

II. NEONATOLOGY

SISTEMIC CONGENITAL SYPHILIS CLINICAL AND BIOLOGICAL STUDY

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Abstract

One of the multisystemic infection transmitted to the fetus via the placenta is the congenital, syphilis caused by *Treponema pallidum*. High incidence of the disease led to routinely screening for all pregnant women. Clinical signs in neonatal period appear in the first 5 weeks of life, but signs of the disease may occur late, after the first 2 years of life. Diagnosis based on neonatal serologic testing is complicated by the transplacental transfer of maternal Ig antibodies, which can cause a positive test in the absence of infection. Three significant cases of congenital systemic syphilis treated in the Clinic of Neonatology are presented in this paperwork.

Key words: congenital syphilis, neonatal diagnosis.

Introduction

Congenital syphilis is a multisystemic infection caused by *Treponema pallidum* and transmitted to the fetus via the placenta.

The rate of transmission is higher in women with primary and secondary syphilis than in those with tertiary syphilis. Up to 40 % of pregnant women with untreated primary syphilis have presented spontaneous abortions.

High incidence of the disease led to routinely screening for all pregnant women.

Two-thirds of the new born with syphilis are asymptomatic at birth. Clinical signs of disease can occur during fetal period, neonatal period or later in childhood with an additional perinatal mortality of 25% - 30% cases. Without treatment, in severe forms of the disease, intrauterine death occurs in 25% of cases with an additional perinatal mortality of 25% - 30%.

Clinical signs in neonatal period appear in the first 5 weeks of life and they are: ulcerative skin lesions on the palms and sole (occur in severe forms of the disease and they are highly contagious),

hepatosplenomegaly, anemia, jaundice, hydrocephalus, lymphadenopathy, mucopurulent rhinitis, meningitis and mental retardation.

In septicemic forms, X-ray examination reveals metaphizal bone destruction and periosteal reaction.

Signs of the disease may occur late, after the first 2 years of life such as: frontal bossing, micrognathia, the palate pointed arch, Hutchinson’s triad, saddle nose, rhagades, optic atrophy which leads to blindness.

Because most infants born with congenital disease are free of clinical symptoms at the time of birth, final diagnosis is determined by laboratory tests. Most used are serological tests and direct fluorescent antibody test.

Diagnosis based on neonatal serologic testing is complicated by the transplacental transfer of maternal Ig antibodies, which can cause a positive test in the absence of infection. However a neonatal titer > 4 times the maternal titer would not generally result from passive transfer and diagnosis is considered confirmed or highly probable. Therefore evaluating the new born baby must follow these steps: historical data on maternal infection, physical examination, hemoleucograma, treponemic and nontreponemic serological tests, cardio-pulmonary and long bones X-ray and liver tests. *T. pallidum* can be identified in skin lesions, umbilical cord, placenta or autopsy.

Material and method

Study was conducted in the Clinic of Neonatology on three cases of congenital systemic syphilis which have been hospitalized in the same period. In the following we will present significant data of the three cases.

CASE 1

Patient MD, male, aged 2 weeks, delivered at term by normal vaginal route with green slimy amniotic fluid, IA at 5’-8, Bw = 1980 g.(birth weigh)

The patient was admitted with extremely severe condition, intense jaundice, facial cyanosis, marked abdominal distention, hepatosplenomegaly, repeated crises of apnea and cyanosis requiring oxygen mask. On the left lower limbs he presented a erythematous macular rash with a diameter between 0.5 -2 cm and with the tendency of spreading to the rest of the body.

Serological tests confirmed the suspicion of congenital syphilis and haematochemical investigations emphasize thrombocytopenia with leukocytosis and hepatocytolitic syndrome (TGO=123U/L, TGP=112U/L). All cultures were sterile.

After about a week of treatment (antibiotic + penicillin, etamsylatum, calcium, vitamins, plasma, dexamethasonum, arginine, aspatofort) CRP and aminotransferase values begin to decline slowly, jaundice gradually decreases in intensity, new born had a significant weight gain and clinical status in evolution was satisfactory (w = 3300g).

CASE 2

Patient K.S.- male new born baby, aged 1 day, weighing 2860 g, delivered at term by caesarean intervention, IA = 9,G1,P1.

On clinical examination the patient was noted to have a extremely severe condition, jaundice, perioronazal (facial) cyanosis, labored respiration with retraction of the intercostal muscles, ritmic heart sounds, pulse=130 beats / min. The abdomen was soft, it protruded durind inspiration, the edge of the liver was palpable approximately 2.5 cm below the right costal margin. Anterior fontanelle (2/3 cm) was normotensive.

Biochemical investigations have confirmed the diagnosis: systemic congenital syphilis (leuKocytosis, thrombocytopenia, CRP positive, VDRL and TPHA positive, increased LDH, hiperbilirubinemia).

Abdominal ultrasonography revealed increased liver volume, gallbladder with bold walls, normal biliary tract and normal spleen volume.

Treated with penicillin-10 days, in association with other antibiotics, and with ursodeoxycholic acid, calcium, vitamins and plasma, evolution is greatly improved with the exception of transaminase that increase progressively reaching a maximum at about a month of hospitalization, as follows: TGO = 535 U/l.

