# Atypical Lipomatous Tumor Originating From the Nasopharynx in a Patient with Chronic Lymphocytic Leukemia

Case Report

Mehmet Emre Sivrice[1], Vural Akın[1], Bekir Büyükçelik[1], Hasan Yasan[1], Mehmet Kıran[2], Veysel Atilla Ayyıldız[3]

[1]Department of Otorhinolaryngology and Head and Neck Surgery, Süleyman Demirel University Faculty of Medicine, Isparta, Turkey
[2]Clinic of Pathology, Isparta City Hospital, Isparta, Turkey
[3]Department of Radiology, Süleyman Demirel University Faculty of Medicine, Isparta, Turkey

## Abstract

Atypical lipomatous tumor, also known as well-differentiated liposarcoma, is rare in the head and neck region. The primary and most effective option in the treatment of this malignancy with a good prognosis is excision with clean surgical margins. Therefore, it is important to distinguish this malignancy from lesions that require more aggressive treatment. In this article, we present the case of an atypical lipomatous tumor originating from the nasopharynx and almost completely obstructing the oropharynx in a 38-year-old male patient with chronic lymphocytic leukemia. To the best of our knowledge, this is the first report in the literature of an atypical lipomatous tumor case originating from the nasopharynx.

**Keywords:** Nasopharynx, atypical lipoma, liposarcoma, chronic lymphocytic leukemia, neoplasms, case report

***ORCID IDs of the authors:***

*M.E.S. 0000-0002-2396-6794;*
*V.A. 0000-0002-0050-4837;*
*B.B. 0000-0002-3620-2443;*
*H.Y. 0000-0002-5470-6784;*
*M.K. 0000-0003-3292-2213;*
*V.A.A. 0000-0003-0252-9023.*

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**Corresponding Author:**
Mehmet Emre Sivrice;
emresivrice@gmail.com

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## Introduction

Liposarcoma is the most common type of sarcoma in adults. In the classification of head-neck tumors published by the World Health Organization in 2017, liposarcomas are histologically classified into three subgroups: well-differentiated, myxoid, and pleomorphic. The well-differentiated liposarcoma is also known as an atypical lipomatous tumor (1, 2). The head and neck location comprises only 5.6% of all liposarcoma cases (3). In this region, they can originate in the soft tissues of the pharynx, the larynx, the oral cavity, and the neck (1, 2, 4). There are no reported cases of atypical lipomatous tumors localized in the nasopharynx. In this article, we present the case of a patient with chronic lymphocytic leukemia (CLL) with an atypical lipomatous tumor originating from the nasopharynx.

### Case Presentation

A 38-year-old male patient presented to our clinic with a mass in his throat. He stated that he had noticed the mass for the first time two years ago and the mass had grown over time. He reported that he had increasing dyspnea, dysphagia, and obstructive sleep apnea and that in recent months, he could only swallow liquid

foods. He had a history of CLL. A well-circumscribed mass with yellow-pink-colored mucosa and local vascularity was observed. The mass was completely obstructing the passage between the soft palate and the base of the tongue and pushing the uvula to the anterosuperior. The pedicle of the mass protruded from the inferomedial region of the torus tubarius on the right side of the nasopharynx (Figure 1). The patient had contrast-enhanced magnetic resonance imaging (MRI) performed 22 months ago and computed tomography performed 11 months ago (Figures 2, 3). The patient underwent orotracheal intubation with a video laryngoscope. The soft palate was suspended with a feeding tube inserted through the nostril. The area where the mass originated from the nasopharyngeal wall was also visualized by nasal endoscopy. The pedicle of

