomonic symptoms of the disease is *rhinitis*, i.e. congenital syphilitic coryza, which takes the form of dense secretion from the nose, making it much harder for a child to breathe and suck. If no treatment is applied, the nose bone structure is damaged and deformed permanently, resulting in the development of the characteristic saddle nose. Another condition observed in children with congenital syphilis is jaundice accompanied by hepatomegaly and liver fibrosis due to interstitial liver inflammation. In 90% of cases, radiological examination confirms *ostochondritis* of the long bones, their widening and damage of their epiphysis which looks as if "moths fed on it". In addition, children have skin symptoms, wart-like lesions, the same as in the case of secondary syphilis, and characteristic Hochsinger's rhagades around lips which tend to transform into radiating fissures and clefts, which turn into scars when healed [3, 4].

Late congenital syphilis is diagnosed most often after 5–20 year of life at the earliest. Its symptoms are mainly the fixed results of early congenital syphilis. If the patient has never undergone any treatment, serology reagents are always positive. The symptoms of late congenital syphilis include the Hutchinson's triad, namely: interstitial keratitis, Hutchinson's incisors, and eighth nerve deafness (n. VIII). In addition, saddle nose – the result of syphilitic coryza, and Parrot's scars around lips

– the result of healing wounds – are observed. Late symptoms might manifest themselves as Olympic brow (marked thickening of the bony prominence of the forehead), gothic palate, sabre shin, and unilateral enlargement of the sternoclavicular portion of the clavicle. All of these deformations of bones stem from periostalgia [3, 4].

In order to commence treatment of syphilis in pregnant women, first they need to have a positive result of the VDRL serology reagent screen test. Treatment with large doses of penicillin begins even prior to positive FTA and FTA-ABS tests. If treponemal reagents do not confirm the screen test result, treatment is terminated. Procaine penicillin is used intramuscularly (*i.m.*) in a single daily dose of 1,200,000 units for 20 consecutive days. The therapy needs to be repeated in the second half of the pregnancy. If the patient had been treated for syphilis before conception, prophylactic treatment needs to be applied in a dose of 1,200,000 units (*i.m.*) of procaine penicillin.

Indications for treating the newborn for congenital syphilis include: greater VDRL titre than the mother's, IgM antibodies in blood, syphilis-characteristic clinical and radiological signs and confirmed and untreated syphilis in the mother or syphilis treated in the last trimester of pregnancy.

Having confirmed the indications, treatment should be implemented as soon as possible, comprising the application of:

- in early syphilis in the 1st year of life 50,000 units of crystal penicillin *i.v.* for 15 days in two daily doses, in the 2nd year of life 300,000 units of procaine penicillin *i.m.* for 15 days as well;
- in treatment of late congenital syphilis, i.e. after two years of living, procaine penicillin is used depending on the child's weight: from 600,000 units to 1,200,000 units for 30 days [3].

The duration of penicillin therapy depends on the results of syphilis serology reagents, which should be taken quarterly until their negative results are obtained or their titre drops fourfold [4].

The authors concluded that diagnosing syphilis at an early stage of pregnancy and commencement of specialist treatment does not preclude infecting the foetus or the occurrence of congenital syphilis symptoms. Running tests even before conception would not only decrease the risk of the disease spreading further but would also allow the mother to give birth to a healthy child.

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Received: 22.02.2012

Revised: 6.03.2012

Accepted: 25.06.2012

Praca wpłynęła do Redakcji: 22.02.2012 r.

Po recenzji: 6.03.2012 r.

Zaakceptowano do druku: 25.06.2012 r.