AMYLOIDOSIS AND DIFFICULT AIRWAY: A CASE REPORT

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Abstract
Amyloidosis is a disease which can affect any tissue in the body with extracellular deposition of low molecular weight protein fibrils. Amyloidosis 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 suprathyroid 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

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’ etiology is still unclear. It was first described in 1854 by pathologist Rudolph Virchow1. Amyloidosis can exist to be systemic or localized form2.

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 larynx3. 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 bilateral 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.
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.](image1)

![Figure 2 and 3: After fiberoptic nasotracheal intubation patient's appearance.](image2)

<table>
<thead>
<tr>
<th>Table 1. Serum protein electrophoresis</th>
<th>%</th>
<th>Normal Ranges</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum albumin</td>
<td>45.52</td>
<td>55.8-65</td>
</tr>
<tr>
<td>α 1</td>
<td>6.52</td>
<td>2.2-4.6</td>
</tr>
<tr>
<td>α 2</td>
<td>20.06</td>
<td>8.2-12.5</td>
</tr>
<tr>
<td>β</td>
<td>9.19</td>
<td>7.2-14.2</td>
</tr>
<tr>
<td>γ</td>
<td>18.71</td>
<td>11.5-18.6</td>
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microglobulin is elevated in patients' systemic are rarely affected. 

amyloidosis affects the kidneys; however the heart and nervous disease. This type of amyloidosis most commonly occurs during chronic infection or inflammatory conditions similar to serum amyloid A (SAA) during acute phase reactants. AA and AL are the most common forms of amyloidosis. Twenty AL amyloidosis.

Amyloid deposition can be seen in many organs and can lead to a disruption of these organs. AA and AL are the most common forms 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. 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. Localized amyloidosis in larynx has been well reported in the literature. But the localization of tongue is uncommon. Anesthesiologist may experience in many different ways with amyloidosis.

Amyloid deposition in the tongue is the most common site of head and neck in systemic 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.

Amyloid deposition in the tongue has been well reported in the literature. But the localization of tongue is always a systemic amyloidosis.

O'Reilly et al. reviewed the cases of localized tongue amyloidosis. 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. 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. Difficult airway due to macroglossia was reported in the literature but no cases were with amyloidosis. 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. 

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.

REFERENCES