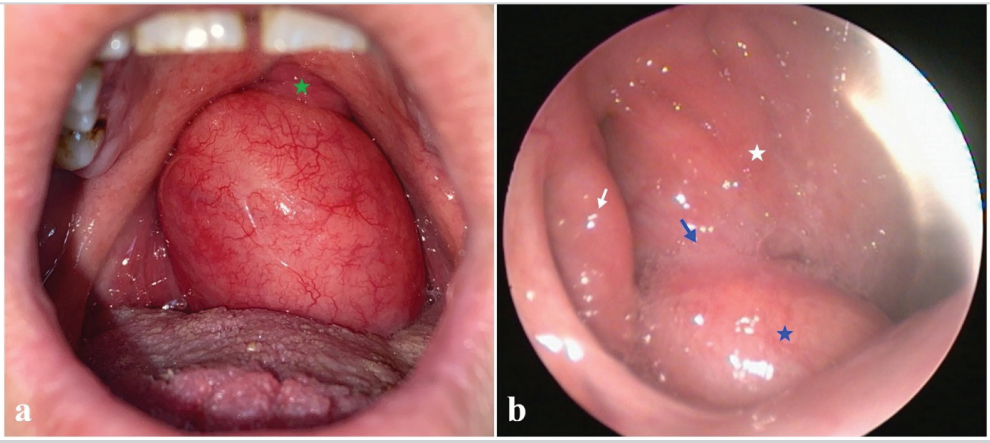


**Figure 1.** Gross appearance of the lesion: a) Oropharyngeal view, b) Nasopharyngeal view (green asterisk: uvula; white arrow: torus tubarius; white asterisk: posterior wall of the nasopharynx; blue asterisk: mass; blue arrow: peduncle of the mass)

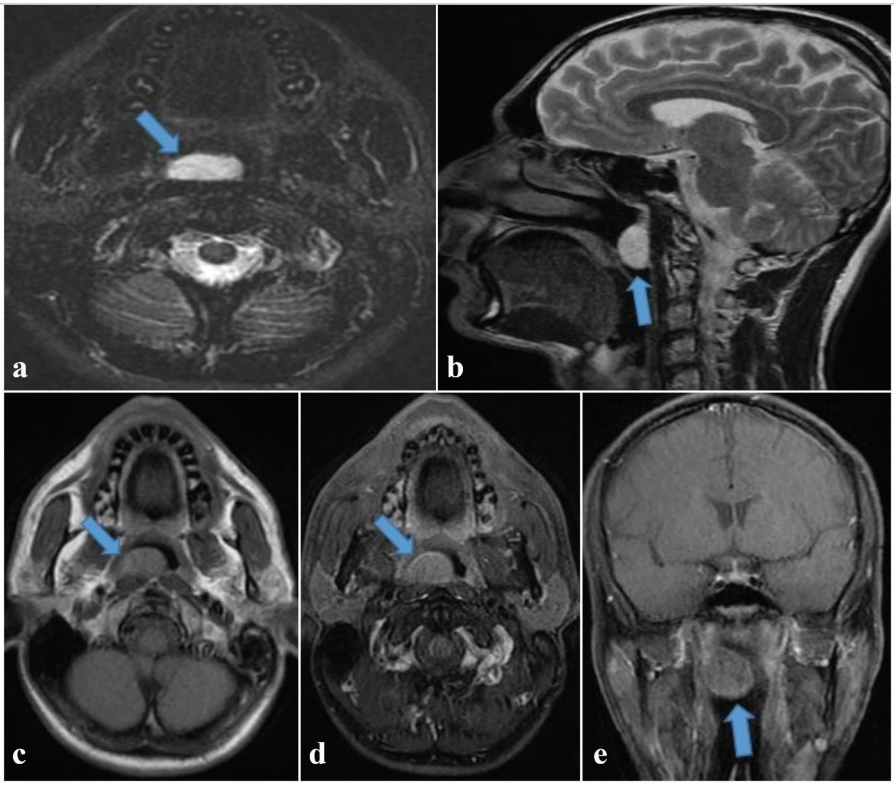


**Figure 2.** Contrast-enhanced nasopharyngeal MRI images taken 22 months before admission: a) Fat-suppressed T2W axial view, b) T2W sagittal view, c) T1W axial view, d) Contrast-enhanced T1 view, e) Fat-suppressed T1 coronal view (blue arrow: mass). MRI revealed a mass on the right side of the midline extending from the posterior inferior wall of the nasopharynx to the oropharynx, hyperintense in fat-suppressed T2W, hypointense in T1W, and without contrast enhancement on postcontrast fat-suppressed T1-weighted images. The MRI demonstrated that the mass was a 23×13×27 mm homogeneous lesion with an oval shape, smooth contours, and no contrast-enhancing septal structure

