A CASE REPORT OF CONGENITAL NEPHROTIC SYNDROME SECONDARY TO CONGENITAL SYPHILIS

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ABSTRACT

The incidence of congenital nephrotic syndrome secondary to congenital syphilis is very uncommon in clinical practices these days. We present a case of a two-month-old girl, who admitted with fever, cough, distended abdomen. The laboratory informed normal kidney function with Albumin 11.9 g/l; Protein 43.6 g/l; Cholesterol:1.64 mmol/l, Proteinuria 2.8g/l, urine Protein/Creatinin = 10g/mmol, VDRL (+), TPHA (+). Abdominal ultrasound showed ascitis and hepatosplenomegaly. The patient was diagnosed as congenital nephrotic syndrome secondary to congenital syphilis. She was prescribed Albumin infusion, Diuretics, Penicillin. She presented good evolution and the syndrome was solved after 2 weeks of treatment.

Keywords: syphilis, nephrotic syndrome.
OVERVIEW

I. CONGENITAL NEPHROTIC SYNDROME

Nephrotic syndrome is the most common glomerular disease in children, caused by the loss of blood protein through glomerular filter membrane due to many different causes. Typical manifestations of this syndrome are: edema, high urinary protein (> 50mg/kg/24 hours or urinary protein/urinary creatinine > 200mg/mmol), serum protein < 56g/l and serum albumin < 25g/l, accompanied by increased lipid and serum cholesterol [3].

When these symptoms start in the first 3 months of a child's life, it is called congenital nephrotic syndrome. This form is very rare and divided into 2 different groups based on pathogenic mechanism [3]:

- Primary congenital nephrotic syndrome: due to mutation of genes encoding glomerular filter membrane components, common genes are: NPHS1, NPHS2, WT1, LAMB2, LAMB3...

- Secondary congenital nephrotic syndrome: after some infections in the neonatal period such as syphilis, Toxoplasma, Cytomegalovirus, HIV, hepatitis B...

Congenital syphilis that causes congenital nephrotic syndrome has been known for a long time. The incidence of nephrotic syndrome in syphilis patients is quite low, usually under 3% according to different studies. Sopana Niemsiri studied 455 children with congenital syphilis at Bangkok children's hospital, Thailand from 1977-1991 and found 11 children with nephrotic syndrome, incidence of 2.4% [4].

In addition to characteristic manifestations of nephrotic syndrome, manifestations of nephritis may also occur. Patients often have urinary protein and hematuria but manifestations of severe nephrotic syndrome is less common. This syndrome may appear in the neonatal period but is common in the period of 1 to 4 months of age. Common pathological damage is membrane kidney disease. It is treated with Penicillin, if permanent kidney damage has not been formed [3].

II. CONGENITAL SYPHILIS

Syphilis is a disease that is transmitted sexually, through blood and from mother to child during pregnancy, caused by Treponema pallidum.

Congenital syphilis is the result of transmission from mother-to-child, in utero through placenta or through vaginal tract when the baby was born, the mother infected and not treated appropriately before and during pregnancy.

1. Clinical manifestations

1.1. Early congenital syphilis: clinical manifestations that appear before 2 years of age. Most clinical symptoms in untreated children appear within the first 3 months of life (usually in the first 5 weeks) [2].

About 60-90% of newborns with congenital syphilis will have no symptoms at birth. In children with symptoms, they may be

