Anesthetic Management in a Case of Dyskeratosis Congenita

Diskeratozis Konjenitalı Bir Hastada Anestezi Yönetimi

Abstract

The most severe complications developed in patients with dyskeratosis congenita (DKC) are bone marrow failure (BMF), difficult airway, and pulmonary fibrosis. Accordingly, in DKC patients to be operated, the clinical and laboratory data should be evaluated for presence of BMF-related pancytopenia, difficult airway and opportunistic infections, and the systemic examination prior to the administration of general anesthesia should be made carefully. The preoperative preparation should include thorough examination of the lungs as well as assessment for postoperative respiratory distress. The operation should be attempted when the patient's clinical and laboratory values are optimal. Supportive equipment should be prepared in advance for patients expected to suffer difficult airway. This paper will discuss the preoperative preparation and anesthetic management in a pediatric case of DKC complicated with bone marrow failure where the patient was scheduled for dental treatment under general anesthesia.

Keywords: dyskeratosis congenita; anesthetic management

Öz

Diskeratozis konjenita (DKC) hastalarında gelişen en ciddi komplikasyonlar, kemik iliği yetmezliği (KİY), zor havayolu ve pulmoner fibrozdur. Bu nedenle, opere edilecek DKC hastalarında hastanın klinik ve laboratuvar verileri KİY ile ilişkili pansitopeni, zor havayolu ve fırsatçı enfeksiyon varlığı bakımından değerlendirilmeli ve genel anesteziden önceki sistemik muayene dikkatle gerçekleştirilmelidir. Preoperatif hazırlıklar kapsamında akciğer ayrıntılı muayene edilmeli ve hasta postoperatif solunum sıkıntısı açısından da değerlendirilmelidir. Operasyona hastanın klinik ve laboratuvar değerleri optimal olduğunda girişilmelidir. Havayolu problemleri beklenen hastalar için yardımcı ekipman hazır bulundurulmalıdır. Bu yazıda genel anestezi altında diş tedavisi planlanan, KİY ile komplike bir pediyatrik hastada gerçekleştirilen preoperatif hazırlık ve anestezi yönetimi tartışılacaktır.

Anahtar Sözcükler: diskeratozis konjenita; anestezi yönetimi

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INTRODUCTION

Dyskeratosis congenita (DKC) is a rare disorder that usually shows autosomal recessive inheritance. The typical clinical manifestations include nail dystrophy, reticular skin pigmentation, and oral leukoplakia. The most severe complications developed in patients with DKC are bone marrow failure (BMF) and pulmonary fibrosis (1). Particularly, the masses in the airway can cause intubation difficulty. In addition, dental caries, gingiva hyperplasia, alveolar bone loss, and malignant neoplasms are seen in the mouth frequently. Gingiva hyperplasia can cause bleeding of fragile gingival lesions during intubation (2,3). Accordingly, in DKC patients to be operated, extensive preoperative examination of the mouth is necessary. Also, the clinical and laboratory data of the patient should be assessed and the systemic examination prior to administering general anesthesia should be made carefully.

This paper will discuss the preoperative preparation and anesthetic management in a pediatric case of DKC complicated with bone marrow failure where the patient was scheduled for dental treatment under general anesthesia.

CASE REPORT

A seven-year-old boy presented at our hospital for dental treatment. The treatment was planned to be under general anesthesia since he could not cooperate with the dentist. Oral and systemic examination was performed. The relatives of the patient who had been informed about the procedures gave signed consent. The patient's anamnesis revealed that he was diagnosed with DKC at the age of 7 months, he suffered from meningitis twice at the age of 1 and 2, and he frequently had to be taken to hospital due to pulmonary infections. The physical examination revealed diffuse, hyperpigmented skin lesions and generalized dry skin. The breath sounds were coarse, and the patient had a cough and abundant secretion during the pulmonary examination. However, there was no active infection. The results of systemic examination were normal. The patient had been receiving intravenous immunoglobulin every 21 days and subcutaneous human granulocyte colony-stimulating factor (Neupogen Roche 30MU) every two days for the last 16 months due to DKC-associated immunodeficiency. Neupogen is a glycoprotein regulating the production and release of functional neutrophils from the bone marrow. Neupogen, containing r-metHuG-CSF (Filgrastim), causes marked increases in peripheral blood neutrophil counts within 24 hours, with minor increases in monocytes. It activates bone marrow and affects neutropenia. The patient's laboratory assessment revealed pancytopenia. He was referred to the Pediatric Hematology Department. His laboratory values reached normal levels (white blood cells: 3,700; platelets: 100,000) after two weeks of routine treatment, and the dental operation was scheduled.

