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Infective Endocarditis Caused by Odontogenic Infection with Dentinogenesis Imperfecta in Jeune Syndrome

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Abstract

Osteogenesis imperfecta is main symptom in an autosomal recessive Jeune syndrome. Odontogenic infection by dentinogenesis imperfecta and poor oral hygiene is not known and Infective Endocarditis (IE) as a life threatening complication is the first report in Jeune syndrome.

Case: A 13-year-old female patient presented with fever, disturbed consciousness, and convulsion. She had Jeune syndrome with mitral regurgitation and mental retardation. Cerebral hemorrhage, vegetation of mitral valve and poor oral hygiene demonstrated clinical definite diagnosis of IE by Duke Criteria. Bacterial aneurysm would cause to be cerebral hemorrhage that was no worsening by Computed Tomography. Vancomycin or Linezolid was treated after MRSA was detected in blood culture. Decayed teeth were removed as source of IE. Consciousness level and inflammation response were improved and no sign of infection was confirmed by frequent echocardiogram and blood culture.

Outcome: Patient was discharged the hospital after she had no symptoms with appropriate treatment.

Conclusion: Observation of oral hygiene and consideration of IE as the differential diagnosis are beneficial in a severe infectious disease of unknown origin with osteogenesis imperfecta, Jeune syndrome. Antibiotics therapy, oral hygiene and teeth extraction are effective for IE caused by decayed teeth with dentinogenesis imperfecta.

Keywords: Cerebral hemorrhage; Infective endocarditis; Jeune syndrome; Odontogenic infection

Introduction

Jeune syndrome, or asphyxiating thoracic atrophy, is an autosomal recessive skeletal dysplasia with multi-organ involvement. It is characterized by a narrow thorax and short limbs at birth by osteogenesis imperfecta. The small bell-shaped thorax often results in respiratory distress and recurrent infections in neonatal period and infancy. The outcome of Jeune syndrome is fetal in early childhood (60-80%). Liver, pancreas, renal and retinal abnormality have also been reported as a multisystem disorder. The thoracic malformation tends to be less severe with age and less respiratory infection [1-4].

In the treatment of severe infectious diseases, it is difficult to identify the specific origin of the infection. Therefore, empiric antimicrobial therapy often tends to be selected. Regarding unknown origin of infection, the condition of oral hygiene may provide an important clue to the pathogenesis of infectious disease. A history of valvular disease is significantly associated with Infective Endocarditis (IE), which could be lethal disease if the diagnosis and treatment are delayed. Taken together, when a poor oral hygiene is found in patients with vulvular disease, IE should be suspected.

Dentinogenesis imperfecta associated with osteogenesis imperfecta [5] can lead to a poor oral hygiene. However, there have been few reports on Jeune syndrome, and the clinical information of oral and systemic health is uncertain. We herein report for the first time on IE caused by odontogenic infection in a 13-year-old female with Jeune syndrome.

Case

A 13-year-old female patient presented with fever, decreased level of consciousness, and convulsion in one day before admission. She had Jeune syndrome, mitral regurgitation, and mental retardation in the medical history. Funnel chest (Figure 1a) and scoliosis developed due to osteogenesis imperfecta of Jeune syndrome. She was transferred to our intensive care unit after CT examination which revealed cerebral hemorrhage (Figure 1b). The level of consciousness was assessed as E4V1M4 using the Glasgow Coma Scale on day 1. She also presented with neck stiffness and systolic murmur. Vital signs were shown in the following: heart rate 137 b.p.m. (regular rhythm); blood pressure, 120/63 mmHg; body temperature, 39.0 oC; respiratory rate 50/min and percutaneous oxygen saturation: 96% on room air. Laboratory data indicated acute inflammation and post-convulsion; White blood cell: 19,000/μL, C-reactive protein: 17.43 mg/dL, Procalcitonin: 86 ng/ml and Creatine kinase: 1709 IU/L. Oral environment was seriously poor with decayed teeth (Figure 2a). Bacterial meningitis was suspected initially by neurogical symptom, and Meropenem (MEPM) and Cefotaxime (CTX) were administered as empiric therapy on day 1. Because cerebral hemorrhage was not progressive by CT on ICU day 2, conservative therapy without operation was selected. As her clinical history of mitral regurgitation and high Brain natriuretic peptide: 451.5 pg/mL, were observed, IE was suspected as an infectious source. Echocardiography identified vegetation of mitral valve (Figure 1c).

One major Duke Criteria (vegetation by echocardiography) and 3 minor criteria (mitral regurgitation, fever and cerebral hemorrhage) resulted in clinical definite diagnosis of IE [6]. After MRSA was detected

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in blood cultures, MEPM was switched to Vancomycin (VCM). Her level of consciousness was recovered and she was discharged ICU to a general ward in day 5. Linezolid (LZD) was administered instead of VCM because of renal dysfunction on day14 and continued on day 49. Inflammatory marker and fever was improved on day 14. Other antibiotics, such as Tazobactam piperacillin (TAZ/PIPC), MEPM or CTX, were administered for 22 days. Decayed teeth were suspected mostly as the source of infection, and they were extracted on day 70 (Figures 2b and 2c). We identified no bacterial infection in subsequent blood culture 5 times and also no change of vegetation of mitral valve was confirmed. She was discharged hospital without unfavorable symptoms on day 92.

