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# A rare case of a laryngeal leiomyosarcoma with a lymph node metastasis

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## ABSTRACT

**INTRODUCTION:** Leiomyosarcoma is a rare mesenchymal tumor that originates from smooth muscle cells. Head and neck LMSs represent only 3% of all leiomyosarcomas with less than 50 cases of laryngeal LMS reported in the literature till now.

**CASE PRESENTATION:** We report a case of 50-year-old male presented at our ENT department for a chronic hoarseness. Clinical examination investigations found small submucosal lesion in the right vocal cord. Treatment consisted of CO<sub>2</sub> Laser excision of the lesion. The evolution was marked by the appearance of a tumefaction in the left submandibular region and a severe dyspnea requiring an emergency tracheotomy. Paraclinical examination investigations found a supraglottis–glottis–subglottis tumor. A total laryngectomy with bilateral functional neck dissection was performed and the histopathological examination found a laryngeal leiomyosarcoma.

**CONCLUSION:** LMS of the larynx a very rare malignancy. The accurate diagnosis is histological. Surgery is the mainstay of treatment. Its prognosis is correlated to local recurrence and distant metastases.

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## 1. Introduction

Leiomyosarcoma (LMS) is a rare mesenchymal tumor that originates from smooth muscle cells. It accounts for 5–10% of all soft tissue sarcomas.

Its laryngeal location was described for the first time by Jackson in 1939 [1] and since then less than 50 cases have been reported in the literature [2,3].

Through this case and a literature review, we discuss epidemiological, clinical, radiological and therapeutic features of this rare entity. This work is reported by following the surgical case report (SCARE) guidelines [4,5].

## 2. Case report

A 50-year-old male presented at our ENT department for a chronic hoarseness evolving for the past one year with no associated dyspnea or dysphagia.

The medical history found chronic smoking (16 pack/year) and occasional alcoholism weaned 20 years, no history of radiation exposure or previous surgery, no pharmacological allergies, no psychosocial problems, no family genetic disease, and specially no history or family history of tumor.

Nasofibroscopy showed a small, smooth submucosal lesion in the right vocal cord without impaired mobility of the latter.

Direct laryngoscopy under general anesthesia was realized by an ENT professor and showed the same lesion in the right vocal cord. Because of these results a CO<sub>2</sub> Laser excision of the lesion was performed.

The histopathology of the lesion demonstrated an Angioleiomyoma with (CD34+/Ck-/ Ki67 < 1%) on Immunohistochemistry (IHC).

Post-operatively the patient was lost to follow-up. 14 months later, the patient presented with a severe dyspnea requiring an emergency tracheotomy (Fig. 1).

The clinical examination found a patient in a fairly good general state, the larynx structures were normal and a painless mass located in the left submandibular. It was slightly fixed to deep structures, measuring 4 × 3.5 cm (Fig. 1).

Neck CT scan highlighted a tissue process of the supraglottis–glottis–subglottis areas completely obstructing the larynx, with the presence of left jugulo-carotid adenopathy (Fig. 2).

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Fig. 1. Clinical aspect of the left angulomandibular mass.

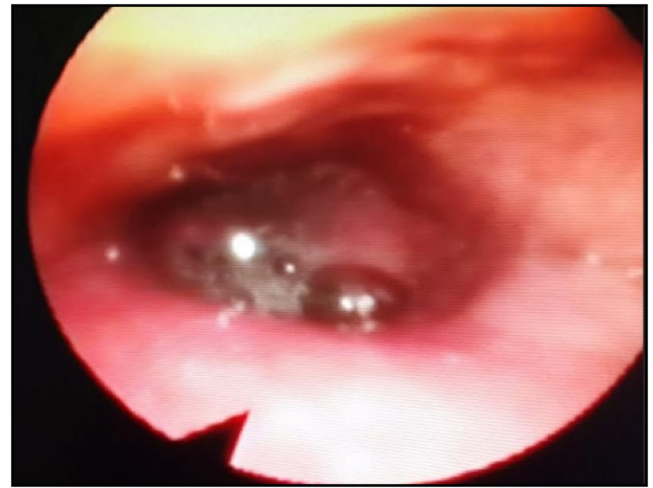


Fig. 3. Nasofiberscopy showing an infiltrative and totally obstructive laryngeal mass with salivary stasis.

A second Direct laryngoscopy under GA revealed a whitish budding supraglottis–glottis tumor that was completely obstructing the laryngeal lumen with a significant salivary stasis (Fig. 3).

