Anesthetic Management of a 12-Year-Old Patient with Proteus Syndrome

Proteus Sendrom'lu 12 Yaşında Bir Hastada Anestezi Yönetimi

Proteus Sendromu'nda Anestezi Yönetimi / Anesthetic Management in Proteus Syndrome

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Özet

Proteus sendromu vücudun herhangi bir dokusunda asimetrik, orantısız aşırı doku büyümesi ile birlikte olan son derece nadir bir konjenital hastalıktır. Proteus sendromu olan hastaların çoğu genellikle aşırı doku büyümeleri ve ilgili deformiteler için ameliyata gereksinim duyar. Bununla birlikte, kistik akciğer gibi pulmoner bulgular ve kifoskolyozu içeren vertebral anormallikler yüksek anestezi riskine yol açabilir. Proteus sendromu olan hastalar preoperatif ayrıntılı bir şekilde değerlendirilmeli, anestezi sırasında ve sonrasında dikkatlice yönetilmelidir. Bugüne kadar Proteus sendromlu 200 olgu bildirilmiş olmasına rağmen, bu hastalarda anestezi yönetimi ile ilgili az sayıda yayın vardır. Bu yazıda, herhangi bir anestezi ilişkili komplikasyon olmaksızın, ayak rekonstrüksiyonu için başarılı bir şekilde ameliyat edilen Proteus sendromlu 12 yaşındaki bir erkek çocuk olgusu sunulmaktadır.

Anahtar Kelimeler

Havayolu Yönetimi; Anestezi; Proteus Sendromu

Abstract

Proteus syndrome is an exceptionally rare congenital disorder with asymmetrical disproportionate overgrowth that includes any tissue of the body. Most of the patients with Proteus syndrome are generally needed to operate for overgrowths and related deformities. However, pulmonary manifestations such as cystic lung and vertebral abnormalities including kyphoscoliosis can lead to high anesthesia risk. The patients with Proteus syndrome should be evaluated in detail preoperatively, and managed carefully during and after anesthesia. Although up to 200 cases with Proteus syndrome have been reported to date, there are few reports on anesthetic management in such cases. In this paper, we present a Proteus syndrome in a case of 12-year old boy who was succesfully operated for foot reconstruction without any anesthesia-related complication.

Keywords

Airway Management; Anesthesia; Proteus Syndrome

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Introduction

Proteus syndrome (PS), first described by Cohen [1], is an extremely rare hamartomatous disorder with an estimated prevalence of approximately 1:1.000.000 [2]. It affects more males than females [3], and is characterized with overgrowth of multiple tissues. Clinical features typically start in the first 18 months of life [4]. Its diagnosis is based on clinical features and radiological findings as per the criteria recommended by Biesecker [3]. Surgery is usually indicated due to overgrowth lesions which lead to complications such as organ deformities, function loss or cosmetic problems. However, anesthetic management of the patients with PS is very difficult due to the presence of kyphoscoliosis, otolaryngologic conditions and cystic lung malformation that are predisposed to severe respiratory complications during and after anesthesia.

Case Report

A 12-year-old boy with PS, weighing 40 kg, presented for left foot reconstruction. On physical examination, there was a disproportionate asymmetric overgrowth and toes macrodactyly on his left foot (Figure 1). He also had hemihypertrophy through

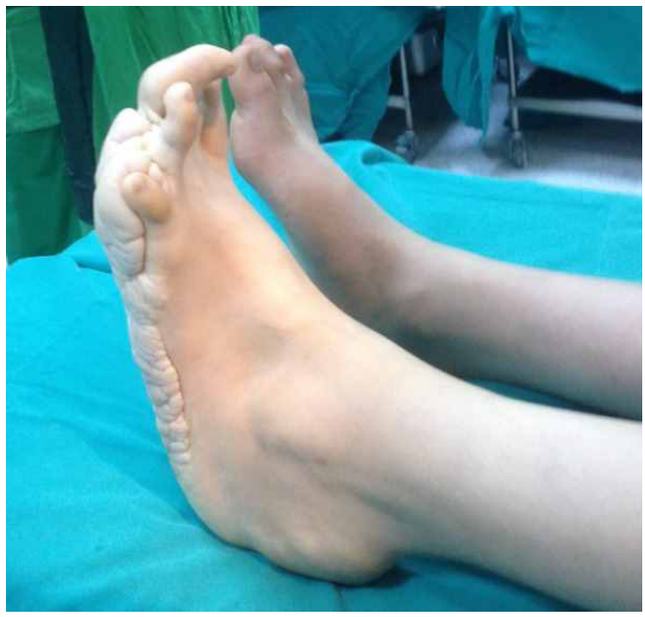

Figure 1. Macrodactyly and asymmetric overgrowth on the left foot

the left limb. Magnetic resonance imaging (MRI) of the left foot revealed hyperostosis of the calcaneus, talus, cuneiform bone and 3.-4. metatarsal bones with subluxation in talotibial joint. This overgrowth was started at 3 years old, and increased in time. However, the patient was diagnosed with PS at 9 years old. There was no abnormality in vertebral MRI, however calvarial asymmetry was detected in brain MRI with a normal electroencephalogram. He had café-au-lait spots on various parts of his body, including neck, arms and chest (Figure 2). Ultrasonography showed bilateral grade 2 ureterohydronephrosis and multipl millimetric renal cysts. Cardiac and pulmoner examination were not revealed any abnormalities, with normal electrocardiography and chest X-ray.

