

Chronic Rhinosinusitis and Endonasal Deformity in X Linked Agammaglobulinemia: A Case Report

X Geçişli Agammaglobulinemi Vakasında Kronik Rinosinüzit ve Endonazal Deformite: Olgu Sunumu

*Süay ÖZMEN, MD, **Ömer Afşin ÖZMEN, MD, ***Taner YILMAZ, MD

* Bursa Dörtçelik Children Hospital,

** Uludağ University Faculty of Medicine, Department of Otorhinolaryngology, Bursa

*** Hacettepe University Faculty of Medicine, Department of Otorhinolaryngology, Ankara

ABSTRACT

X linked agammaglobulinemia (Bruton's disease, XLA) is a rare infantile immunodeficiency syndrome due to a B-cell defect. It is characterized by the absence of B lymphocytes, the total absence or severe deficiency of all immunoglobulin classes, and recurrent bacterial infections with pyogenic pathogens. Immunoglobulin replacement therapy and antibiotics do not suffice in some cases, making sinus surgery to advance the drainage necessary. We present 16-year-old patient with hypogammaglobulinemia, treated with functional endoscopic sinus surgery (ESS) and septorhinoplasty. After one year of follow-up, there was no evidence of nasal obstruction and nasal drainage at the right side, pus drainage from the left side continued. To our knowledge in the literature there was no report of septoplasty or rhinoplasty performed in a patient with XLA. The aim of the present case report was to emphasize that septorhinoplasty operation might be performed without any problems in an immunodeficient patient.

Keywords

Agammaglobulinemia; sinusitis; rhinoplasty

ÖZET

X geçişli agammaglobulinemi (Bruton hastalığı, XLA) infant dönemde açığa çıkan B hücre defektine bağlı gelişen nadir bir immün yetmezlik sendromudur. B lenfosit eksikliğine bağlı tüm immunoglobulin tiplerinde ağır yetersizlik mevcuttur ve piyojenik bakteriler ile reküren bakteriyel enfeksiyonlar görülür. Hastada akut ve reküren bakteriyel enfeksiyonlarla gelişen kronik rinosinüzit ve akciğer enfeksiyonları görülür. Bazı vakalarda immunoglobulin takviyesi ve antibiyotik tedavisi yeterli gelmez ve sinüs cerrahisi ile drenajın sağlanması gerekir. 16 yaşında hipogammaglobulinemisi bulunan bir hasta sunulmuştur. Fonksiyonel endoskopik sinüs cerrahisi ve septorinoplasti ile tedavi edilmiştir. Bir yıllık takip sonrasında nazal obstrüksiyon ve sağ nazal pasajda enfeksiyon bulgusu yoktur. Sol nazal pasajdan püy gelişi devam etmektedir. Bildiğimiz kadarı ile literatürde septoplasti veya rinoplasti yapılan XLA hastasına rastlanmamıştır. Bu vaka sunumunun amacı immün yetmezliği olan bir hastada septorinoplasti ameliyatının sorunsuz bir şekilde yapılabileceğini göstererek, bu vakalardaki yaşanabilecek tereddütü azatmaktır.

Anahtar Sözcükler

Agammaglobulinemi; sinüzit; rinoplasti

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Correspondence

Süay ÖZMEN, MD

Bursa Dörtçelik Children Hospital, Bursa

E-mail: suayozmen@yahoo.com

INTRODUCTION

XLA is a severe life threatening disease characterized by the absence of B lymphocytes, the total absence or severe deficiency of all immunoglobulin classes, and recurrent bacterial infections with pyogenic pathogens. Typically, the disease is first seen during early infancy or childhood and, unless treated aggressively with intravenous gamaglobulin, antibiotics or both, results in significant morbidity. The patients suffer from acute and recurrent bacterial infections with chronic rhinosinusitis and chronic lung disorders. Immunoglobulin replacement therapy and antibiotics do not suffice for the treatment of the infection in some cases, making sinus surgery necessary to improve drainage. The aim of the present case report was to emphasize that septorhinoplasty operation might be performed without any problems in an immunodeficient patient, in order to abate the hesitancy in these cases.

CASE REPORT

A 16-year-old boy presented with complaints of nasal obstruction, continuous nasal discharge and cough. He had a history of nasal trauma at the age of five. He had been followed up in the Pediatric Immunology Department with the diagnosis of XLA, receiving intravenous immunoglobulin replacement therapy every four weeks. He also had been prescribed antibiotics several times with the diagnosis of sinusitis which were failed to get complete recovery. Physical examination revealed a bony hump, obstruction of left nasal cavity due to deviated septal cartilage, purulent drainage in his right nasal passage and post-nasal purulent drainage. Paranasal CT scan showed septal deviation, opacification in all paranasal sinuses and obliteration of osteomeatal units (Figure 1 and 2). Preoperative photographs were taken (Figure 3a and 3b). Preoperatively he was consulted with the Pediatric Immunology Department and was given intravenous immunoglobulin treatment. The patient was operated under general anesthesia. Initially, bilateral anterior and posterior ethmoidectomy was done, together with opening of maxillary sinus ostia. Then an external septorhinoplasty was performed. Septal cartilage was extremely deviated causing complete obstruction of the left nasal cavity. Alar and upper lateral cartilages and septal cartilage were exposed. Mu-



Figure 1. Paranasal CT scan demonstrating septal deviation.