Biopsy and histopathological examination of the specimen have confirmed a laryngeal leiomyosarcoma (Fig. 4).

Clinical and radiological work-up has not found distant metastases.

A multidisciplinary ORL-Oncology meeting has been held where a total laryngectomy with extemporaneous examination of the adenopathy was decided (Fig. 5), followed by radiotherapy.

The extemporaneous examination was consistent of a sarcoma metastasis, given these results, total laryngectomy was completed by bilateral functional neck dissection performed by an ENT professor.

The patient adhered well to the treatment received with a good tolerance to the surgery and post-operative care.

The postoperative period was unremarkable. Adjuvant radiotherapy was considered. After 2 years of follow-up, no further recurrence was noted.

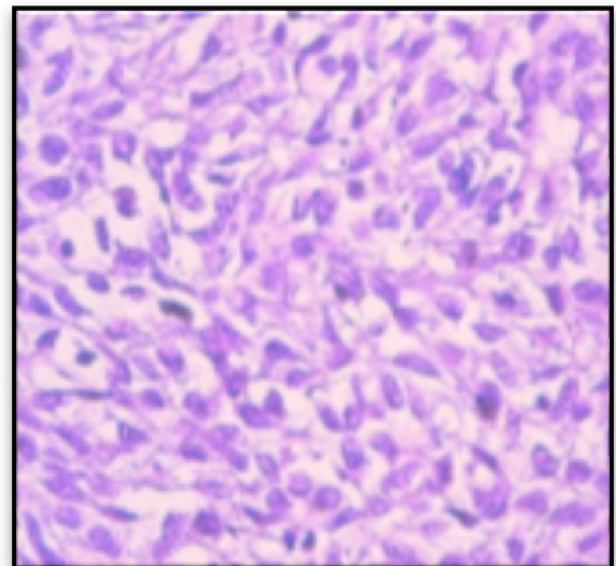


Fig. 4. Histological picture showing the appearance of a leiomyosarcoma with fusiform cells and high mitotic index.

3. Discussion

LMS of the larynx is a very rare malignancy that originates from blood vessel, smooth muscle or from the heterotopic mes-

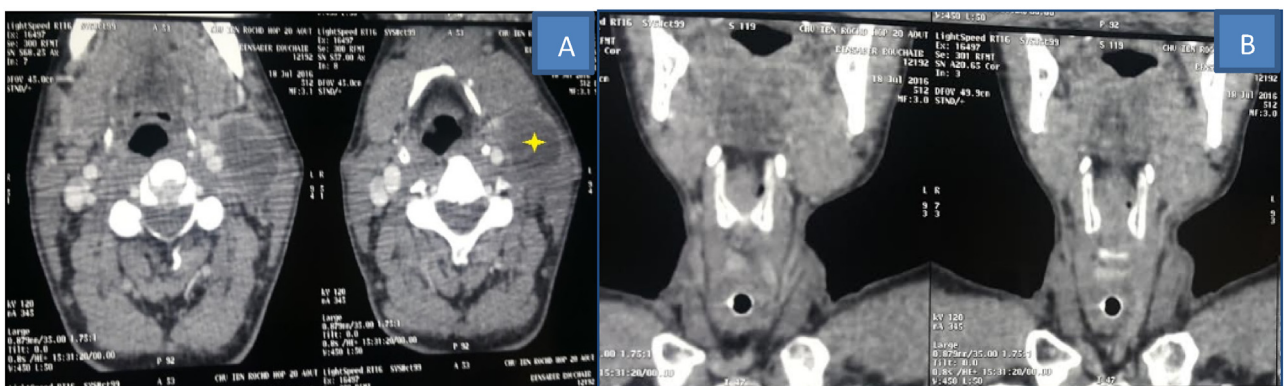
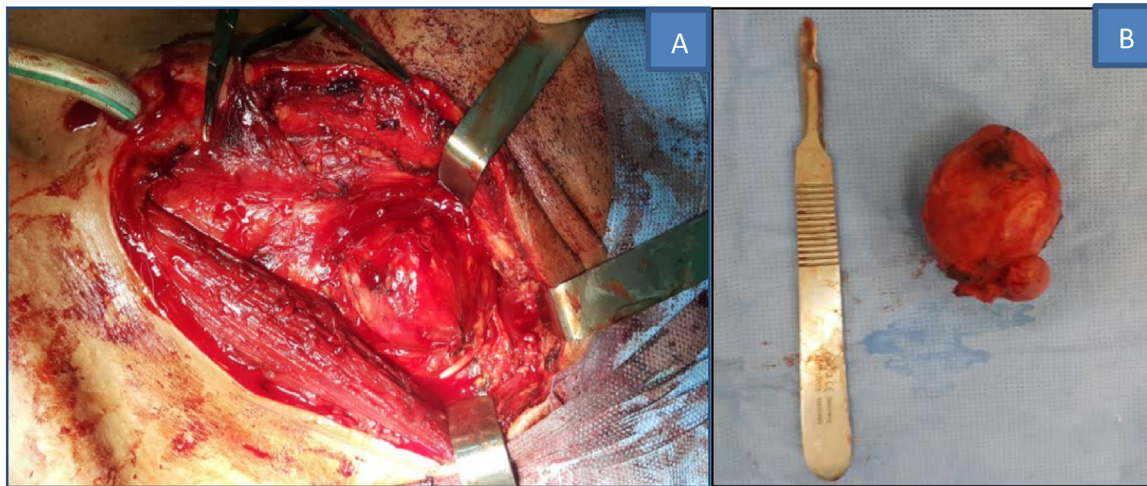


