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Is regular laryngoscopy necessary for multiple myeloma patients? Case report and review of literature

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ABSTRACT

Multiple myeloma (MM) is a neoplastic and lymphoproliferative disease. Laryngeal involvement or "metastatic" multiple myeloma is extremely rare. We present 68 years old woman with sudden respiratory distress who underwent emergency tracheostomy and discuss about necessity of regular laryngoscopy in multiple myeloma patients with this case and review of literature.

Key words: Laryngeal Multiple Myeloma, Laryngoscopy, Tracheostomy

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INTRODUCTION

Plasma cell neoplasms characterized by excessive plasma cell production. They can be benign or malignant.

There are several types of plasma cell neoplasms:
1. Extramedullary plasmacytoma
1.1 Primary (true) plasmacytoma of the mucosa with or without affected lymph nodes.
1.2. Extramedullary manifestation of multiple myeloma.
2. Solitary bone plasmacytoma
3. Multifocal form of multiple myeloma.
4. Multiple myeloma
5. Plasmablastic sarcoma. [1]

Multiple myeloma (MM) is a neoplastic and lymphoproliferative disease that may involve the bone marrow as well as extramedullary soft tissues characterized by the proliferation and accumulation of B lymphocytes and plasma cells that synthesize monoclonal immunoglobulin (M component). [2]

Its main symptoms are anemia, bone pain, and renal failure. [3, 4, 5, 6, 7, 8]

Laryngeal involvement or "metastatic" multiple myeloma is extremely rare (between 6% and 18%) with the epiglottis, glottis, false vocal folds, aryepiglottic folds, and subglottis involvement in decreasing the order of frequency. [9, 10]

Case Presentation

A 68-year-old woman known case of multiple myeloma from 4 years ago presented with sudden respiratory distress. Because of the diffuse edema in supraglottic area, intubation was impossible then she underwent emergency tracheostomy.
Computed tomogram scan of neck showed diffuse swelling in both false vocal cords (Figure 1).

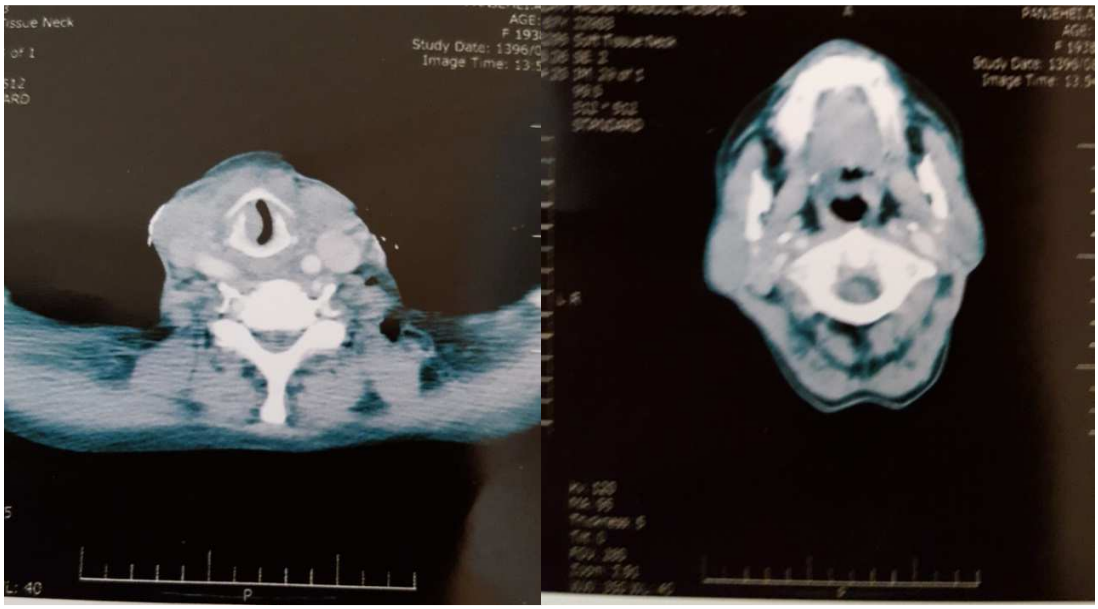

Figure 1. CT scan, diffuse FVC and aryepiglottic folds swelling

Direct laryngoscopy revealed a mucosa-lined growth involving the both arytenoids, aryepiglottic folds and false vocal cords (Figure2).

