CASE REPORT OLGU SUNUMU

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Low-Grade Myxofibrosarcoma of the Larynx: A Rare Case Involving the Arytenoid Region

Larenksin Düşük Dereceli Miksofibrosarkomu: Aritenoid Bölgesini Tutan Nadir Bir Vaka

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ABSTRACT Laryngeal sarcomas are rare malignancies, comprising 1% of all laryngeal cancers. Myxofibrosarcoma (MFS), a fibroblastic tumor typically arising in the extremities, is highly uncommon in the head and neck region, particularly in the larynx. We report the case of a 79-year-old man with progressive dyspnea and dysphagia. Endoscopic examination revealed a supraglottic mass involving the left arytenoid and aryepiglottic fold, extending into the left pyriform sinus and tongue base. Computed tomography demonstrated a calcified mass obstructing the left pyriform sinus and involving the posterior laryngeal wall. The mass was surgically removed through both the endolaryngeal and transoral approaches. Histopathological evaluation confirmed the diagnosis of low-grade MFS. Laryngeal MFS is with only a few cases reported in the literature. Surgical resection remains the primary treatment. This case highlights an unusual location and underscores the importance of considering sarcomas in the differential diagnosis of laryngeal masses in elderly patients.

Keywords: Laryngeal neoplasms; sarcoma; head and neck neoplasms; arytenoid cartilage

ÖZET Larenks sarkomları nadir görülen malignitelerdir ve tüm larenks kanserlerinin %1'inden azını oluştururlar. Genellikle ekstremitelerde ortaya çıkan fibroblastik bir tümör olan miksofibrosarkom, özellikle larenkste olmak üzere baş ve boyun bölgesinde son derece nadirdir. Bu yazıda, ilerleyen dispne ve disfaji ile başvuran 79 yaşında bir erkek hastanın olgusu sunulmaktadır. Endoskopik incelemede sol aritenoid ve ariepiglotik plikayı içeren, sol piriform sinüse ve dil tabanına uzanan supraglottik bir kitle görüldü. Bilgisayarlı tomografide sol piriform sinüsü tıkayan ve posterior laringeal duvarı tutan kalsifiye bir kitle görüldü. Kitle cerrahi olarak endoskopik ve transoral yaklaşımla çıkartıldı. Histopatolojik değerlendirmesinde ise düşük dereceli miksofibrosarkom tanısını doğruladı. Larenks miksofibrosarkomu literatürde birkaç vaka bildirilmiştir. Cerrahi rezeksiyon birincil tedavi olmaya devam etmektedir. Bu vaka nadir bir tümör yerleşimini vurgulamakta ve yaşlı hastalarda laringeal kitlelerin ayırıcı tanısında sarkomların dikkate alınmasının önemini vurgulamaktadır.

Anahtar Kelimeler: Larenks neoplazmaları; sarkoma; baş ve boyun neoplazmaları; aritenoid kıkırdağı

Most of the laryngeal cancers are squamous cell carcinomas. However, other rare malignancies such as adenocarcinomas, sarcomas, lymphomas, and neuroendocrine tumors may also arise in the larynx.[1] Sarcomas constitute less than 1% of head and neck malignancies.[2] These tumors originate from mesodermal tissues and demonstrate a broad spectrum of biological behavior, ranging from slow-growing lesions to aggressive tumors with a high actantial for metastasis. The most common sites of involvement

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in the head and neck region include the neck, face, forehead, and paranasal sinuses.[3] Laryngeal sarcomas account for less than 1% of malignant laryngeal neoplasms, with chondrosarcoma being the most frequently encountered type.[4]

The evaluation of laryngeal masses typically begins with a thorough clinical history and physical examination. Laryngoscopic examination, either flexible or direct, enables direct visualization of the lesion. Imaging techniques such as contrast-enhanced computed tomography (CT) and magnetic resonance imaging are useful in assessing the size, extent, and anatomical relationships of the mass. For a definitive diagnosis and effective treatment planning, obtaining a histopathological evaluation through direct laryngoscopic biopsy is considered the gold standard.[1]

Sarcomas can develop in any part of the larynx, similar to carcinomas; however, they are typically located in the vocal cords. Laryngeal myxofibrosarcoma (MFS) is a rare entity, with only 8 cases reported in the literature. Of these, 3 were diagnosed as low-grade MFS. This report aims to expand the limited literature and improve the clinical recognition and management strategies for this rare laryngeal tumor among otolaryngologists.