The Mallampati score of the patient whose neck and jaw movement was normal was 1. An infusion of 0.9% NaCl was started after the patient was placed on the operation table, and vascular access was established. As the patient was monitored, his mean arterial blood pressure, heart rate, end-tidal carbon dioxide, and peripheral oxygen saturation (SpO2) were measured. Anesthesia was induced by using 1 μg/kg of fentanyl and 1–3 mg/kg of propofol intravenously. After confirmation of face-mask ventilation, rocuronium (0.6 mg/kg) was given intravenously. The mask ventilation and intubation were successful. There was no bleeding in the mouth. Anesthesia was maintained with use of 50% nitrous oxide in oxygen and sevoflurane 2% by inhalation.

DISCUSSION

DKC was first defined by Zinsser in 1906 (4). Eighty to ninety percent of the patients die at a young age due to the associated complications. Pancytopenia-related bleeding and opportunistic infections are common in patients with DKC (5). Accordingly, the patients' clinical and laboratory data should be assessed carefully prior to any procedure.

Seventy-five percent of the patients followed up due to DKC progress to pancytopenia. Of these, 80% develop it before the age of 20 and 50% before the age of 10 (6). Additionally, patients may develop life-threatening malignancies such as myelodysplastic syndrome, acute myeloid leukemia, and solid tumoral formations (6). In DKC patients to be operated, pancytopenia, if present, should be treated prior to general

anesthesia. The white blood cell counts should be normal and platelets at a healthy level for the scheduled surgical procedure. If necessary, patients should be treated with Neupogen. In our case, the dental treatment including tooth extraction was performed after pancytopenia treatment. No intraoperative or postoperative complication occurred.

The most common pulmonary conditions in patients with DKC include pulmonary fibrosis, characterized by impaired gas exchange and abnormalities of the pulmonary vasculature (5). The estimated mortality rate due to such pulmonary complications is 10–15% (6). However, previous studies have shown that such complications develop because of using drugs with pulmonary toxicity, especially in patients scheduled for bone marrow transplantation (7). Another pulmonary problem is the frequent infections developed due to immunodeficiency (7,8). The DKC patients to undergo general anesthesia should be assessed for pulmonary respiratory functions and presence of infection. Surgical operations should be attempted only when the lungs function optimally. The healthcare team should be prepared for intraoperative and postoperative complications. Additionally, it should not be forgotten that postoperative ventilator support may be required in presence of pulmonary fibrosis. Aiming the minimal suppression of the pulmonary functions, anesthetics with a bronchodilator effect on the lungs should be preferred. Mirere et al. who performed emergency abdominal surgery in a patient with DKC drew attention to the fact that this rare disease might progress to involve multiple organs including the skin, gastrointestinal tract, bone marrow, and respiratory system and to the necessity of sound preoperative examination and intraoperative monitoring. There were only findings of chronic pulmonary infection in our case. Neither any problems except for increased secretion nor postoperative pulmonary complications were observed.

Supportive equipment should be prepared in advance when the patient is expected to suffer from difficult airway based on the oral examination findings. In addition, patients with gingival hyperplasia should be scheduled for atraumatic intubation in order to avoid bleeding and bone damage due to alveolar bone loss (9). In our case, no abnormal findings were recorded during the preoperative intraoral examination, and the intubation was performed successfully.

CONCLUSION

There are two important points to take into consideration in DKC patients to undergo general anesthesia. First, the patient should be screened for presence of BMF-related pancytopenia and opportunistic infections. Then the patient should undergo a thorough pulmonary examination for difficult airway and postoperative respiratory distress. The operation should be scheduled for a time when the patient's clinical and laboratory values are optimal.

REFERENCES

1. Nishio N, Kojima S. Recent progress in dyskeratosis congenital. Int J Hematol 2010;92:419–24.
2. Mitre CI, Corda DM, Dunca F, Iancu C. Anesthesia in a patient with dyskeratosis congenita presenting for urgent subtotal gastrectomy. J Clin Anesth. 2015;27(7):612–5.
3. Davidovich E, Eimerl D, Aker M, Shapira J, Peretz B. Dyskeratosis congenita: dental management of a medically complex child. Pediatr Dent. 2005;27(3):244–8.
4. Savage SA, Alter BP. Dyskeratosis congenita. Hematol Oncol Clin North Am. 2009;23:215–31.
5. Alter BP, Giri N, Savage SA, Rosenberg PS. Cancer in dyskeratosis congenita. Blood. 2009;113:6549–57.
6. Dokal I. Dyskeratosis congenita in all its forms. Br J Haematol. 2000;110:768–79.
7. Fernández García MS, Teruya-Feldstein J. The diagnosis and treatment of dyskeratosis congenita: a review. J Blood Med. 2014;5:157–67.
8. Karunakaran A, Ravindran R, Arshad M, Ram MK, Laxmi MK. Dyskeratosis congenita: a report of two cases. Case Rep Dent. 2013;2013:845125.
9. Baran I, Nalcaci R, Kocak M. Dyskeratosis congenita: clinical report and review of the literature. Int J Dent Hyg. 2010;8(1):68–74.