Fig. 2. Cervical CT scan in axial (A) and coronal (B) section with contrast injection showing a totally obstructive laryngeal tumor with left jugular–carotid adenopathy (star).



**Fig. 5.** Intraoperative aspect of the subangulomandibular metastatic lymph node (A) and the resection specimen (B).

enchymal tissue in the larynx [5]. LMSs account for 10% of head and neck sarcomas [6,7]. Head and neck LMSs represent only 3% of all leiomyosarcomas [8]. Head and neck LMSs occur most often in paranasal sinuses, scalp and cervical esophagus [7,5]. Smoking and alcohol are predisposing factors in the genesis of laryngeal squamous cell carcinoma, but their role in the genesis of leiomyosarcoma is still uncertain [9]. It occurs in adult between 30–80 years with maximum frequency during the 5th decade, preferably in male gender with a male/female ratio of 4:1 [10]. It may be a result of a malignant transformation of laryngeal angioleiomyoma which is highly unlikely [11]. Only two reports about malignant transformation of the benign angioleiomyoma in the literature have been reported so far, separately in the index finger and in the forearm [12].

Clinically, laryngeal LMS cannot be distinguished from other laryngeal malignancies, with non-specific symptoms, however, it is more obstructive which may require an emergency laryngectomy or tracheostomy [13]. The reported duration of the symptoms may vary from weeks to years [14].

Laryngeal LMS originates mostly in the supraglottic or glottic area. In a previous study of 31 patients, its presence in the larynx was reported as glottis (48%), supraglottis (32%), and supraglottis-glottis (6.5%) [15]. In the current case, we found a budding supraglottis-glottis tumor that was causing a complete airway obstruction.

Head and neck LMS are poorly lymphophilic and rarely presented by lymph node metastasis. Patients with this condition do not usually have palpable lymphadenopathy at the time of diagnosis [16]. Thus, the case of our patient represents an exceptional case to report for the first time.

CT and MR imaging evaluate the local extent and the size of the tumor as well as the lymph node status and the vascular component of the lesion [10]. Immunohistochemistry and ultrastructural findings provide the accurate diagnosis. LMSs are usually positive for alpha smooth muscle actin and negative for cytokeratins and epithelial membrane antigens.

Given the scarcity of cases reported of LMS in the literature, no therapeutic protocol has been established yet. Treatment modalities depends on size, location, extension of the tumor beside the age and the comorbidities that may have the patient. It is mainly surgical, thus only large radical resection by total laryngectomy with tumor free-margins provide a long survival [2]. The rate of local recurrence following surgical excision ranges from 35 to 50% [17]. Radiation therapy is less effective as a mainly therapeutic modality but still considered as a therapeutic adjunct to surgery. It has also a

role in recurrence and in residual disease. Chemotherapy also has a limited role [7]. After a review of the literature, the 5-year survival is estimated at 40% in the absence of metastasis. A long period of follow up is mandatory since the risk of recurrence persist long time after treatment [17].

#### 4. Conclusion

LMS of the larynx a very rare malignancy that can clinically mimic laryngeal carcinomas. The accurate diagnosis is mainly histological. The treatment is based on surgery and radiotherapy. The prognosis is essentially conditioned by the quality of surgical excision and correlated with the occurrence of local recurrence and the presence of metastases.

#### Declaration of Competing Interest

The authors report no declarations of interest.

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#### Ethical approval

I certify that this kind of manuscript does not require ethical approval by the Ethical Committee of our institution.