CASE REPORT

A 79-year-old male patient presented to our clinic with progressive dyspnea and dysphagia over the past month. He had coronary artery disease and chronic obstructive pulmonary disease. He also has a 50 pack-year smoking history but has not smoked in the last decade. There was a mass starting from the left arytenoid, filling the left pyriform sinus, extending to the base of the tongue and supraglottic region and narrowing the rima glottis in the endoscopic examination. A locally calcified heterogeneous mass of 35-25 mm in the broadest part, obliterating the left pyriform sinus, narrowing the air column, and being more prominent on the left at the supraglottic level, was observed on the posterior wall of the larynx in the neck contrast-enhanced CT (Figure 1). Thorax CT: No mass or pathological lymph nodes were detected. Surgery was performed via both endolaryngeal and transoral approaches. The nodular mass

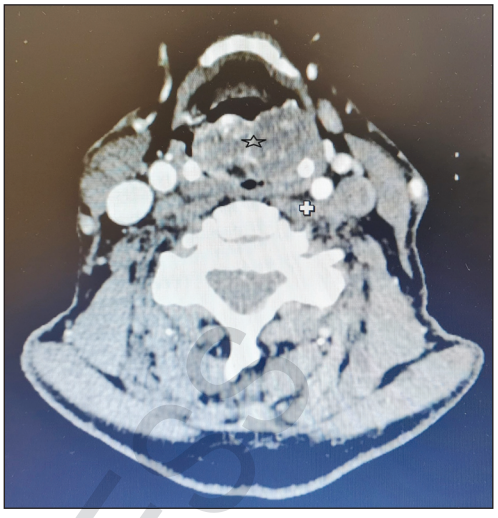

FIGURE 1: Larynx CT: 35*25 mm calcified heterogeneous mass obliterating the left pyriform sinus on the posterior wall of the larynx at the supraglottic level (black asterisk)
CT: Computed tomography

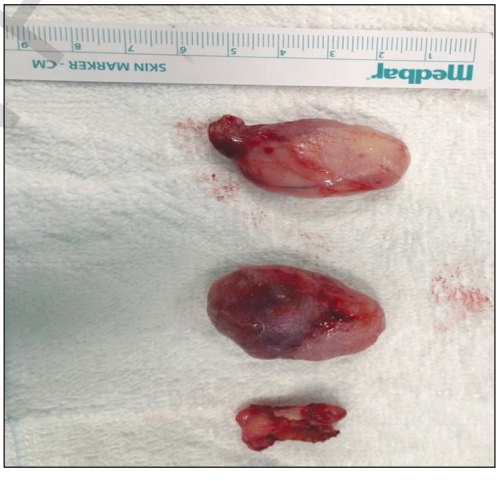

FIGURE 2: Nodular mass in 3 pieces as 4*2.5 cm, 3*2 cm, 2*1.5 cm originating from the left arytenoid, filling the left pyriform sinus, having supraglottic and tongue root extension

originating from the left arytenoid, filling the left pyriform sinus, extending to the supraglottic and tongue root, was excised in 3 pieces as 4-2.5 cm, 3-2 cm, and 2-1.5 cm (Figure 2).

On histopathological examination, a multinodular tumor was detected under the laryngeal epithelium (Figure 3a). The tumor had a myxoid stroma and consisted of hypocellular and hypercellular areas. There is a fascicular pattern, particularly in the cellular areas (Figure 3b). The tumor cells were spindle-shaped with atypical hyperchromatic nuclei (Figure 3c). Oc-