MRI: Magnetic resonance imaging

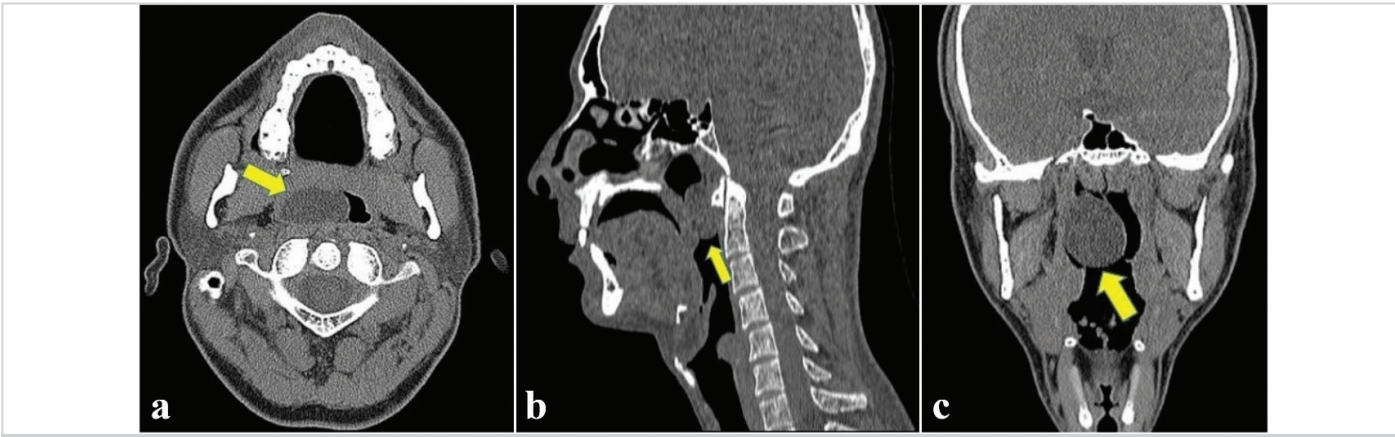


**Figure 3.** Non-contrast paranasal sinus computed tomography images taken 11 months before admission: a) axial, b) sagittal, and c) coronal (yellow arrow: Mass). The paranasal sinus CT report described the mass as a homogeneous lesion with dimensions of 29×14×30 mm and an average density of -5 Hounsfield Units. It was an oval-shaped, well-contoured capsulated mass structure extending from the posteroinferior wall of the nasopharynx to the oropharynx on the right side of the midline

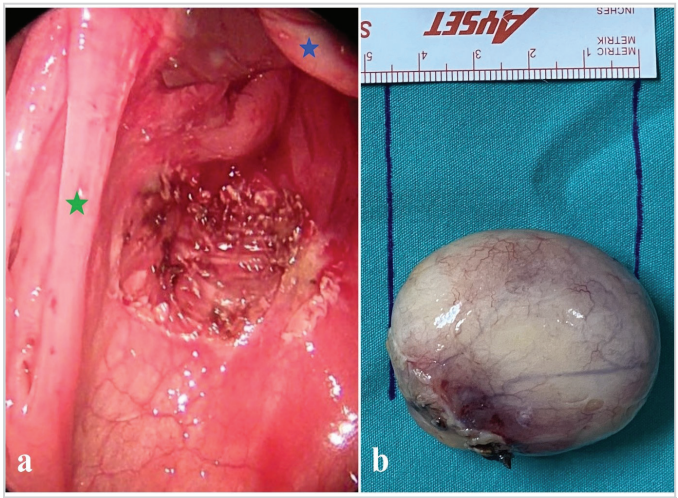


**Figure 4.** a) The location of the pedicle after removal of the mass (green asterisk: posterior plica; blue asterisk: uvula), b) gross specimen

