Cukurova Medical Journal
ÇUKUROVA ÜNİVERSİTESİ TIP FAKÜLTESİ DERGİSİ

Cukurova Med J 2016;41(4):804-807
DOI: 10.17826/cutf.254551

OLGU SUNUMU/CASE REPORT

Amyloidosis and difficult airway: a case report

Amiloidozis ve zor hava yolu: olgu sunumu

Ebru Biricik[1], Ersel Güleç[1], Feride Karacaer[1], Özgür Sürmelioğlu[2], Dilek Özcengiz[1]

[1]Cukurova University Faculty of Medicine, Department of Anesthesiology and Critical Care, [2]Department of Head and Neck Surgey, Adana, Turkey

Cukurova Medical Journal 2016;41(4):804-807.

Abstract
Amyloidosis is a disease which can affect any tissue in the body with extracellular deposition of low molecular weight protein fibrils. Amyloid deposition can appear in many organs and tissues. Amyloidosis of tongue occurs most commonly in systemic form as rubbery macroglossia due to extra-deposition of amyloid within the suprahyoid muscle. Anesthesiologist can experience the negative consequences of amyloidosis in many conditions. Difficult airway due to amyloid deposition in tongue is an unexpected condition. We aimed to report a case that had respiratory distress with extremely big tongue due to amyloidosis. We performed a fiberoptic intubation for emergent tracheostomy without any complication.

Key words: Amyloidosis, difficult airway, anesthesia

Öz
Amiloidozis, düşük molekül ağırlıklı amiloid moleküllerinin ekstrasellüler birikimi ile karakterize bir hastalıktır. Amiloid birikimi kalp, karaciğer, böbrek, cilt, barsak, otonom sinir sistemi, dil ve karpal tünelde görülebilir. Dilde amiloidozis, amiloidin suprahyoid kaslarda ekstrasellüler depozisyonuna bağlı makroglossi çoğunlukla amiloidin sistemik formuna bağlı görülmektedir.Anestezistler amiloidozisle pek çok farklı durumda karşılaşabilirler. Dilde amiloid birikimine bağlı zor hava yolu beklenmedik bir durumdur. Fiberoptik bronkoskopi ile acil trakeostomi açılmasını sağladık ve herhangi bir komplikasyon gözlenmedi. Burada amiloidozise bağlı aşırı büyük dilden kaynaklanan solunum sıkıntısını sunulması amaçlanmıştır.

Anahtar kelimeler: Amiloidozis, zor hava yolu, anestezi

INTRODUCTION

Amyloidosis is a disease influencing the tissues in the body with the extracellular deposition of low molecular weight protein fibrils. Amyloid deposition can lead to deterioration of tissues and organ failure. This deposition can identified histochemically with kongo red. Amyloidosis' s etiology is still unclear. It was first described in 1854 by pathologist Rudolph Virchow[1]. Amyloidosis can exist to be systemic or localized form[2].

Head and neck involvements are common in both localized and systemic forms of amyloidosis. As well as it can affect many organs, head and neck involvement of amyloidosis is mostly localized in the larynx[3]. The second most common involvement is the tongue by its localized form.

In this case report, we aimed to present a case with an amyloidosis localized in tongue and caused to a sudden respiratory distress and required tracheostomy.

CASE

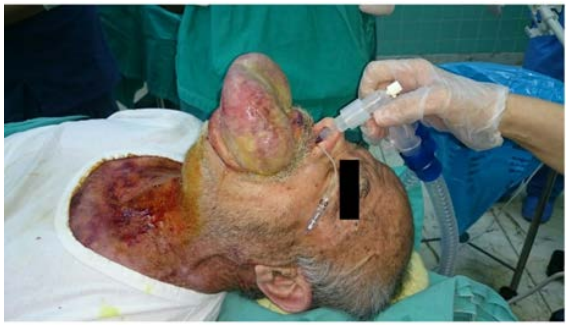
Seventy-six years old male patient with respiratory distress admitted to the emergency service. He had a tongue completely filling the oral cavity and a difficult breathing with orthopnea. He was trying to take breathe with leaning forward (Figure 1). There were many ulcers on the tongue. He had ecchymosis on the chest wall and hands. By auscultation, there were bilaterally crackle lung sounds and bilaterally decreased breath sounds in the bases of the lung. The patient's mental status was confused and self-care was quite poor.