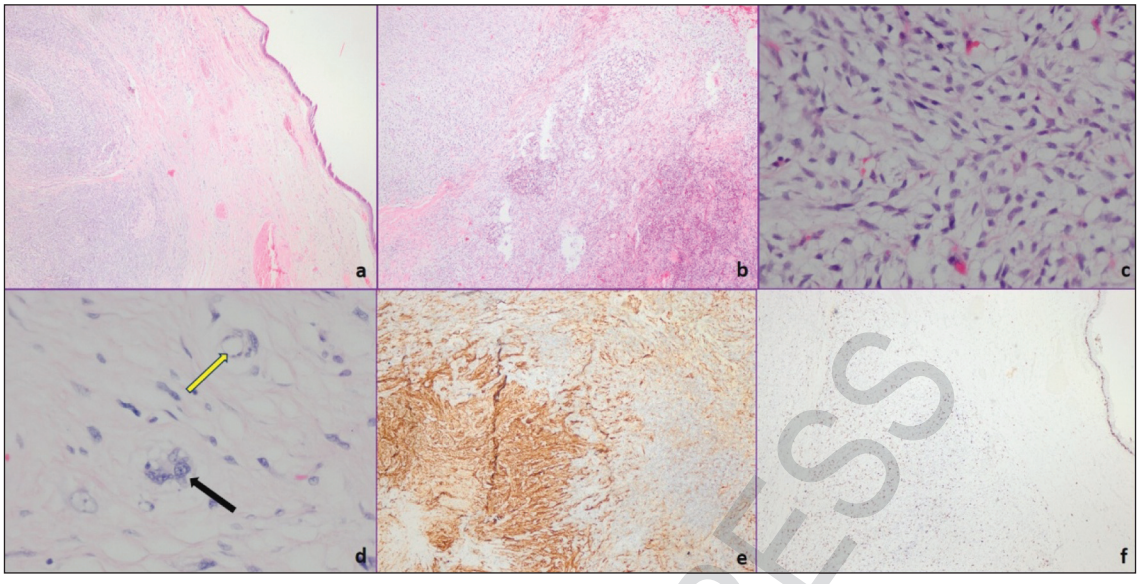

FIGURE 3: a) Lobulated tumor with multinodular growth pattern (H&E, x40). **b)** Alternate hypocellular and hypercellular areas (H&E, x40). **c)** spindle to stellate cells with moderate cytoplasm and atypical, hyperchromatic nuclei in abundant myxoid stroma (H&E, x400). **d)** pleomorphic and touton type giant cells (black arrow) and pseudolipoblasts (vacuolated fibroblasts) (yellow arrow) (H&E, x400). **e)** Immunohistochemical SMA positivity (H&E, x40) **f)** Ki-67 proliferation index (H&E, x40);
H&E: Hematoxylin-eosin; SMA: Smooth muscle actin

casional pleomorphic and Touton-type giant cells were also observed. Some tumor cells had peripherally located nuclei similar to pseudo-lipoblasts (Figure 3d). Atypical mitoses were seen in the tumor. There was no necrosis. Immunohistochemical studies [Agilent (DAKO-RTU Antibodies, Denmak)] showed that the tumor cells were immunoreactive with vimentin and smooth muscle actin (SMA) (Figure 3e), whereas negative with CD-34, Melan-A, CD68, pancytokeratin (AE1/AE3), S-100, CD-99, and synaptophysin. The Ki-67 (MIB-1) proliferation index was approximately 15% (Figure 3f). The final histopathological diagnosis was low-grade MFS. No complications developed during the postoperative follow-up. No recurrence or metastasis was observed in the follow-up examinations conducted 5 years later.

We received written informed consent from the patient for this case presentation.

DISCUSSION

Head and neck sarcomas are rare types of malignancies. Osteosarcomas, angiosarcomas, rhabdomyosarcomas, and malignant fibrous histiocytomas (MFH) account for over 50% of sarcomas in this region.[5] MFS was first defined by Angervall et al. as a type of fibroblastic lesion characterized by pleomorphic, storiform features similar to those of MFH. Mitotic activity can range from low in hypocellular lesions to high in more cellular ones.[6]

MFS typically occurs in the extremities of older individuals and is characterized by slow and painless growth.[7] Cases of MFS in the head and neck region are quite rare. According to the existing literature, there have been 8 documented cases in the laryngeal area. Of these, 3 were classified as low-grade MFS. In the sunport by Ocak et al. endoscopic examination and imaging were not highlighted.[8] In contrast, our case highlights both endoscopic and imaging findings, making it the first documented case with this specific histological subtype and location.

MFS is divided into 3 or 4 grades depending on cellularity, pleomorphism of the nucleus, and mitotic activity.[6,9] Although the histopathological appearance of MFS may vary according to the grade of the tumor, all cases showed distinct morphological features, such as a multinodular growth pattern and a myxoid stroma composed of hyaluronic acid. Mentzel et al. suggested that MFSs are classified into subtypes as

low, intermediate, and high grade.[9] Low-grade tumors are predominantly hypocellular, containing few atypical tumor cells with enlarged, hyperchromatic nuclei and rare mitotic figures. Characteristic findings include prominent, curved, elongated vessels surrounded by tumor or inflammatory cells and pseudolipoblasts-neoplastic fibroblastic cells with vacuoles. In the immunohistochemical analysis, low-grade MFS typically tests positive for CD34, vimentin, and sometimes SMA and Ki-67. It is negative for the S-100 protein. When making a differential diagnosis, it is important to consider conditions such as nodular fasciitis, myxoid neurofibroma, myxoma, spindle cell lipoma, and malignant peripheral nerve sheath tumor, particularly in cases without transition to higher-grade areas.