Figure 2. Paranasal CT scan demonstrating opacification in all paranasal sinuses.



Figure 3a. Preoperative anterior view.

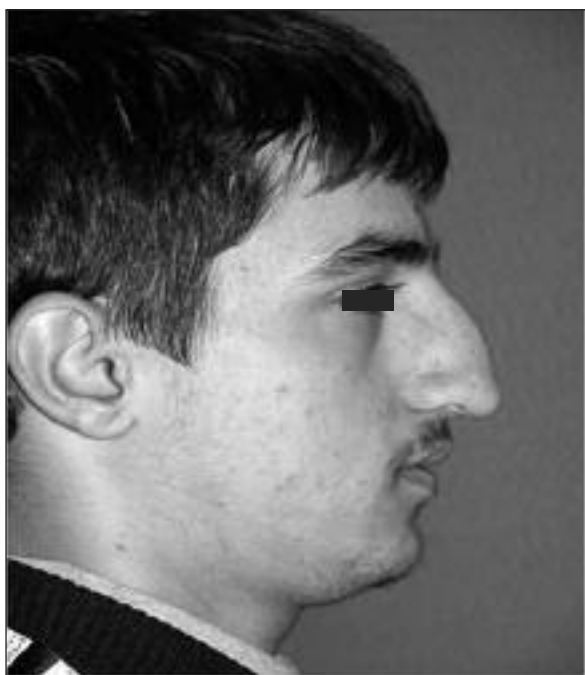


Figure 3b. Preoperative lateral view

coperichondrium and mucoperiosteum were elevated bilaterally. Posterior chondrotomy was made. Septal cartilage was separated from maxillary crest and was taken outside. Bony hump was reduced. The septal cartilage was straightened and replaced to its normal anatomic position, and fixed to the upper lateral cartilages and nasal spine with three sutures. No complications were seen in the intraoperative and postoperative periods. After one year of follow-up, there was no evidence of nasal obstruction and nasal drainage at the right side, pus drainage from the left side continued. His postoperative photographs were taken (Figure 4a and 4b). Informed consent was taken from patient's parents to use his photographs.

DISCUSSION

Early diagnosis, appropriate immunoglobulin replacement therapy, intensive chest therapy, and antibiotics have changed the clinical history of patients with humoral immunodeficiency,¹ however many still present with chronic productive cough, the hallmark of chronic bronchitis or sinusitis.²

Many immunodeficient patients have a history of repeated sinus surgery before the recognition of their immune defect. Conventional management of sinusitis is of little benefit in patients with chronic re-

fractory sinusitis with an underlying immunodeficiency.³

In order to elucidate the optimal treatment of chronic sinusitis in a patient with immunodeficiency, antibiotic regimens, probably combined with surgical drainage are to be determined.

Patients with naso-septal deformity managed only by septoplasty had accentuation of nasal pyramid deformity; those treated by septorhinoplasty showed a



Figure 4a. Postoperative anterior view.



Figure 4b. Postoperative lateral view.

good aesthetic and functional result after long-term follow-up.⁴

In the literature, few studies were made to reveal the efficacy of the surgical treatment of chronic sinusitis in the patients with immunodeficiency. In immunodeficient patients treated with antibiotics and immunoglobulin replacement therapy, functional endoscopic sinus surgery is successful in only half of the patients.⁵ To our knowledge, this is the first report of septorhinoplasty performed on an XLA patient in the literature.

In this case, the patient's nasal obstruction and drainage were reduced significantly. However, nasal drainage from the left side was not recovered. It was dedicated to insufficient sinus surgery due to narrowed

nasal passage by septal deviation. Revision ESS is planned to the left side.

We successfully performed ESS, septoplasty and rhinoplasty in an immunodeficient patient. No complication was seen in the intraoperative and postoperative periods. After one year of follow-up, our patient does not have nasal obstruction and he is satisfied with his appearance. He only complains of postnasal drainage at night.

The aim of the present case report was to emphasize that septorhinoplasty operation might be performed without any problems in an immunodeficient patient, in order to abate the hesitancy in these cases. ESS, septoplasty and rhinoplasty can be performed safely on immunodeficient patients if necessary precautions are taken.

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