Yazışma Adresi/Address for Correspondence: Dr. Ebru Biricik, Cukurova University Faculty of Medicine, Department of Anesthesiology and Critical Care, Adana, Turkey, Email: ebrubiricik01@gmail.com
Geliş tarihi/Received: 16.03.2016 Kabul tarihi/Accepted: 04.05.2016

Oxygen 100% of 4 L/min was administered by a nasal cannula to the patient who had a peripheral oxygen saturation value of 85%. By the consultation of surgeon, we decided to perform an urgent surgical tracheostomy. Pseudoepitheliomatous hyperplasia, chronic active inflammation, and amyloid (+) were identified in tongue biopsy before six months. The patient's relatives reported that he had swelling throughout the body, especially the tongue. He had also a history of bronchial asthma as well as amyloidosis. He has been taking steroid drug (prednisolone 500 mg per a day) but not regularly.

Laboratory blood sample tests resulted in hemoglobin 9.7 g/dL, white blood cells 5100, hematocrit 29.7% and normal electrolyte levels. The patient's serum protein electrophoresis test results are shown in Table 1. Left ventricle ejection fraction was found to be 70% by the echocardiography (ECHO) measurement. A severe left ventricular diastolic dysfunction and pleural effusion were identified by the ECHO. After receiving informed consent from relatives of the patient, he was taken into the operating room. We did not consider a local anesthesia under sedation because he had a respiratory distress not to allow the supine position. Furthermore, the surgeon did not prefer to perform tracheostomy under local anesthesia due to the position of orthopnea. Thus, we decided to perform endotracheal intubation using fiberoptic bronchoscopy. His fasting period of time was enough for anesthesia. The patient was monitored using pulse oximetry, noninvasive blood pressure, and electrocardiography.

Figure 1: Preoperative patient's appearance. He had huge tongue and he tried to take breath with leaning forward.



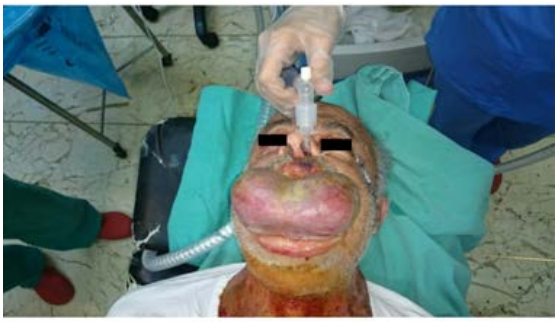


Figure 2 and 3: After fiberoptic nasotracheal intubation patient's appearance.

Table 1. Serum protein electrophoresis

	%	Normal Ranges
Serum albumin	45.52	55.8-65
α 1	6.52	2.2-4.6
α 2	20.06	8.2-12.5
β	9.19	7.2-14.2
γ	18.71	11.5-18.6