After this time, jaundice decreases in intensity and the liver size is progressively reduced.

CASE 3

Patient V.I. , Female, aged 1 day, delivered at term by normal vaginal route, with green amniotic fluid,

G2, P1, gestational age-33/34 weeks, polihidramnios, Bw = 2440 g, IA=5 at 1 minute, 6 at 5 minutes.

It was admitted in the first days of life with severe condition, cyanosis, petechial elements on the legs and body, saddle nose. Balanced cardio-pulmonary, pulse 132 beats / min, SaO₂-99%, abdominal distension caused by gas accumulation, the liver was palpable to the right iliac tank. Eruptive pustular papules on the abdomen and thorax.

Biochemical investigations (high leukocytosis, thrombocytopenia, hepatocitolitic syndrome: TGO = 210 U/L, TGP = 21 U/L, elevated inflammatory tests) and serological tests (VDRL, TPHA-positive) confirmed the diagnosis.

In evolution remain thrombocytopenia, leukocytosis, hepatocitolitic syndrome and intrainfectious anemia, beginning to improve after about 2 weeks from the onset.

Results and discussion

Although the literature does not mention an increased incidence of septicemic congenital syphilis, our clinic has faced, in a short period of time, 3 cases with similar clinical and biological features and also with a relatively good evolution despite data quoted in the literature that emphasizes a bad prognosis : death in 40% of cases.

Among the patognomonic features of the disease, common to our patients we mention:

- Intense jaundice, with elevated bilirubinemia. At first unconjugated bilirubina and in evolution, installed colestasis led both to increased conjugated bilirubin and also elevated levels of gGT and FA.
- Hepatomegaly accompanied by high levels of hepatic enzymes which after approximately one month start to decline under established treatment.
- Elevated values of inflammatory tests had been also present at all three patients and they slowly decreased under antibiotic therapy.

Treatment was conducted in accordance with the current protocols: penicillin for 10 days. It also has been done etiopatogenic treatment of concurrent infections, correction of acidobazic and hidroelectrolitic imbalances and correction of haematological disorders.

Evolution of the patient was initially serious: status toxicoseptic, needs for oxygen mask, positive serology, biological values considerably raised but then slowly becoming favorable, with improving of general status, jaundice remission and normalization of transaminases. The haematological parameters have also been corrected reaching values that corresponded to the patient's age.

Conclusions

1. The most commonly form of disease encountered in medical practice are asymptomatic, incidentally discovered by serologic test for syphilis.
2. All the cases of manifest syphilis had a serious evolution requiring special care and treatment.

3. Although the status of patients was extremely severe, joining various pathology and highly modified laboratory indices, the evolution under treatment was favorable.

References

1. Alford CA Jr, Cronic congenital and perinatal infections. in Avery GB editor, Neonatology, Pathophysiology and Management of the newborn, Philadelphia, JB Lippincott, 1987
2. Babcock D.S., Cranial ultrasonography of infants, Baltimore, Wiliams and Wilkins, 1981
3. Bale JF, Sato Y, Eisert D, Progressive postnatal subependimal necrosis in infant with congenital cytomegalovirus infections, *Pediatr Neurol*, 1986, 2, 367-370
4. Bains MK, Hosseini-Ardehali M. Palatal perforations: past and present. Two case reports and a literature review. *Br Dent J*. 2005;199:267–269. [PubMed]
5. Chawla W, Pandit PB, Nkrumach FK, Congenital syphilis in the newborn, *Arch Dis Child*, 1988, 63, 1393-1394
6. Dykes FD, Ahmann PA, Lazzara A, Cranial ultrasound in the detection of intracranial calcifications, *J Pediatr*, 1982, 100, 406-407
7. Platou RW, Hill AJ jr, Ingraham NR jr, Early congenital syphilis. Treatment of 252 patients with penicillin, *JAMA*, 1947, 133, 10
8. Lugo A, Sanchez S, Sanchez JL. Congenital syphilis. *Paediatr Dermatol*. 2006;23:121–123. doi: 10.1111/j.1525-1470.2006.00194.x.
9. Gurlek A, Alaybeyoglu NY, Demir CY, et al. The continuing scourge of congenital syphilis in 21st century: a case report. *Int J Paediatr Otorhinolaryngol*. 2005;69:1117–1121. doi: 10.1016/j.ijporl.2005.03.007.
10. Tikhonova L, Salakhov E, Southwick K, for the Congenital Syphilis Investigation Team. et al. Congenital syphilis in the Russian Federation: magnitude, determinants and consequences. *Sex Transm Infect*. 2003;79:106–110. doi: 10.1136/sti.79.2.106. [PubMed]
11. Li, Y.; Gonik, B. Is congenital syphilis really congenital syphilis. *Infect Dis Obstet Gynecol*. 2006. pp. 1–4. Article ID 81629.
12. Humphrey MD, Bradford DL. Congenital syphilis: still a reality in 1996. *Med J Australia*. 1996;165:382–385. [PubMed]

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