All routine laboratory tests were in normal ranges. His blood pressure, heart rate, EtCO2, and oxygen saturation were 115/65 mmHg, 105/min, 41 mmHg, and 99% respectively. Airway assessment was consistent with Mallampatti class 3. Informed

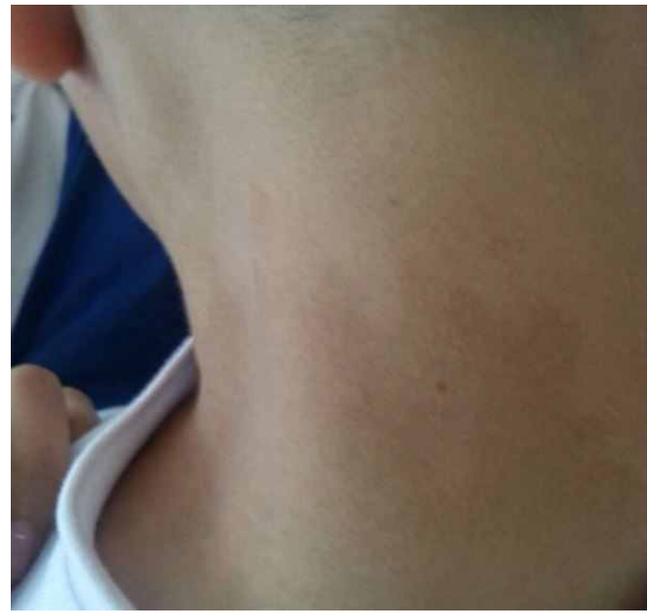

Figure 2. Multiple café-au-lait spots can be seen on the neck.

consent form was taken from his parents.

Monitoring included electrocardiography, noninvasive blood pressure and pulse oximetry. Anesthesia was induced by fentanyl (2,5 µg/kg), lidocaine (1 mg/kg) and propofol (3 mg/kg) with 100% oxygen. After rocuronium (0,8 mg/kg) endotracheal intubation was performed with using a 6,5 no spiral endotracheal tube uneventfully. Anesthesia was maintained with sevoflurane 2% MAC with nitrous oxide (50%) in oxygen (50%) at 3 l/min total gas flow. In addition, rocuronium was used two times in maintenance of anesthesia. The operation was performed in right-side position and lasted 3 hours without any complication, atropine 0,5 mg and neostigmine 1,5 mg was used for recurarization, and finally the patient was extubated successfully. The patient was given tramadol (1 mg/kg) for postoperative pain control. The postoperative course was also uneventfully, and he was discharged on tenth day.

Discussion

PS is a complex congenital hamartomatous disorder with overgrowth of multiple tissues in a mosaic manner. For correct diagnosis, the following three criteria are necessary; mosaic distribution, progressive nature, and sporadic occurence [5]. Skeletal system, connective tissue and fat are the most commonly affected areas [5]. However, this syndrome can involve any part of the body. To date, many dermatological manifestations such as cerebriform connective tissue nevi, Café-au-lait spots, cutaneous, venous and lymphatic malformations and patchy dermal hyperplasia were reported [6]. Our patient also had Café-au-lait spots on the arms, neck and chest. In addition, hemihypertrophy was also noted in his left limb which was led to walking disturbance.

PS is associated with a number of tumors. Ozkınay et al. reported a case of PS with various tumoral lesions including cavernous hemangiomas, pigmented nevi, lipomas and lymphangiomas [7]. Of these, lipomas are the most common tumor type that usually located in the abdominal or chest wall. Our patient had several subcutaneous fat tissue hipertrophies. Especially

two types of tumor, monomorphic adenomas of the parotid glands and bilateral ovarian cystadenomas, are used in the diagnostic criteria because of their specificity [5].

Central nervous system manifestations such as brain malformations, mental retardation and seizures can be presented in up to 40% of patients [8]. In our case, calvarial asymmetry due to hiperostosis was detected in brain MR. In addition, he had mild mental retardation which was led to poor school performance.

The typical facial phenotype including coarse facies, prominent forehead, widely spaced eyes, minor ptosis, low nasal bridge, relative lengthening of the face, and persistently open mouth may be present in some patients with PS. In a case series reported by Angurana [6], 3 of 6 PS cases had facial dysmorphism. Our patient had coarse facial lines and open mouth.

Patients with PS may also have overgrowths through the airway, and this condition can lead to a difficult intubation. Pennant et al. [9] reported such a case who has to be performed fiberoptic-aided intubation under inhalational anesthesia. Airway assessment of our case was consistent with Mallampatti class 3, but no problem occurred during the intubation. Additionally, various vertebral deformities such as kyphoscoliosis and dysplastic vertebra also be found in these patients, and may affect the pulmonary ventilation. In our patient, no vertebral abnormality was detected radiologically. A portion of patients with PS may have some pulmonary manifestations that may cause difficulties during anesthesia. Of these, cystic changes, emphysema, atelectasis, and fibrosis are the most frequently reported pulmonary complications. In addition, asymmetric growth in the ribs may cause restrictive lung disease. In our case, no abnormality was detected in the lung parenchyma and ribs. Patients with PS are also at increased risk of pulmonary embolism because of vascular malformations. Deep venous thrombosis and pulmonary embolism are also one of the most common causes of death, especially in young children [8]. For this reason, patients who will be operated for any reason should be undertaken perioperative anticoagulation prophylaxis [5]. Additionally, spinal anesthesia can be a reliable choice of anesthesia in suitable cases according to its low risk of developing deep vein thrombosis [10].

Urological abnormalities such as nephromegaly and renal cysts are found to be at a ratio of 9% [3]. Our patient had also bilateral grade 2 ureterohydronephrosis, but renal function tests were normal.

In conclusion, PS is a rare clinical entity which can lead to develop difficulties in anesthetic management due to presence of overgrowth through the airway, pulmonary manifestations and vertebral abnormalities. Therefore, detailed preoperative anesthetic evaluation should be necessary for these patients.

Competing interests

The authors declare that they have no competing interests.

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