

PERTUSSIS; STILL A CLINICAL DIAGNOSIS

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Pertussis is an acute, contagious respiratory infection characterized by paroxysmal coughing episodes. The most important and severe complication in infants is respiratory failure due to pneumonia. We report the case of a 3-month-old infant without any immunization who presented with cough, whooping and cyanosis, and developed pneumonia on the second day and a leukemoid reaction on the sixth day of admission. The relation between pneumonia and mortality, especially in infants, and the prognostic impact of a leukemoid reaction are emphasized.

Key Words: Pertussis, Leukemoid Reaction, Mortality.

BOĞMACA; KİLİNİK TANININ ÖNEMİ

Boğmaca, öksürük nöbetleriyle karakterize akut, bulaşıcı bir solunum yolu enfeksiyonudur. Bebeklerde en önemli ve ciddi komplikasyonu pnömoniye bağlı solunum yetmezliğidir. Nöbetlerle boğulur tarzda öksürük ve siyanoz ile başvuran, hiç aşı olmamış, yatışının ikinci gününde pnömoni, altıncı gününde lökomoid reaksiyon geliştiren üç aylık bebek sunulmuştur. Özellikle bebeklerde pnömoninin mortalite ile ilişkisi ve lökomoid reaksiyonun prognostik önemi vurgulanmıştır.

Anahtar Kelimeler: Boğmaca, Lökomoid Reaksiyon, Mortalite.

INTRODUCTION

Pertussis is one of the most highly communicable infectious diseases. It begins with mild upper respiratory symptoms and is characterized by classic severe, repetitive, progressive coughing and whooping episodes, post-tussive vomiting and poor feeding. Mostly it is caused by a Gram-negative rod called *Bordetella pertussis* (1-3). Absolute lymphocytosis is characteristic, which is sometimes accompanied by a specific leukocytosis called a leukemoid reaction ($> 50,000$ cells/ μ l). Although the time of onset of the leukemoid reaction is not properly defined, its severity is associated with poor prognosis (4). Vaccination has considerably reduced both the disease incidence and the severity of this reaction (1,2).

Particularly infants, who are prone to other respiratory infections, develop less recognizable and indistinguishable symptoms. Making an accurate diagnosis in this group needs awareness of the disease, and aggressive efforts are essential to identify both the cases and the contacts.

CASE REPORT

A three-month-old girl was admitted with nonproductive, progressive, whooping cough followed by vomiting, brief apnea and cyanosis. While having coryza-like symptoms during the previous week, she began to have 15-20 episodes of coughing and cyanosis was noted in the previous 2 days. It was learned that she had had a varicella infection for two months and had received no vaccination at all. On physical examination she looked exhausted, with mild fever, tachypnea, intercostal retractions and circumoral cyanosis. Auscultation of the chest showed rough breath sounds and bilateral fine crackles at the base. On admission, her white blood cell count (WBC) was $15.3 \times 10^9/L$ with predominant lymphocytes (80%), and oxygen saturation was 84% (pulse oximetry); flattening of the ribs and mild inflation were noted on her chest X-ray. Despite supportive treatment on the second day she deteriorated with an increase in the severity and frequency of the episodes and respiratory distress. The blood picture at that time was unremarkable, while perihilar granular infiltration and atelectasis in the right upper zone were observed on her chest X-ray. Although the results of nasopharyngeal culture and IgG and IgA antibodies against *B. pertussis* (ELISA) were pending, based on clinical findings she was diagnosed with pneumonia due to pertussis, and clarithromycin 15 mg/kg/d and salbutamol were added to the treatment. On the sixth day, while receiving the above treatment, she was found to have an increase in WBC ($89 \times 10^9/L$) with predominantly lymphocytes. At that time although the diagnosis of pertussis was confirmed by the laboratory results and this blood picture was thought to be a lymphoid leukemoid reaction consistent with the disease, a

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bone marrow aspiration was performed, which showed hypercellularity with a predominance of lymphoid cells and without any evidence of any malignancy. Then 3000 cc/m² fluid was added to the treatment and WBC was monitored daily, which remitted, with a gradual decrease to 17 x 10⁹/L within one week (12th day of admission) (Table 1).

On day 15 she was discharged with normal physical examination findings, normal WBC and resolution of pneumonia on X-ray.

Table 1. White blood cell counts (WBC) of the patient.

Day	WBC (x 10 ⁹ /L)
1 (admission)	15.3
6	89
7	84
8	79
9	63
10	57
11	52
12	48
13	35
16	24.8
19	17.7

DISCUSSION

Pertussis, which is a serious lower respiratory infection, is more common and more severe particularly in infants who are non- or incompletely immunized (5-8). Paroxysmal coughing is the major symptom in the clinical diagnosis. Whole cell pertussis vaccine in four doses, with an efficacy of 70%–90%, prevents severe disease (1). Nasopharyngeal culture, specific agglutination tests and polymerase chain reaction could be used to confirm the diagnosis (9-11). Leukocytosis and typical absolute lymphocytosis may accompany the disease, mostly at the beginning. Leukocytosis may be exaggerated as a special finding called leukemoid reaction, especially in nonimmunized patients, which can be explained by the exposure to higher doses of the toxins. Extreme leukocytosis and accompanying thrombocytosis are noted in many fatal cases (4). The mortality rate is below 1%, but it can be as high as 40% in infants less than 5 months old. Death is usually due to pneumonia or other respiratory complications like asphyxia or encephalopathy, and the leukemoid reaction is related to poor prognosis in non- or incompletely immunized infants (4,8).

Since there is no exact time for the onset of the leukemoid reaction in pertussis it can be challenging to make a differential diagnosis, like in our patient. As the typical clinical and laboratory findings like leukocytosis may not be present in infants the diagnosis may be missed or delayed (7,10,11). Furthermore, it must also be taken into account that both serologic

and culture results are obtained a few days later, which might be important in treating a patient especially as young as ours.

As our patient was an infant who had no immunization with repetitive, paroxysmal coughing episodes, absolute lymphocytosis, pneumonia and positive culture for pertussis, it was easy to make the diagnosis. However, as the WBC was higher than expected and it seemed to be a little late for a real leukemoid reaction associated with pertussis, serology and culture results as well as a bone marrow aspiration were needed to confirm the ultimate diagnosis. As in our patient, for those with paroxysmal cough, one should have a high index of suspicion for pertussis and daily monitoring of leukocytosis is necessary for a timely diagnosis and treatment.

After 1974, the incidence, mortality and morbidity of vaccine-preventable diseases in developed countries decreased due to enlarged immunization programs guided by WHO. This ultimate goal was also considered in our country with the beginning of national vaccine campaigns in 1985. However, it still has not been achieved, as found in the Demographic and Health Survey held in 2003. It is reported that despite the fact that there has been an improvement (9% increase compared to the previous survey held in 1998) in the percentage of fully immunized children, 3% of children aged 12-23 months never received any vaccine. Moreover, it is also found that 25% of children who received the first dose of the vaccine did not complete the other doses, which is the sine qua non concerning prevention (12). This shows the importance of maintaining and following full coverage vaccination programs in childhood as occurred in our patient; if ever postponed, the vaccine must be given at as early an age as possible.

In conclusion, although the presentation of our patient, with the exception of the late onset of the leukemoid reaction, was consistent and clinical suspicion is important in the diagnosis of pertussis, we emphasize the necessity of performing all the diagnostic studies in all hospitalized infants with signs of lower respiratory infection and daily monitoring of leukocyte counts, especially in nonimmunized patients, to make a differential diagnosis and to predict and overcome poor prognosis.

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