- Large liver and / or large spleen
- Jaundice
- Release of nasal discharge: clear fluid, or possibly fluid with blood, pus. Release usually lasts.
- Skin rash: pink or red maculapapular rash.
- Whole body lymph nodes.
- Skeletal abnormalities: Abnormal X-ray of long bones occupies 60-80%. Usually affected bones are femur, arm bone, leg bone.
- Nephrotic syndrome
- Central nervous system: Syphilis infection of the central nervous system may or may not be symptomatic. Non-asymptomatic form is manifested by abnormalities of cerebrospinal fluid. Symptomatic form is rare in children, but may develop in children who are not treated at the neonatal stage, including two clinical forms: acute syphilis meningitis (with manifestations of vomiting, fontanelle swelling, increased head loop, increased cell, cerebrospinal fluid protein) or chronic vestibular-meningitis syphilis (hydrocephalus, cranial nerve paralysis, papilloedema, optic nerve atrophy, nervous retardation, convulsion).
- Respiratory system: fuzzy inflammatory lesions in two lung fields are characteristic images of pneumonia in congenital syphilis patients.
- Blood system: anemia, thrombocytopenia, increased or decreased leukocytes [2].

1.2. Late congenital syphilis: conjunctivitis, deafness, Hutchinson's teeth, protruding forehead, nasal septum, mental retardation [2].
2. Definition of case

Under the guidance of the US Centers for Disease Control (CDC), congenital syphilis is defined as follows:

2.1. Can be infected if:

- The child is a child of a mother with syphilis who is not treated or treated ineffectively until the time of childbirth, regardless of the child's symptoms, or
- The child has serological tests for positive syphilis such as VDRL, rapid plasma reagin (RPR), and one of the following criteria:
  • Any manifestations of congenital syphilis on examination (mentioned above).
  • Any manifestations of congenital syphilis on long bone film.
  • VDRL serum test in cerebrospinal fluid is positive.
  • Increased leukocytes or protein in the cerebrospinal fluid (excluding other causes) [1].

2.2. The case is diagnosed as congenital syphilis when Treponema pallidum is detected by darkfield microscopy, fluorescent antibody or other specific staining of lesions, placenta and umbilical cord... [1].

CASE

The patient was a 2-month-old female child, the second child, giving birth normally, premature birth 33 weeks, birth weight 2.8kg. After birth, the child did not show any abnormalities. The child's father was 33 years old, Chinese, doing business; the child's mother was 25 years old, Vietnamese, working as a housewife.

From 1 month of age, the mother saw the baby appear release of clear nasal discharge, sometimes with phlegm cough. In addition, the mother saw the baby's belly becoming bigger and bigger. About 3 days before being admitted to the hospital, the child had a lot of cough with mild fever, loose stool 6 times/day, yellow stool without bloody mucus.

Examination at admission: The child was alert, tired, had a fever of 39 degrees C. Pale skin. Unknown face edema, dyspnea, concave chest breathing, breathing rate 60 times/minute, SpO2 93%. Two lungs had moist rales. Regular heart rate. Abdominal distention, clear collateral line circulation, unknown liver and spleen. Soft tumor in the right groin area suspected by hernia.

Tests: Blood leukocytes: 30200, neutrophils: 66%, HGB: 75gl, platelets: 159000. Blood biochemistry: Albumin 11.9g/l; Total protein 43.6g/l; Cholesterol: 1.64 mmol/l; Tryglicerid: 0.88 mmol/l, GOT: 67 UI/l, GPT: 36 UI/l. The first sample urinary protein was 2.8g/l, then quantified the urinary protein/urinary creatinine ratio = 10g/mmol. Basic coagulation test gave normal results. Stool test was negative. Chest X-ray: blurred images of two lung fields consistent with Pneumonia. Abdominal ultrasound: much free abdominal fluid, multiple sound reduction blocks in the liver, 16mm large block, in the heart of the liquidous necrosis hole; large liver and spleen. Abdominal CT: scattered images of reducing the proportion of posterior right lobe liver parenchyma, after injection without infusion.

