

# Tracheal Lymphoma Presenting with Acute and Progressive Dyspnea

## Akut ve İlerleyici Solunum Sıkıntısı ile Belirti Veren Trakeal Lenfoma

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### ABSTRACT

The most common causes of acute airway obstructions are infections, foreign bodies, laryngeal tumors, and allergic reactions. Primary tracheal tumors are among very rare causes of acute airway obstructions. Tracheal involvement of lymphoma is also not common. In this case report, a 58 year-old woman who presented with acute and progressive airway obstruction was reported. A tracheal mass was detected on computed tomography scan, an emergency tracheostomy was performed, an incisional biopsy was taken, and the histopathological examination revealed a mantle cell variant of primary non-Hodgkin lymphoma of the trachea. No other involved site or lymph node could be detected. This is the second primary tracheal mantle cell lymphoma case in the literature. (*Gazi Med J 2011; 22: 127-9*)

**Key Words:** Mantle-cell lymphoma, Non-Hodgkin's lymphoma, tracheal neoplasms, dyspnea

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### ÖZET

Akut havayolu obstrüksiyonlarının en sık nedenleri arasında enfeksiyonlar, yabancı cisimler, larengeal tümörler ve allerjik reaksiyonlar yer alır. Primer trakeal tümörler oldukça nadir karşılaşılan akut havayolu obstrüksiyon nedenlerindedir. Trakeanın lenfoma tarafından tutulumu da nadir görülür. Bu olgu sunumunda akut ve ilerleyici havayolu obstrüksiyonu nedeniyle başvuran 58 yaşında bir kadın hasta bildirilmektedir. Hastada bilgisayarlı tomografi tetkiki sonucunda trakeayı tutan bir kitle tespit edilmiş, acil trakeotomi açılmış, insizyonel biyopsi alınmış, patolojik inceleme sonucunda mantle hücreli lenfoma tanısı konulmuştur. Hastada başka bir bölgede ya da lenf nodunda lenfoma tutulumu saptanmamıştır. Hasta, literatürdeki ikinci primer trakeal mantle hücreli lenfoma olgusudur. (*Gazi Med J 2011; 22: 127-9*)

**Anahtar Sözcükler:** Mantle-hücreli lenfoma, non-hodgkin lenfoma, trakeal neoplaziler, dispne

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### INTRODUCTION

Several etiological factors can cause acute upper airway obstruction. The most common causes are infection, neoplasms, trauma, foreign bodies and angioedema (1). Primary malignant tumors of the trachea are uncommon. The most frequent tumors of the trachea

are squamous cell carcinoma and adenoid cystic carcinoma (2). Primary presentation of extranodal lymphoma affecting the trachea is quite unusual. According to a literature review combining data from seven series including 425 cases of tracheal tumors reported between 1930 and 1989, the frequency of lymphoma is 0.23% (only one case) among all tracheal tumors (3).

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Mantle cell lymphoma is a distinct subtype of B-cell lymphoma and accounts for approximately 3-10% of all lymphomas (4). Three cases of tracheal mantle cell lymphoma have been reported to date (5-7). In only one of them, the trachea was the primary site of involvement (7). In the other two cases, the trachea was the site of relapse after initial treatment and one of the multiple involvements (5, 6). The previous case of primary tracheal mantle cell lymphoma was a 75 year-old patient who also presented with acute dyspnea (7). In this paper, the second primary tracheal mantle cell lymphoma case presenting with acute and progressive dyspnea is reported.

### CASE REPORT

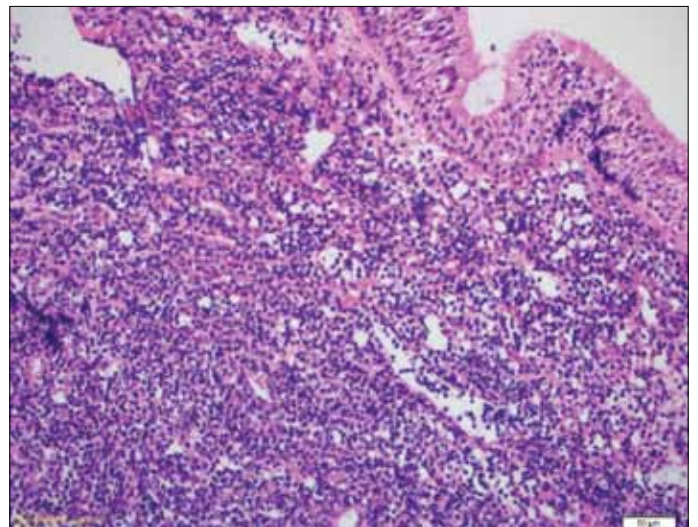
A 58-year-old woman was admitted to our clinic with acute and progressive dyspnea, stridor and confusion. The patient was from Pakistan and could not cooperate due to her state of confusion and language problems. Her history was taken from the patient's son. He stated that a mild dyspnea had been present for one week but had worsened within the preceding 24 hours. He also stated that she had become confused within the last few hours. The medical history of the patient was unremarkable except for diabetes mellitus. She was dyspneic and had biphasic stridor at presentation. In the laryngoscopic examination of the patient, the larynx was normal however there was a regular bordered endotracheal mass 1 cm below the glottic level and obstructing  $\frac{3}{4}$  of the trachea from the posterior segment. Her oxygen saturation was between 60%-70%. Her blood glucose level was 375 mg/dL and serum creatinine level was 2.6 mg/dL. In the urine test, glycosuria was detected. She was diagnosed with prerenal acute renal failure, diabetic ketoacidosis and acute airway obstruction. Renal failure was thought to be related to the worsening of her diabetic situation. On laryngeal computed tomography images, a mass narrowing the tracheal lumen posteriorly through a 5 cm segment in the subglottic area and the trachea was detected (Figure 1). The patient underwent an emergency tracheostomy and direct laryngoscopic examination. Multiple biopsies were taken from the tracheal mass. The patient had treatment for diabetic ketoacidosis and renal failure. Pathological examination of the specimen was reported as a diffuse pattern of mantle cell lymphoma (Figure 2, 3). In the immunohistochemical study, CD20, CD5, Cyclin-D1, BCL-2 receptors were positive and CD10, CD3, CD30 and CD21 receptors were negative. Lymphoma was evaluated as a coincidental situation and it was thought that lymphoma was not related to the diabetic situation or renal failure. However, together with the tracheal lymphoma, other two systemic factors probably had roles in the deterioration of the clinical situation and general health status of the patient. The patient was referred to the medical oncology department in our institution for further management.

