An unusual case of acute rheumatic fever presenting with unilateral pulmonary edema

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The differential diagnosis of acute rheumatic fever (ARF) in children, especially those younger than five years, may be difficult, even with strict application of the updated Jones criteria. They are likely to present with atypical symptoms that can mimic various conditions. Herein we report a 27-month-old girl with ARF presenting unilateral pulmonary edema secondary to severe mitral regurgitation. Taking into account atypical clinical presentations in these younger ARF patients will prevent the delay in the diagnosis and will have an impact on treatment to reduce morbidity and mortality of the disease.

Key words: acute rheumatic fever, pulmonary edema, mitral regurgitation.

Acute rheumatic fever (ARF) is a systemic illness that may occur in children following group A beta hemolytic streptococcal pharyngitis. The incidence of ARF and rheumatic heart disease has not decreased in developing countries. The retrospective studies demonstrate the highest figures for cardiac involvement and recurrence rates. Therefore, ARF appears to be a complicated health problem in these countries[1,2].

The modified Jones criteria provide guidelines for making the diagnosis of ARF. There are five major criteria, such as carditis and polyarthritis, and four minor criteria including clinical and laboratory features. The presence of two major or of one major and two minor criteria indicates a high probability of ARF if supported by evidence of preceding group A streptococcal infection[3]. Even with strict application of the Jones criteria, overdiagnosis as well as underdiagnosis of ARF may occur.

Acute rheumatic fever is uncommon in children younger than five years, with only approximately 5% of children with ARF under five years of age at presentation. These patients are more likely to show atypical presentation[4]. When present, cardiac involvement is more often moderate to severe (as opposed to mild). Herein, we report a 27-month-old girl with ARF presenting unilateral pulmonary edema secondary to severe mitral regurgitation.

Case Report

A 27-month-old girl was referred to the hospital with a five-day history of tachypnea and dyspnea. She had upper respiratory tract infection two weeks earlier and was treated with procaine penicillin for seven days. Despite the treatment, her symptoms worsened and she was hospitalized in a local clinic because of bronchopneumonia two days later. On her previous admission, chest X-ray revealed right pleural effusion. She was administered sulbactam-ampicillin and amikacin. However, her clinical condition progressively worsened, and she was referred to our hospital. Her parents also mentioned that she had knee pain one month earlier that caused difficulty in walking and disappeared spontaneously after a few days. However, no clinical sign of arthritis was present. Otherwise, her past medical history was unremarkable.

On admission, she appeared ill with a temperature of 37°C, respiratory rate 66/min, heart rate 162/min and blood pressure 92/67 mmHg. Her chest examination revealed right-sided fine inspiratory crackles. Cardiac examination showed pansystolic murmur on the apex radiated over the precordium. The liver was palpated 3 cm below the costal margin and there was tenderness on her right ankle.

Complete blood count showed the following: hemoglobin (Hb), 9.3 g/dl; white blood cell count, 12500/mm^3; mean corpuscular volume, 75 fl; hematocrit, 29%; platelet, 305000/mm^3; erythrocyte sedimentation rate, 40 mm/h; and elevated antistreptolysin O (ASO) titer, 193 IU/ml. Her serum electrolyte, creatinine and liver enzyme levels were all normal. Chest X-ray showed cardiomegaly and patchy alveolar infiltrates on the right lung consistent with pulmonary edema. Echocardiographic evaluation revealed severe mitral regurgitation (4+) and mild aortic valvular regurgitation. Throat swab and blood cultures were negative. Anti-DNase B was 401 E/ml (normal: <188 E/ml). Anti-nuclear antibodies (ANA), rheumatoid factor and anti-ds-DNA were negative.

A diagnosis of ARF was made and prednisolone therapy was started for four weeks. She responded to the therapy and her pulmonary symptoms improved within one week. She is now four years old and very well without any clinical symptom. Her echocardiographic examination revealed mild mitral regurgitation. She has been on benzathine penicillin administration for secondary prevention.

Discussion

Acute rheumatic fever is an immunologically mediated sequela of group A streptococcal infection, which can affect a number of different tissues such as heart, joint and brain. ARF can mimic many other diseases of various organs. The diagnosis of ARF is based on certain clinical criteria. However, it is still underdiagnosed as well as overdiagnosed in different settings. In contrast to older patients, children younger than five years are more likely to present with unusual clinical findings. In addition to moderate to severe carditis, pulmonary edema associated with mitral valve regurgitation is usually bilateral and of cardiac origin[5,6]. As seen in the present case, a misdiagnosis of pneumonia is often made, since the possibility of cardiac disease usually remains obscure.

Schnyder et al.[5] reported a 9% prevalence of predominantly right upper lobe pulmonary edema (UPE) in ARF cases with severe mitral valve regurgitation. However, other causes of UPE should be considered in the differential diagnosis. Therefore, we ruled out other conditions in our patient by considering that she had no clinical history consistent with previous unilateral lung disease, no history of prolonged lateral decubitus position, no hydrothorax or pneumothorax, and no invasive catheter insertion.

Because no clinical and laboratory finding is pathognomonic for ARF, the diagnosis can be established by the Jones criteria and an absolute requirement for evidence of antecedent group A streptococcal infection. Positive throat cultures are obtained only in about 11% at the time of presentation[8]. In contrast, the detection of ASO in the serum of the patient or an increase in its titer is usually a strong indicator of recent streptococcal infection. ASO, an internationally standardized test, is widely used in the detection of group A streptococcal infections and their sequelae. Elevated or rising titers are seen in 80% or more of the cases with ARF. However, ASO titers can vary depending on the geographic location, age group of the study population and the climatic conditions. A positive test result can be achieved in 95-100% of the cases, if three different antibodies (ASO, anti-DNase B, antihyaluronidase) are measured. Therefore, it is proposed that when ARF is suspected clinically, multiple antibody tests should be performed[3].

In conclusion, we suggest that UPE secondary to severe mitral regurgitation may be an atypical presentation of the clinical picture of ARF in children younger than five years. Therefore, even with strict application of the updated Jones criteria, the diagnosis of ARF may be difficult, especially when carditis is an isolated manifestation of the disease. However, such an atypical presentation should be taken into account and this approach will prevent the delay in the diagnosis and will have an impact on treatment to reduce morbidity and mortality of the disease.

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