The patient was diagnosed with Pneumonia - Anemia - Liver abscess - Congenital nephrotic syndrome. The child was indicated for infusion of erythrocytes, infusion of albumin and antibiotics Cefoperazol in combination with Metronidazol for the treatment of pneumonia - liver abscess. Shortness of breath, fever, loose stool gradually stabilized in the first 3 days of treatment. Also tests to find out the causes of congenital nephrotic syndrome were indicated. The results were HBsAg, anti-HCV, HIV negative, CMV 4610 copies/ml, VDRL positive, TPHA positive twice (ratio of 1: 10246). Parents also had VDRL tests, the father gave positive results and the mother gave negative results, but the mother told that after giving birth she used an antibiotic for 5 days (unknown drug name).

With the patient’s clinical and test results, based on the diagnostic criteria mentioned above, we concluded that the child suffered from congenital syphilis and secondary congenital nephrotic syndrome caused by syphilis. The child was treated with Penicillin G at a dose of 200 000 UI/l for 10 days and Captopril. After 2 weeks of treatment: blood Albumin 38.6 g/l, 64.3 g/l, urinary protein/urinary creatinine 40mg/mmol. In addition, abdominal ultrasound showed no hepatosplenomegaly, normal liver parenchyma, normal hemoglobin. Lungs no longer had lesions. The patient was scheduled for re-examination after 1 month of treatment, the results of blood biochemistry were within normal limits, abdominal ultrasound was normal, urinary protein/urinary creatinine was
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50mg/mmol (this value is normal for 3-month-old children), the patient had no abnormal manifestations of the nervous system, normal mental and movement development.

We gave patients with a X-ray of long bones and found normal results, hearing and eye examination and found no abnormalities. The patient indicated for cerebrospinal fluid puncture and kidney biopsy but because the family did not agree to do the procedure (with the main reason that the patient was small), we could not evaluate the damage of the nervous system caused by syphilis (although the child did not have neurological symptoms) as well as histopathological damage of kidney caused by syphilis in this patient. However, the patient's complete response to penicillin treatment supported the diagnosis of secondary kidney failure caused by syphilis.

DISCUSSION

This patient was admitted in the status of infection (pneumonia, liver abscess) with gastrointestinal disorder. In this patient, a large amount of peritoneal effusion accompanied by biochemistry with blood albumin and blood protein decreased significantly in the nephrotic threshold, while basic coagulation was normal, urinary protein was high in two tests, although blood cholesterol and triglycerides were normal, in a 2-month-old female child, it was eligible to conclude congenital nephrotic syndrome. Infection status (pneumonia, liver abscess), gastrointestinal disorder can be explained by complications of nephrotic syndrome as well as congenital syphilis. According to the 1991 study by Sopana Niemsiri Bangkok Children's Hospital, Thailand in 11 children with nephrotic syndrome secondary to congenital syphilis, it was also found that 4 out of 11 children (36%) had normal blood cholesterol, 4 out of 11 children (36%) had complications of pneumonia, 3 out of 11 children (27%) had infection and septicemia, 3 out of 11 children (27%) had hepatitis [4].

The patient had manifestations of hepatosplenomegaly, anemia, nephrotic syndrome, pneumonia accompanied by positive VDRL test; positive THPA test twice at a ratio of 1:10246, according to the above definition of case, the patient was eligible to be diagnosed with congenital syphilis. In addition, the father's VDRL test gave positive results and the mother's gave negative results; but because the mother had a pre-history of antibiotic use after birth, their enlargement may be suspected by the mother who was infected but treated. The patient's response to penicillin treatment (indicators for blood test, urine test, abdominal ultrasound) was also an additional support for the diagnosis of syphilis in this patient.

Unfortunately, in this patient, we did not perform a kidney biopsy to know pathological damage because the family did not agree, but we hope the patient's kidney damage has been treated and does not lead to permanent kidney damage.

This patient also did not have a CSF test because the family did not agree. Until now, this patient has no neurological abnormalities. We will continue to follow-up for clinical and subclinical developments of the patient.

CONCLUSION

Manifestations of nephrotic syndrome in congenital syphilis patients are rare.

Penicillin is still the standard treatment of syphilis so far. When treated early, kidney damage will be completely restored.

REFERENCES