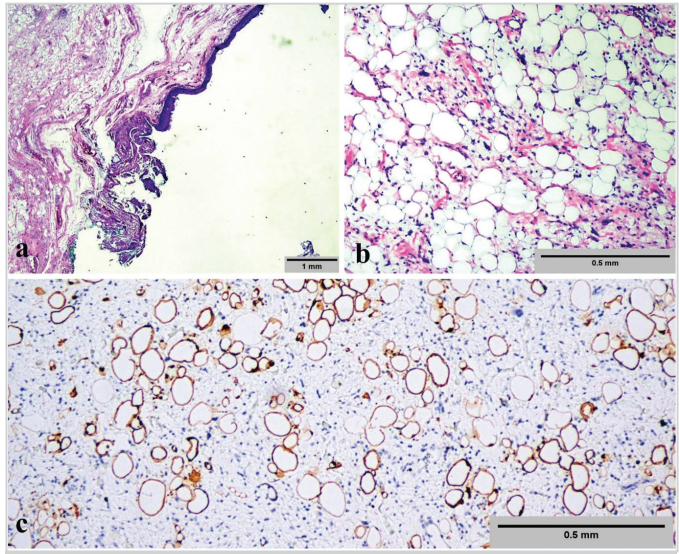


**Figure 5.** Histologic features of the mass: a) a well-circumscribed and thinly encapsulated lesion adjacent to the surface epithelium (H&E stain x20), b) variably sized adipocytes (H&E stain x100), c) positive staining with S100 was observed in adipocytes (S100 DAB, x100)

the mass was dissected with a transoral approach using bipolar cautery (Figure 4). Histopathological examination of the mass reported an atypical lipomatous tumor. Surgical margins were clean, and lymphovascular invasion was not observed. The tumor had patchy staining with CD34. Ki-67 positive cells were 5% (Figure 5). At the one-month follow-up, the patient's breathing and swallowing complaints were completely resolved (Figure 6). We asked for the results of the patient's genetic examination tests performed for CLL in previous years. He was positive for 11q22.3 deletion. No recurrence was observed in the patient's six-month follow-up examination. He was followed up clinically at three-month intervals.

Informed consent was obtained from the patient.

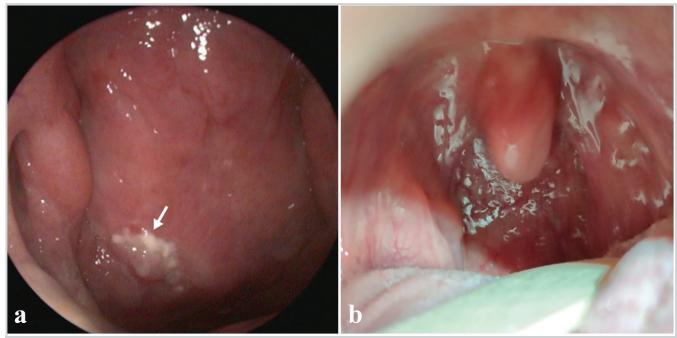


**Figure 6.** a) Nasopharynx and b) oropharynx view one month after the operation (white arrow: granulation tissue at excision site)

## Discussion

Radiation, trauma, and genetic factors are blamed for the etiology of liposarcomas (2). Our patient did not have a history of radiation or trauma, but in his genetic tests for CLL, 11q22.3 (ATM) deletion was positive. This deletion has been reported as associated with a poor prognosis for CLL patients (5). No study has shown the relationship between this deletion and liposarcomas. 11q22.3 deletion has been associated with some solid cancers as well as hematological malignancies (6). This deletion, for which a direct relationship with the development of atypical lipomatous tumors could not be established, needs to be evaluated with further studies and case series. Moreover, the association of CLL with an atypical spindle cell lipomatous tumor was reported in one case (7). The fact that another case of atypical lipomatous tumor with CLL has been reported in the literature also makes our case interesting.

Liposarcomas develop more frequently in men, and the average age at diagnosis is 60 years (1, 2). These sarcomas are characterized by slow-growing, submucosal, well-circumscribed, and painless masses (1, 2). They are usually encapsulated (2). Symptoms vary by region of origin. Those located in the oral cavity, the larynx, and the pharynx may cause dysphagia and dyspnea, depending on their size (1, 2). In their article published in 2012, Papacharalampous et al. (8) reported a case of myxoid-round cell liposarcoma with moderate differentiation located in the nasopharynx. This was the case of a 58-year-old man, and chemoradiotherapy was applied to the patient because curative surgery was not possible. No recurrence was observed in the 14-month follow-up of the patient. In the same article, the authors stated that they had identified four more cases of nasopharyngeal localization in the English literature review. Two of these cases were female and two were male, the youngest was 12 years old and the oldest was 58 years old. In one of these cases, the histopathological subtype could not be determined, while two of them showed myxoid features, and one was a well-differentiated sclerosing subtype. In these five cases, cure could not be achieved with surgical excision alone. Radiotherapy was also applied in all cases, and chemotherapy was added to the treatment in two cases (8). In 2022, Nishith et al. (9) reported a case of low-grade dedifferentiated liposarcoma originating from the nasopharynx. This was a 36-year-old male patient and chemoradiotherapy was preferred. Unlike the other nasopharyngeal liposarcomas reported in the literature, our case could only be treated with excision. The reasons for this are that the lesion was pedicled and well-differentiated.