### DISCUSSION

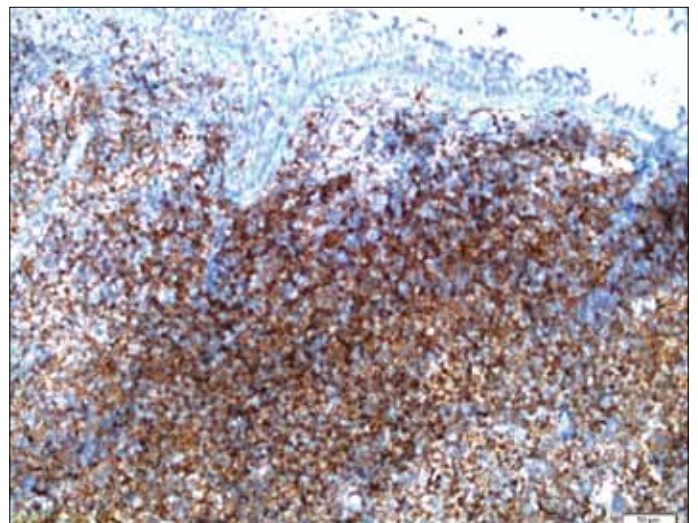
Tracheal neoplasms account for less than 0.1% of all malignancies (8). In spite of their low incidence, these tumors represent a potentially lethal phenomenon. Tracheal tumors may be primary or more commonly, secondary to the invasion of tumors arising from neighboring structures. Primary tracheal tumors develop very rarely, with an incidence of about 0.1 per 100,000 person and over 90% of these are malignant. More than 90% of the cases are squamous cell and adenoid cystic carcinomas (9). Since they are often misdiagnosed



**Figure 1.** Axial neck CT scan showing an intraluminal mass narrowing the trachea posteriorly



**Figure 2.** Mantle cell lymphoma. Normal respiratory epithelium and diffuse infiltration composed of atypical lymphoid cells with hyperchromatic nucleus, scant cytoplasm is seen (H&E x200)



**Figure 3.** Mantle cell lymphoma. CyclinD1 is positive (IHC x200)

as asthma or chronic lung disease, the diagnosis can be delayed for years. Laryngotracheal involvement of lymphoma is an uncommon cause of upper airway obstruction. Primary non-Hodgkin's lymphoma of the trachea is even more rarely encountered, representing 0.23% of all tracheal tumors (10). According to a review by Takami et al. (11) the most common presenting symptoms of tracheal lymphoma cases are dyspnea, cough, stridor, and wheezing. Hemoptysis is an uncommon symptom, due to the submucosal localization of the tracheal lymphomas. Airway obstruction is reported in 87% of the patients, half of whom require an emergency intervention.

According to the WHO classification, lymphomas are categorized as B cell lymphomas, T cell lymphomas and Hodgkin's disease. B cell lymphomas are precursor B lymphoblastic lymphoma, extranodal marginal zone lymphoma, mantle cell lymphoma, follicular lymphoma, diffuse large B cell lymphoma and Burkitt's lymphoma (12). Mantle cell lymphoma is a subtype of non-Hodgkin lymphoma, which is commonly seen in older male adults. A minority of these patients present with extranodal disease, most often involving the gastrointestinal tract, occasionally the Waldeyer ring, but rarely they present with the involvement of other extranodal sites, such as the skin or the ocular adnexa. The mantle cell lymphoma presenting in extranodal sites has cytologic and immunophenotypic features similar to the mantle cell lymphomas in lymph nodes. Neoplastic cells are usually small to medium-sized with slightly irregular, dark nuclei and scant cytoplasm. When the gastrointestinal tract is involved, the most common manifestation is multiple lymphomatous polyposis involving the mucosa, sometimes superficially involving the submucosa. The neoplastic cells are typically CD20<sup>+</sup>, CD5<sup>+</sup>, CD10<sup>+</sup>, CD23<sup>+</sup>, cyclin D1<sup>+</sup> and bright surface immunoglobulin<sup>+</sup> (12).

The case presented herein is the second reported case of mantle cell lymphoma in the literature. Primary tracheal tumors do not always progress slowly; as in our case, they may also present with acute and progressive dyspnea. After maintenance of airway, a biopsy should be taken to rule out any neoplastic condition in any suspicious case.

### Conflict of Interest

No conflict of interest was declared by the authors.

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