For nasotracheal intubation, a nasal decongestant (oximethasoline HCL) and a dose of 10 μg/kg atropine IV, as an antisialagogue agent, were administered. Then, at a dose of 0.02 mg/kg midazolam and 2 mg/kg ketamine IV were administered. We predicted to a ventilation by face mask would not be possible, and therefore, did not want to lose airway control after anesthesia performed. So we aimed to maintain patient's spontaneous breathing due to difficult airway. A nasal oxygen supplementation was continued as 4 L/min. While patient was on half-sitting position, endotracheal intubation was performed with fiberoptic laryngoscopy after used lubricant through left nostril. During fiberoptic intervention, we did not found any anatomical abnormality or lesion in nasotracheal airway. When examined the right and left bronchus, intense secretions were observed in both bronchi and then we aspirated the secretions. We did not experience any complication during fiberoptic intervention. Then patient intubated safely (Figures 2, 3). After endotracheal intubation, surgical tracheostomy was performed to the patient under general anesthesia. General anesthesia was performed with 0. 5 mg/kg rocuronium, %2 sevoflurane, %50-50 N2O-O2. When patient can safely ventilate via tracheostomy, surgery and general anesthesia were ended.

The patient was transferred to the postoperative care unit and then he was followed in intensive care unit for two days in terms of any adverse events. We did not observe any complication about fiberoptic tracheal intubation in postoperative period. Appropriate amyloid therapy was adjusted by endocrinology clinic.

DISCUSSION

Amyloid deposition can seem in many organs and tissue and can lead to a disruption of these organs[4] AA and AL are the most common form of amyloidosis. Twenty-eight different forms of amyloid protein were isolated (particularly cerebral, neurodegenerative transthyretin, dialysis, diabetes, Alzheimer's, Creutzfeldt-Jakob disease). AA amyloidosis is characterized by the overproduction of acute phase reactants similar to serum amyloid A (SAA) during chronic infection or inflammatory disease. This type of amyloidosis most commonly affects the kidneys; however the heart and nervous system are rarely affected.[9,10] Serum β2 microglobulin is elevated in patients with systemic AL amyloidosis.

Head and neck amyloidosis is often seen in case of localized amyloid of the AL type[5]. Localized amyloidosis in larynx has been well reported in the literature. But the localization of tongue is always with a systemic amyloidosis.[6] Amyloid deposition occurs at aerodigestive tract in 90% of patients with a systemic amyloidosis.[5] Amyloidosis of the oral cavity is uncommon. Anesthesiologist may experience in many different ways with amyloidosis[4-8] Hoarseness, dysphonia, and stridor are the most common symptoms of primary laryngeal amyloidosis. Dyspnea can also occur as an important symptom. Although it can be respiratory distress with laryngeal amyloidosis, there is no case of emergency difficult airway in the literature[11-15]. Amyloid deposition in the tongue is the most common site of head and neck in systemic amyloidosis[16,17].

O'Reilly et al. reviewed the cases of localized tongue amyloidosis[18]. Localized amyloid depositions can be nodular or flat in tongue. Infiltrative lesions tend to recurrence. But if severe obstructive symptoms are present, surgical management may be necessary. Amyloidosis associated with macroglossia occurs most commonly in systemic form as rubbery macroglossia[8,20]. In clinical evaluation, tongue can be significantly large, rigid or nodular.

We report a case having respiratory distress with extremely big tongue due to amyloidosis. It is the first report in literature on difficult airway with amyloidosis in tongue. We performed a fiberoptic bronchoscopy to the patient under sedation on half sitting position. In this case, the surgeon did not perform a glossectomy. However, glossectomy is the treatment options of macroglossia[21]. Difficult airway due to macroglossia was reported in the literature but no cases were with amyloidosis[22-24]. Besides, a difficult condition experienced by anesthesiologists due to amyloidosis has been published. Minoque et al. reported that they experienced an airway obstruction after the insertion of endotracheal tube to the patient with laryngotracheal amyloidosis undergoing microlaryngoscopy and laser surgery. Subsequently, the airway had become obstructed and also ventilation impossible. They had to perform supraglottic jet ventilation and identified an obstruction in rigid bronchoscopy. The obstruction was due to the flaps of mucosal tissue occluding the trachea both anteriorly and posteriorly[25]. Amyloidosis can present with so many clinical

situations for anesthesiologist. In this case report, we aimed to emphasize that anesthesiologist can experience a difficult airway with amyloidosis.

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