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ENTCase

CONGENITAL CYSTIC MASS WITH ATYPICAL LOCATION IN AN ELDERLY PATIENT

YAŞLI BİR HASTADA ATİPİK YERLEŞİMLİ KONJENİTAL KİSTİK KİTLE

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Özet

Tiroid kalıntılarından sonra boyunda en sık görülen doğumsal boyun kitlesi brankial ark anomalileridir. İkinci brankial ark anomalisi en sık görülenidir, üçüncü ve dördüncü tip ise nadirdir. Bunlar öncelikle iyi huylu kitleler olmasına rağmen primer veya metastatik tümör oluşumu nadirdir. Genellikle boyun ve faringeal bölgede görülmekle birlikte, özellikle yaşlı hastalarda larinkste son derece nadir görülmektedir. Çalışmamızda kronik lenfositik lösemi ile takipli 82 yaşındaki hastanın alt dudağında iyileşmeyen ülser gelişmesiyle polikliniğimize başvurması ve alt dudakta skuamöz hücreli karsinom ile birlikte nazofaringeal ve boyun bölgelerinde lenfoma ile laringeal bölgede sağ ariepiglottik kıvrımı işgal eden son derece nadir görülen brankial yarık kisti olan olguyu sunmak amaçlandı. Bu kist endoskopik olarak başarıyla çıkarıldı. Beş aylık takip sürecinde nüks görülmedi. Laringeal brankial ark anomalilerinin bu yaş grubunda nadir görülmesi yönüyle de literatürde ilk birkaç vaka arasında yer almaktadır. Laringeal bölgeden köken alan brankial yarık kistleri nadir olduğundan baş boyun maligniteli hastalarda bu bölgedeki kistik lezyonları değerlendiren klinisyenlerin ayırıcı tanısında brankial yarık anomalilerini de dikkate alması gerekir.

Anahtar kelimeler: Boyun kitlesi, brankial ark anomalileri larenks lenfoma konjenital

Abstract

After thyroid remnants, the most common congenital neck mass in the neck is branchial arch anomalies. The second branchial arch anomaly is the most common, while the third and fourth types are rare. Although these are primarily benign masses, primary or metastatic tumor formation is rare. They are generally seen in the neck and pharyngeal region but are extremely rare in the larynx, especially in elderly patients. In our study, we aimed to present the case of an 82-year-old patient with chronic lymphocytic leukemia, who applied to our clinic with a non-healing ulcer on his lower lip, squamous cell carcinoma on the lower lip, lymphoma in the nasopharyngeal and neck regions, and an extremely rare branchial cleft cyst occupying the right aryepiglottic fold in the laryngeal region. This cyst was successfully removed endoscopically. No recurrence was observed during the five-month follow-up period. Since laryngeal branchial arch anomalies are rare in this age group, they are among the first few cases in the literature. Since branchial cleft cysts originating from the laryngeal region are rare, clinicians evaluating cystic lesions in this region in patients with head and neck malignancies should also consider branchial cleft anomalies in the differential diagnosis.

Keywords: Neck mass, branchial arch anomaly larynx lymphoma congenital

Introduction

In the early weeks of embryonic development, significant structures in the head and neck region begin to form. The causes of various malformations and deformities in the neck arise from insufficient or unsuccessful embryological development of pharyngeal arch structures. As a result, branchial cleft sinuses, fistulas, or cysts develop [1]. Branchial cleft anomalies in the form of neck swellings are more common after birth, especially in the first two decades following an episode of infection. The most common presentation of the second branchial arch anomaly is mass-like swelling on the lateral aspect of the neck. According to Bailey's classification, branchial cleft

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cysts of type 2 have four different subtypes based on the localisation of significant neck vessels; the fourth subtype, which occurs in the parapharyngeal and retropharyngeal areas, is very rare and generally presents with mild symptoms [2]. Laryngeal cysts are rare and can occur in males and females of any age. They are usually benign and exhibit non-threatening behaviour. Laryngeal cysts, also known as saccular and ductal epithelial cysts, are classified according to their origin and location. Researchers have also categorised branchial cysts as ductal cysts [5]. The literature review reported nasopharyngeal carcinoma metastasis as an intrathoracic extension or parapharyngeal mass [2-4]. Laryngeal branchial cleft cyst anomalies are rare. This study presents a case of an advanced-age patient who developed a laryngeal branchial cleft cyst anomaly following multicentric synchronous malignancies.

Case Report

An 82-year-old female patient with a medical history of dermatomyositis and chronic lymphocytic leukaemia presented to our clinic with a nonhealing sore on the lower left lip that had persisted for approximately two months. The authors stated that the study's patient who presented with images obtained written consent. During the otolaryngological examination, an ulcerative lesion on the lateral left aspect of the lower lip, posterior fullness in the nasopharynx, a cystic nodular lesion occupying the right aryepiglottic fold in the larynx, and multiple neck lymphadenopathies, with the largest measuring 3.5 cm. A wedge biopsy of the lesion on the lower left lip and punch biopsy of the posterior nasopharynx revealed squamous cell carcinoma (SCC) in the lower lip and lymphoma in the nasopharynx. Computed tomography (CT) and magnetic resonance imaging (MRI) of the nasopharynx and neck revealed a smooth-bordered cystic nodular lesion, approximately 2 cm in size, filling the right aryepiglottic region with slight contrast enhancement (Figure 1-2).

