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Case Report

How rare is too rare to suspect? A case of a well-differentiated liposarcoma of the piriform recess of the larynx

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ABSTRACT

Liposarcomas are among the rarest soft tissue sarcomas in the larynx, accounting for less than 1% of cases with the piriform recess of the larynx being one of the least common sites. These tumors present significant diagnostic delays due to their comparison to benign lipoma lesions. The aim is to emphasize the diagnostic utility of molecular markers such as murine double minute 2 (MDM2) and cyclin-dependent kinase 4 (CDK4), and the importance of maintaining long-term surveillance in patients with atypical adipocytic tumors in the head and neck region. A 56-year-old female with a prior history of hypopharyngeal lipoma presented with progressive dysphagia, hoarseness, and inspiratory stridor. Flexible endoscopic examination revealed a well-circumscribed mass originating from the right piriform recess. Magnetic resonance imaging (MRI) was performed. Initial transoral excision confirmed well-differentiated liposarcoma (WDL) via histopathological analysis and positive MDM2 and CDK4 immunochemistry, further supported by FISH. Following multidisciplinary evaluation in sarcoma tumor board, a second, wide-field excision with clear margins was performed using CO_2 transoral laser microsurgery (TOLMS). Postoperative recovery was complicated by dysphagia. At 24-month follow up, the patient remained clinically disease-free with preserved speech and swallowing functions following dedicated rehabilitative therapy. Involvement of the piriform recess, confirmed by MDM2 and CDK4 gene amplification, and the successful management through CO_2-TOLMS features both the diagnostic complexity and the therapeutic significance of prompt, molecular guided intervention in this rare clinical context. Complete surgical excision with negative margins remains the therapeutic mainstay. Long-term clinical surveillance is essential due to the risk of recurrence and the anatomic complexity of the laryngeal region.

Keywords: Well-differentiated liposarcoma, Piriform recess, Laryngeal neoplasms, MDM2 amplification, *CDK4*, Transoral laser microsurgery, Head and neck sarcoma, Histopathology

INTRODUCTION

Liposarcomas are among the rarest soft tissue sarcomas in larynx, accounting for less than 1% of cases.[1]

Within this subset, laryngeal involvement, particularly the piriform recess, is exceptionally rare and represents significant diagnostic challenges due to its nonspecific clinical presentation.[2]

This case involves a well-differentiated liposarcoma (WDL), a subtype characterized by the amplification of the murine double minute 2 (MDM2) and cyclin-dependent kinase 4 (CDK4) genes.[3]

The subtlety of symptoms such as mild dysphagia or hoarseness of voice often contributes to diagnostic delays.[4] The aim is to emphasize the diagnostic utility of molecular markers such as MDM2 and CDK4, and the importance of maintaining long-term surveillance in

patients with atypical adipocytic tumors in the head and neck region.

CASE REPORT

Subject

A 56 year old female with a prior medical history of a hypopharyngeal lipoma resection presented to our clinic with progressive dysphagia, hoarseness of voice, and inspiratory stridor. No routine imaging follow-up had been conducted since her initial benign diagnosis eight years earlier. A flexible endoscopic examination showed a large, smooth, exophytic hypopharyngeal mass arising from the right piriform recess, visible at the supraglottic inlet and partially obstructing the airway. Hypomobility of the right vocal cord was observed (Figure 1).

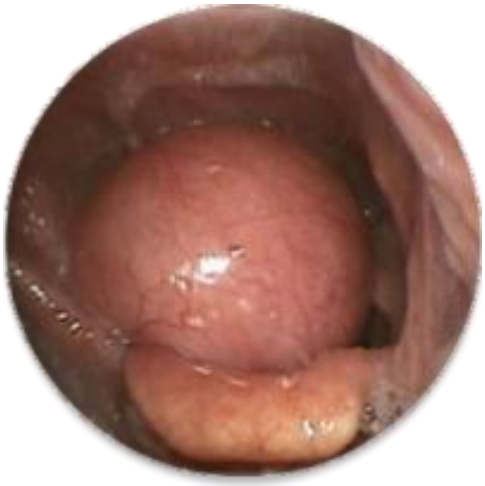

Figure 1: Flexible endoscopic examination. Exophytic mass in the right piriform recess of the larynx.

Radiological investigation

Magnetic resonance imaging (MRI) of the neck was conducted and showed a well-defined, lobulated soft tissue mass measuring 2.9×2.2×4.8 cm, centered in the right piriform recess of the larynx (Figure 2).

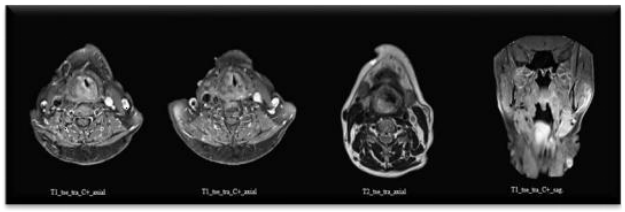

Figure 2: Radiological evaluation, pre-operative MRI neck.

The lesion appeared isointense on T1-weighted sequences and showed mild post-contrast enhancement. There was no evidence of cartilage invasion, nodal involvement, or extralaryngeal extension.

Operation and intraoperative findings

The patient underwent transoral CO_2 laser microsurgery (TOLMS). Complete excision was achieved, with careful preservation of critical structures, including the aryepiglottic fold, to maintain well postoperative swallowing function.