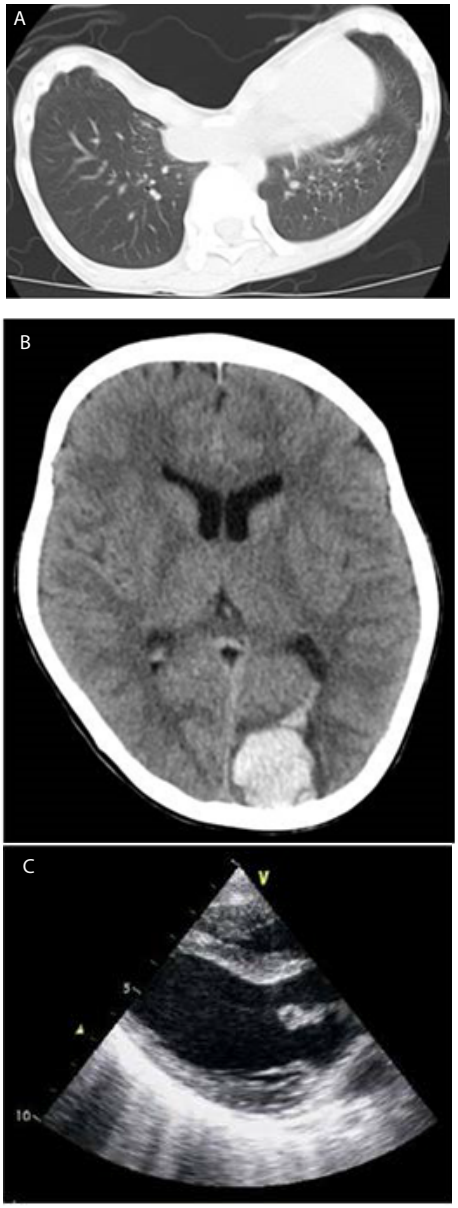

Figure 1: a) Identification of funnel chest, cerebral hemorrhage and mitral vegetation by CT examination and echocardiography. a: Funnel chest in Jeune syndrome.

b) Cerebral hemorrhage was evaluated by CT.

c): Vegetation of mitral valve was detected by echocardiography.

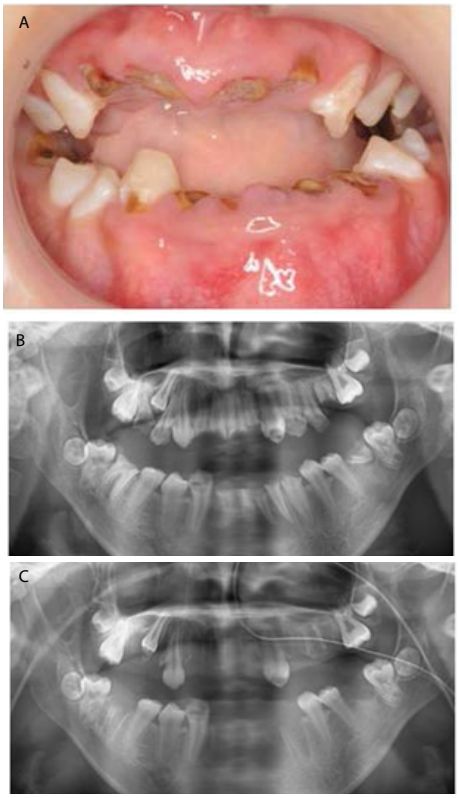

Figure 2:a) Multiple decayed teeth by poor oral hygiene were observed

b) Poor oral hygiene and extraction of decayed teeth by X-ray.

c) X-ray demonstrated decayed teeth before and after extraction.

Discussion

We identified two beneficial clinical issues. Dentinogenesis imperfecta could be associated with osteogenesis imperfecta of Jeune syndrome. IE was caused by odontogenic infection with decayed teeth and dentinogenesis imperfecta. First, dentinogenesis imperfecta could be associated with osteogenesis imperfecta of Jeune syndrome. Jeune syndrome is an autosomal recessive disease, and IFT 80 has been identified as a responsible gene.

The patients with Jeune syndrome mainly present osteogenesis imperfecta exhibiting an abnormality of the thorax, such as funnel chest, and limb abnormalities. It has been suggested that this disease could be associated with imperfect hard tissue formation including dentinogenesis imperfecta. In the severe cases, it is reported that the patients suffer from respiratory or multiple organ failure [1-4]. No serious respiratory complications were observed in this patient with funnel chest (Figure 1a).

Second, IE was caused by odontogenic infection with decayed teeth and dentinogenesis imperfecta.

The intraoral inflammation by poor oral hygiene such as gingivitis can give the opportunity that pathogenic microorganisms easily penetrate into blood vessels in the daily life [7]. In the present case, multiple decayed teeth associated with dentinogenesis imperfecta were considered mostly as the source of infection (Figure 2). Other cause of multiple decayed teeth was caused by a poor oral daily care based on mental retardation. MRSA was the target of bacteria mainly for IE in this case after blood examination. She had chance to exposure

MRSA in hospital and health care institution during frequent visits for taking care of disease. The condition of patient might be low nutritional or immunologic state, indicating compromised host. A higher prevalence of amoxicillin-resistant oral streptococci in dental plaque specimens from Japanese child and adolescents at risk for IE [8]. Cerebral hemorrhage and systolic murmur with fever elevation pushed us to diagnose IE by echocardiography (Figures 1b and1c). Bacterial aneurysm would cause to be cerebral hemorrhage. It has been reported that IE is accompanied with brain complications in ratio of 20-40%, and the brain complications as the initial symptoms in IE was high ratio of 47% [9].

Conclusions

Observation of oral hygiene and consideration of IE as the differential diagnosis are beneficial in a severe infectious disease of unknown origin with osteogenesis imperfecta, Jeune syndrome. Antibiotics therapy, oral hygiene and teeth extraction are effective for IE caused by decayed teeth with dentinogenesis imperfect [10].

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