MRI is extremely helpful in the differential diagnosis of liposarcomas. In well-differentiated liposarcomas, the MRI image of the tumor is similar to that of normal adipose tissue. However, contrast enhancement indicates the level of differentiation. Contrast enhancement is minimal in well-differentiated lesions, while more intense enhancement is observed in more aggressive subtypes (4).

Atypical lipomatous tumors have enlarged adipocytes of varying sizes, with hyperchromatic and enlarged nuclei (1, 2). They can be distinguished from benign lipomas because they contain multivacuolar lipoblasts. Immunohistochemical staining of lipoblasts with S-100 is essential for differential diagnosis (4). The tumor consists of lobules of adipose tissue separated by a thick fibrous band. Mild cytological atypia may be observed in a well-differentiated liposarcoma (10). Necrosis and mitotic activity are rare (4). More than 90% of liposarcomas are MDM2- and CDK4-positive. Likewise, 12q13-15 amplification is positive in atypical lipomatous tumors (1). Detection of MDM2 amplification is a sensitive and specific method for distinguishing well-differentiated liposarcoma from benign lipoma (10). The histopathological examination results of our case are also compatible with the literature. The mass consisted of well-circumscribed, encapsulated lesions with large nuclei and S-100 positive staining adipocytes.

Excision of the lesion with clean surgical margins usually provides effective treatment (1, 2). Localization and tumor grade are important factors in prognosis (1, 2). The rate of distant metastases in atypical lipomatous tumors is reported as less than 6%, and they most commonly metastasize to the lung. Regional nodal metastases are also extremely rare (2, 10). Atypical lipomatous tumors can recur at a rate of 30%, so long-term follow-up is essential (2). Radiotherapy may be preferred in unresectable tumors, although the effectiveness of radiotherapy is unclear, and there is no evidence of the efficacy of chemotherapy (2, 10). Regional lymph node dissection is not recommended (2). These tumors can sometimes differentiate or transform into high-grade sarcomas. This transformation occurs as a result of a delay in treatment or inadequate excision (4).

## Conclusion

To our knowledge, this is the first reported case of an atypical lipomatous tumor originating from the nasopharynx. This pathology, with typical clinical and radiological features, should be included in the differential diagnosis of nasopharyngeal masses. The association with CLL and 11q22.3 deletion needs to be investigated in further studies.

**Informed Consent:** Informed consent was obtained from the patient.

**Peer-review:** Externally and internally peer-reviewed.

**Authorship Contributions**

Surgical and Medical Practices: M.E.S., V.A., B.B., H.Y., Concept: M.E.S., V.A., B.B., H.Y., M.K., V.A.A., Design:

M.E.S., V.A., H.Y., M.K., V.A.A., Data Collection and/or Processing: M.E.S., V.A., B.B., M.K., V.A.A., Analysis and/or Interpretation: M.E.S., V.A., M.K., V.A.A., Literature Search: M.E.S., V.A., B.B., H.Y., M.K., V.A.A., Writing: M.E.S., V.A., B.B., H.Y., M.K., V.A.A.

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**Main Points**

- An atypical lipomatous tumor, also known as well-differentiated liposarcoma, is rare in the head and neck region.
- This is the first case in the literature of an atypical lipomatous tumor originating from the nasopharynx.
- Excision of the lesion with clean surgical margins usually provides effective treatment.
- Further studies are needed to show the relationship of atypical lipomatous tumor with chronic lymphocytic leukemia and 11q22.3 deletion.

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