Microscopic examination

Histology confirmed the diagnosis of a WDL of the larynx, classified according to the WHO 2020 classification of liposarcoma subtypes. Histopathological evaluation of a soft tissue tumor using hematoxylin and eosin (H and E) staining revealed the presence of atypical cells, including multinucleated forms (Figure 3).

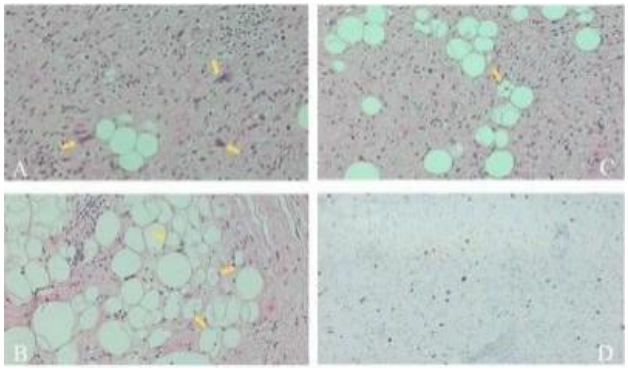

Figure 3: (A) Presence of atypical sometimes multinuclear forms (arrows) H and E stain; (B) area with lipoblast (arrows) H and E stain; (C) lipoblast (arrow) H and E stain; (D) MDM2 positivity by immunohistochemistry.

Lipoblasts were identified, both as isolated cells and in clusters, indicative of adipocytic differentiation. Immunohistochemical analysis demonstrated strong nuclear positivity for MDM2, supporting the diagnosis of a WDL liposarcoma. Fluorescence *in situ* hybridization (FISH) confirmed gene amplification of MDM2 and CDK4, molecular alterations commonly associated with Liposarcoma subtypes. These findings collectively support the diagnosis of a liposarcoma, with molecular and histological features consistent with a malignant adipocytic neoplasm. In this case amplification served as a definitive diagnostic confirmation (Figure 4).

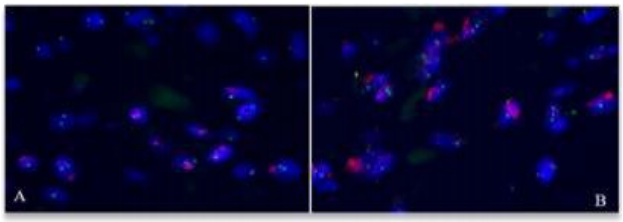

Figure 4 (A and B): (A) MDM2 gene amplification by FISH (B) CDK4 gene amplification by FISH.

Multidisciplinary sarcoma board*:* Following discussion in a multidisciplinary sarcoma tumor board, a second-look TOLMS procedure was performed to ensure complete resection with negative margins. Target biopsies confirmed absence of residual disease.[5]

Follow-up

Postoperative recovery was complicated by dysphagia, requiring nasogastric feeding for two months, and transient dysphonia. The patient underwent rehabilitative speech and swallowing therapy. At the 2-months follow-up, a fiberoptic endoscopic evaluation of swallowing (FEES) confirmed restoration of safe swallowing. Postoperative MRI of the neck showed no residual or recurrent mass, with only minimal airway distortion and no nodal pathology. Serial MRI-imaging of the neck at 3, 6, 12, and 24 months demonstrated sustained surgical success, anatomical stability, and no evidence of disease recurrence (Figure 5).

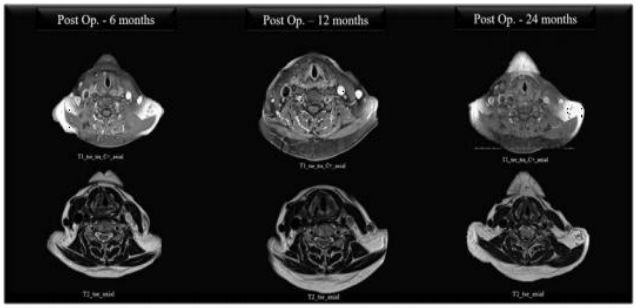

Figure 5: Serial post-operative MRI's at 6 months, 12 months and 24 months.

DISCUSSION

The WDL of the larynx and especially in the piriform recess is exceptionally rare, often presenting with non-specific clinical presentation.[1] Clinical awareness is critical, particularly in patients with slowly growing adipocytic lesions in atypical locations of the upper aerodigestive tract.

Molecular analysis including gene amplification of MDM2 and CDK4 detected by FISH is essential to differentiated WDL from lipomas.[4] An accurate molecular diagnosis directly informs surgical planning and prevents under treatment.

Given the risk of local recurrence, long-term clinical and radiologic follow-up remains the mainstay of management. In selected cases, especially those with localized residual disease, second-look transoral CO_2 laser microsurgery can be curative while preserving function.[5]

CONCLUSION

Involvement of the piriform recess, confirmed by MDM2 and CDK4 gene amplification, and the successful management through CO_2 TOLMS features both the diagnostic complexity and the therapeutic significance of prompt, molecular guided intervention in this rare clinical context.

Clinicians and pathologists should maintain a high index of suspicion when evaluating submucosal laryngeal masses, and integrate molecular diagnostics early in the workup to enable timely and definitive intervention.

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