#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Author contribution

K. Salama: conception and design of the study.  
 B. Merzouki: conception and design of the study.  
 O. Berrada: acquisition of data.  
 Y. Oukessou: drafting the article.  
 S. Rouadi: drafting the article.  
 R. Abada: revising the article.  
 M. Roubal: revising the article.

M. Mahtar: final approval of the version to be submitted.

#### Registration of research studies

Not applicable.

#### Guarantor

Khadija Salama.

#### Provenance and peer review

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#### References

- [1] C. Jackson, C.L. Jackson, Sarcoma of the larynx, in: C. Jackson (Ed.), *Cancer of the Larynx*, W. B. Saunders, Philadelphia, 1939, pp. 164–168.
- [2] M. Soufi, R. Mohsine, Leiomyosarcome primitif de la surrenale, *J. Afr. Cancer* 1 (2009) 168–171.
- [3] Akbar Abbas, Mubasher Ikram, Nausheen yaqoob, Leiomyosarcoma of the larynx: a case report, *Ear Nose Throat J.* 84 (2005) 435–436.
- [4] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus Surgical Case Report (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230.
- [5] Ömer Tarık Selçuk, Levent Renda, Bekir Erol, Üstün Osma, Hülya Eyigor, Hülya Öztürk, A case of laryngeal leiomyosarcoma and review of the literature, *Ann. Maxillofac. Surg.* 5 (July–December (2)) (2015) 274–276, <http://dx.doi.org/10.4103/2231-0746.175772>.
- [6] A.M. Freedman, H.M. Reiman, J.E. Woods, Soft tissue sarcomas of the head and neck, *Am. J. Surg.* 158 (1989) 367–372.
- [7] E. Khadivi, M.H. Taziky, A.H. Jafarian, M. Nasser Sadr, Laryngeal leiomyosarcoma, a case report and review of articles, *Iran. J. Otorhinolaryngol.* 25 (2013) 253–258 [PMC free article] [PubMed].
- [8] A.k. Wadhwa, H. Gallivan, B. J. O'Hara, et al., Leiomyosarcoma of the larynx: diagnosis aided by advances in immunohistochemical staining, *Ear Nose Throat J.* 79 (2000) 42–46.
- [9] Ehsan Khadivi, Mohammad Hossein Taziky, Amir Hossein Jafarian, Msoud Nasser Sadr, Laryngeal leiomyosarcoma, a case report and review of articles, *Iran. J. Otorhinolaryngol.* 25 (4) (2013), Serial No. 73, Oct.
- [10] E. Morera Serna, C.A. Pérez Fernández, C. de la Fuente Jambrina, J. Razquin Muñoz, M.A. Pérez Gil, Laryngeal leiomyosarcoma, *Acta Otorrinolaringol. Esp.* 58 (2007) 445–448 [PubMed].
- [11] C.P. Wang, Y.L. Chang, T.S. Sheen, Vascular leiomyoma of the head and neck, *Laryngoscope* 114 (2004) 661–665.
- [12] D.B. Herren, A. Zimmermann, U. Buchler, Vascular leiomyoma in an index finger undergoing malignant transformation, *J. Hand Surg. Br.* 20 (1995) 484–487.
- [13] K. Kainuma, M. Kikukawa, T. Itoh, M. Osawa, M. Watanabe, Leiomyosarcoma of the larynx: emergency tracheostomy, *J. Laryngol. Otol.* 115 (7) (2001) 570–572.
- [14] R.C. Helmberger, B.P. Croker, A.A. Mancuso, Leiomyosarcoma of the larynx presenting as a laryngopyocele, *AJNR Am. J. Neuroradiol.* 17 (1996) 1112–1114.
- [15] G. Marioni, G. Bertino, L. Mariuzzi, A.M. Bergamin-Bracale, M. Lombardo, C.A. Beltrami, Laryngeal leiomyosarcoma, *J. Laryngol. Otol.* 114 (2000) 398–401.
- [16] R.S. Mindell, T.C. Calcaterra, P. H. Ward, Leiomyosarcoma of the head and neck: a review of the literature and report of two cases, *Laryngoscope* 85 (1975) 904–910.
- [17] Ch.E. Skoulakis, P. Stavroulaki, P. Moschotzopoulos, M. Paxinos, A. Fericean, D.E. Valagiannis, Laryngeal leiomyosarcoma: a case report and review of the literature, *Eur. Arch. Otorhinolaryngol.* 263 (10) (2006) 929–934.

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