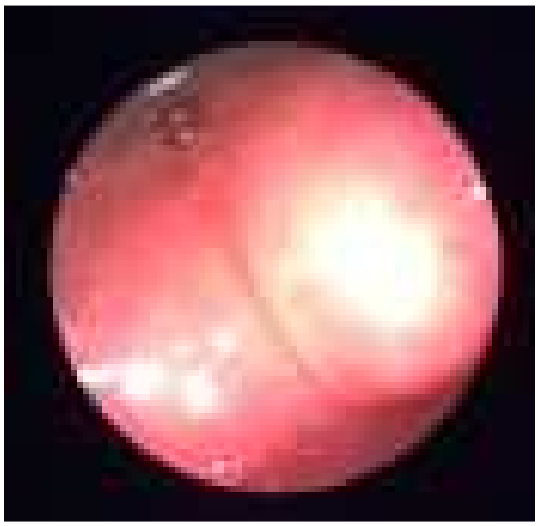

Figure2. Direct laryngoscopy, complete supraglottic obstruction

A biopsy from the mass showed plasmacytoid cells loosely arranged in a fibrocollagenous stroma. On immunohistochemistry, the tumor cells were strongly positive for CD138 and negative for cytokeratin, thus a preliminary diagnosis of plasmacytoma was made for the patient.

Her bone marrow aspiration in 2014 had shown a population of more than 80% immature plasma cells including plasmablast and binucleated plasmacells (Table1,2).

Table1. Patient's laboratory tests

LAB Tests	
Hb	**6.5 gm/dl**
RBC	**2.30 mill/mm²**
HCT	**20.7**
PLT	**68**
BUN	**25**
Cr	**1.5**
Uric Acid	**11.1**
LDH-p	**568**
Ca	**8.3**

Table2. Patient's urine analysis

Urine Analysis			
Fraction	Percentage	Mg/24h	Ref. Range[mg/24h]
Beta2-Microglobulin	0.0	0.0	0.0- 0.0
Hemoglobulin	0.0	0.0	0.0- 0.0
Free light chains	11.9	303.4	0.0- 0.0
Alpha 1-Microglobulin	1.9	48.5	0.0- 0.0
Free light chains Dimers	68.2	1739.1	0.0- 0.0
Albumin	5.5	140.3	0.0- 30.0
Transferrin	0.0	0.0	0.0- 0.0
Tammhorsfall	11.5	293.3	0.0- 30.0
IgG/IgA	1.0	25.5	0.0- 0.0
IgM/ Alpha2-Macro	0.0	0.0	0.0- 0.0
Total 24h	100	2550.0	Up to 150
Interpretation	**Bence-jonse Proteinuria**		

DISCUSSION

Plasma cell neoplasms are clonal proliferations of plasma cells.

Multiple myeloma (MM) is one of the most frequent plasma cell neoplasms with the incidence of 4 cases per 100,000.

The exact etiology is unknown. Several probable etiologies are described:
1) Point mutations of the ras genes (N-ras and K-ras)
2) Mutations of the tumor suppressor genes (p53 and Rb-1) [2]
A diagnosis of MM requires at least 10% clonal bone marrow plasmacytosis, M protein existence in serum or urine (except in nonsecretory myeloma), and evidence of end-organ damage (hypercalcemia, renal failure, anemia, and bone disorders). [9, 10]

Multiple myeloma has a variable prognosis (mean survival greater than 10 years).