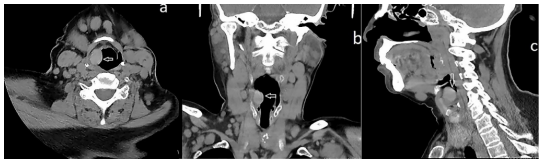

Figure 1

In the neck computed tomography, axial (a), coronal (b), and sagittal (c) sections reveal a cystic mass appearance in the right aryepiglottic region. (white arrow: cystic mass)

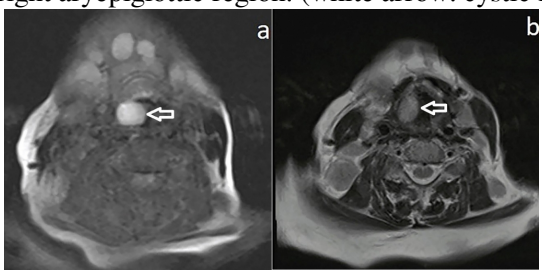

Figure 2

Contrast-enhanced neck magnetic resonance imaging, axial T1-weighted (a), and T2-weighted (b) sections show a nodular cystic mass observed on the right side of the laryngeal region. (white arrow: cystic mass)

A multidisciplinary tumour board meeting discussed this patient with multiple synchronous primary malignancies. The surgical procedure encompassed the meticulous excision of the tumoral mass on the left lateral lower lip, employing an appropriate safety margin, followed by a comprehensive reconstruction utilising the primary V flap technique. Concurrently, the surgical team performed a diagnostic excision of lymphadenopathy in response to multiple lymphadenopathies on the patient's left neck.

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Furthermore, a transoral endoscopic approach is employed to excise the cystic mass in the laryngeal region at the right aryepiglottic area. Figure 3 illustrates endoscopic views of the cystic mass in the laryngeal region before and one month after surgery. Histopathological examination revealed SCC of the lower lip, marginal zone lymphoma in the neck lymph node, and a branchial cleft cyst in the lesion on the right side of the laryngeal region. The surgical team utilised a transoral endoscopic approach to excise the laryngeal cyst. During the five-month follow-up period, no recurrence was observed in the laryngeal area, while the haematology department monitored the diagnosis of nasopharyngeal and neck lymphoma.

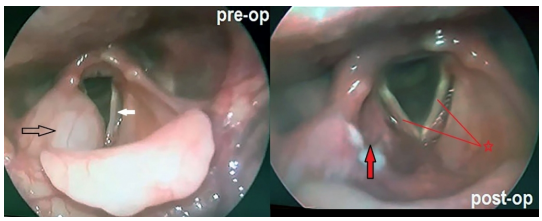

Figure 3

Endoscopic view of the laryngeal area before the surgical procedure for the laryngeal cystic mass and one month after the surgery. (black arrow cystic mass, white arrow: left vocal cord, red arrow: right aryepiglottic area after removal of the cystic mass, red star: the appearance of the vocal cords)

Discussion

Branchial cleft anomalies are developmental cysts whose pathogenesis remains controversial. Researchers have described these cysts as cervical lymphoepithelial cysts. During the pediatric period, the most commonly observed developmental anomalies in the head and neck region were the second most common. Due to inadequate obliteration of embryonic remnants, they can manifest as cysts, sinuses, or fistulas [6]. Branchial arches are fundamental structures that play a role in the embryological development of the face, neck, and pharyngeal structures. Type 2 is the most common branchial cleft anomaly [7]. Type 2 has four subtypes described by Bailey, and the least frequently observed subtype is the fourth [1] branchial cleft anomaly, usually present in the hospital due to swelling in the neck. Branchial cleft anomalies in the neck often present as masses and sometimes extend to the intrathoracic region. Those located in the pharyngeal area can cause disturbances in swallowing and vocal functions. In the diagnostic approach, neck ultrasonography, CT, and MRI are used for evaluation if necessary. It generally develops on the lateral aspect of the neck.

During the literature review, the development of branchial cleft cysts in the laryngeal area was a rare occurrence, according to the observations made by researchers. Treatment involves the excision of the mass in the neck or pharyngeal region followed by close monitoring [1,4].

Ida et al. reported a branchial cleft cyst in the neck specimen of a patient who underwent laryngectomy due to laryngeal carcinoma. A cystic lesion is a third branchial cleft cyst anomaly [8]. Bochnia et al. reported the case of a 55-year-old female patient with a similar presentation. The patient had a laryngeal branchial arch fistula originating from a fourth branchial arch anomaly that involved the right aryepiglottic fold. Researchers have noted a scarcity of laryngeal branchial arch anomalies after the fourth decade of life. [5]. The patient was diagnosed with hypertension and dermatomyositis and underwent transoral laryngoscopy-guided excision of a well-defined cystic mass originating from the right laryngeal aryepiglottic region, along with malignant lesions in the lower lip, nasopharynx, and neck. Histopathological examination revealed a branchial cleft cyst; we detected no recurrence during the 5th-month follow-up. In this case, we identified the cystic mass as a 4th branchial arch anomaly because of its localisation. Transoral excision ensured the avoidance of neck scar formation and potential neurovascular complications.

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This study was presented as a poster at the 15th International Congress of Otolaryngology and Head and Neck Surgery, June 9-11, 2023, in Ankara, Turkey (ONLINE).