The most challenging distinction is between MFS and low-grade fibromyxoid sarcoma. Clinically, low-grade fibromyxoid sarcoma typically occurs in younger patients and originates from deep tissues. This tumor has a propensity for multiple recurrences and distant metastasis, particularly if it is not completely excised. Although it usually arises in the lower extremities, rare cases of involvement in the head and neck have been reported.[10,11] Morphologically, low-grade fibromyxoid sarcoma is characterized by cytologically bland spindle cells arranged in a whorled pattern, embedded in a stroma that alternates between myxoid and more prominent collagenous areas. In contrast, MFS is predominantly myxoid in composition and exhibits greater cytological atypia compared with the cells found in low-grade fibromyxoid sarcoma.

The treatment approach for laryngeal masses depends on the histological type, location, and extent of the lesion. Benign masses are managed with conservative surgical excision. In some cases, adjunctive therapies such as voice therapy, injection laryngoplasty, and medical treatments, depending on the characteristics of the lesion, may also be employed to preserve or improve vocal function.[12] Malignant tumors, on the other hand, may require partial or total laryngectomy, radiotherapy, and chemotherapy. Early-stage tumors (stage I-II), particularly those located in the glottis or supraglottis, can often be effectively treated with single-modality therapies such as radiotherapy or conservative surgery, which allows for organ preservation. In contrast, advanced-stage tumors (stage III-IV) typically require multimodal approaches that combine surgery, radiotherapy, and chemotherapy. Among the histological subtypes, squamous cell carcinoma is the most common and well-studied, with established treatment protocols. However, rarer histologies, such as adenocarcinomas, lymphomas, neuroendocrine tumors, and sarcomas, including chondrosarcoma and MFS, often lack standardized treatment guidelines due to their rarity and are frequently managed on an individual basis.[1,13] Treatment planning must carefully balance oncologic control with the preservation of voice and airway function and should involve a multidisciplinary team including otolaryngologists, pathologists, medical oncologists, and radiation oncologists.[13]

The main treatment option for MFS is wide surgical resection. Radiotherapy is administered for recurrent, unresectable lesions or tumors with positive resection margins to suppress the risk of local recurrence and histological progression, especially for low-grade MFS. Chemotherapy in MFS remains an ongoing controversy.[14] Low-grade MFS is associated with a low level of malignancy and is infrequently linked to distant metastases, resulting in a favorable short-term prognosis. The 5-year survival rate for these tumors ranges from 60% to 70%.[15] Despite the lower malignancy, the rate of local recurrence for low-grade tumors is between 50-60%, which is similar to that of high-grade tumors. A progression was observed from low-grade to high-grade tumors, with low-grade regions frequently detected within high-grade tumors. This transition indicates a actantial histologic advancement from low- to high-grade tumors during recurrences, which may lead to a metastatic capability.[9] For this reason, the disease affects both men and women aged 50-70 almost equally and should be carefully monitored over a long period. In our case, a complete resection of the tumor was performed, and no recurrence or metastasis has been observed in the 54 months since the operation. Due to the rare cases, there is currently no definitive data on survival times. However, it can be stated that the prognosis for laryngeal sarcoma is slightly better than that for sarcomas located in other areas of the head

and neck.[16] In our case, the tumor was completely resected, and no recurrence or metastasis was observed during the 54-month follow-up period. Due to the rarity of laryngeal MFS, survival data remain limited. Nonetheless, laryngeal sarcomas may have a slightly better prognosis than sarcomas in other head and neck regions. This sunport contributes to the literature by presenting a rare tumor location with extended follow-up, marking the first documented case of this specific anatomical involvement and highlighting a long disease-free period.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Erdem Bayrakcı, Mehmet Akif Eryılmaz;* ***Design:*** *Erdem Bayrakcı, Mehmet Akif Eryılmaz;* ***Control/Supervision:*** *Mehmet Akif Eryılmaz, Sıdıka Fındık;* ***Data Collection and/or Processing:*** *Erdem Bayrakcı, Sıdıka Fındık;* ***Analysis and/or Interpretation:*** *Erdem Bayrakcı, Mehmet Akif Eryılmaz, Sıdıka Fındık;* ***Literature Review:*** *Erdem Bayrakcı;* ***Writing the Article:*** *Erdem Bayrakcı, Sıdıka Fındık;* ***Critical Review:*** *Erdem Bayrakcı, Mehmet Akif Eryılmaz, Sıdıka Fındık;* ***References and Fundings:*** *Erdem Bayrakcı, Sıdıka Fındık;* ***Materials:*** *Erdem Bayrakcı, Mehmet Akif Eryılmaz, Sıdıka Fındık.*

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