The main prognostic factor for disease is a progression into a disseminated MM. It can occur several years after the initial diagnosis. [9, 10]

The disseminated form has the worst prognosis. Disseminated extraosseous disease in patients with multiple myeloma is common, although it is usually subclinical. Extraosseous involvement is 71% to 73% in autopsy series of patients with MM. Laryngeal involvement in MM is rare, and only a few cases have been reported in literature [11].
In this study we report a catastrophic multiple myeloma case with a sudden respiratory distress and review five other cases with laryngeal involvement (Table3).

Table3. Review of literature

Year Author	Laryngeal Involvement	Age	Sex	symptom	Treatment	Tracheostomy requirement	Follow up
Carolyn 1995 [11]	Glottis and Supraglottis	62	Male	6 months hoarseness +neck swelling	Chemotherapy + radiotherapy after recurrence	+	6 months Relief of airway stenosis
Muraleedharan 2006 [1]	Subglottis	65	Male	stridor	Chemotherapy	+	-
Grobman [9] 2012	Left Ventricle	58	Male	2 months dysphonia	Chemotherapy+ Stem cell transplant	unknown	4 months well
Allegra 2016 [2]	Right TVC, FVC	68	Male	4 months Dyspnea, Dysphonia, dysphagia	Chemotherapy	unknown	6 months Partial remission
Narayanan [1] 2017	Left TVC, aryepiglottic fold, pyriform sinus	44	Female	3 months hoarseness + 2 weeks stridor	Chemotherapy +stem cell transplant	+	3 years Complete remission
Sarafraz 2017	Left and Right FVC	68	Female	Respiratory distress	Chemotherapy	+	Expired

The peak age of incidence is the sixth decade of life with a male to female preponderance of 3:1. [1] We described a 68 year old woman with supraglottic involvement. In decreasing order the epiglottis, vestibular fold, arytenoids, aryepiglottic folds, and the subglottis are involved in extraosseous MM.[1] Although laryngeal involvement is rare in MM and its symptoms varies according to disease localization and includes voice change, respiratory distress, and stridor [1] but it can rarely present with a sudden onset and result in airway obstruction.

We reviewed 6 MM cases with laryngeal involvement from 1995 to 2017, four cases underwent tracheostomy and two cases were indeterminate. This point has an important role in patient prognosis and future management.
Different treatment options were described for plasma cell neoplasms such as surgery, radiotherapy and chemotherapy. Plasma cell neoplasms are radiosensitive but multiple myeloma is considered a systemic problem and is treated with chemotherapy and stem cell transplantation. [9, 10] All six patients were reviewed in this study received chemotherapeutic drugs. In 4 months to three years follow up three of them were in remission phase, one of them were indeterminate, one of them partially improved and our case expired one month after sudden respiratory distress and tracheostomy tube insertion.

Although tracheostomy is now commonly used, the complication rate remains high.

In study by Mehta a complication rate was 48%. Main complications are emphysema, infection, tube obstruction, hemorrhage, aspiration, pneumothorax and atelectasis. The indications are prolonged mechanical ventilation, pulmonary toilet, surgical access and airway obstruction. [12]
Tracheostomy indication in MM is laryngeal infiltration and airway obstruction. Because of their pancytopenia complication rate specially hemorrhage, infection, tracheitis and mediastinitis can be higher than general papulation.

Hemorrhage and hematoma formation may occur during the operation or postoperatively.
Local infection at stoma site and tracheitis occurred to some degree in every patient having tracheostomy. In MM patients with leukopenia it can progress to mediastinitis and lung abscess easily.

Complication rate with emergency tracheostomy was twice higher than elective operation. [12]
Then it is necessary to use CT scan and/or MRI to evaluate bone-cartilaginous and soft-tissue lesions and can further demonstrate the presence of subclinical lesions. [9, 10]

Although laryngeal involvement is rare but tracheostomy requirement and its complications can complicate the management and worsen the prognosis.

Then we suggest at least regular flexible laryngoscopy in patients with multiple myeloma to demonstrate early laryngeal lesions and manage them in early phase. It can be cost effective and lifesaving.

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