# Schwannoma subglótico: Uma causa rara de dispneia

# Subglottic Schwannoma: A rare cause of dyspnea

Joana Gonçalves • Jorge Miguéis • António J. Ferreira • Nuno Silva • Clara Silva • Ana Amorim • Luís Silva

RESUMO

Uma jovem adulta foi referenciada à consulta de pneumologia por um quadro de tosse persistente, disfonia e dispneia progressiva com 6 meses de evolução. As provas de função respiratória revelaram obstrução e aumento da resistência das vias aéreas, sem resposta à broncodilatação. A TC de alta resolução demonstrou uma lesão infraglótica lobulada, lateralizada à direita, condicionando uma redução do lúmen aéreo em 50 a 75%. O estudo dirigido por video-broncofibroscopia confirmou a presença de duas lesões infraglóticas vascularizadas, efetuando-se biópsia. A excisão cirúrgica completa foi realizada por uma abordagem combinada (via endolaríngea e via cervicotomia medial). O diagnóstico histopatológico revelou tratar-se de um schwannoma laríngeo. Aos 6 meses após a cirurgia, a doente apresentava-se assintomática e, na visualização por laringoscopia indireta, a mobilidade das cordas vocais estava preservada e sem evidência de recorrência tumoral.

Palavras-chave: Subglótico, Schwannoma, Laringe

**Joana Gonçalves**
ENT Department, Centro Hospitalar e Universitário de Coimbra

**Jorge Miguéis**
ENT Department, Centro Hospitalar e Universitário de Coimbra

**António Jorge Ferreira**
Pneumology Department, Centro Hospitalar e Universitário de Coimbra

**Nuno Silva**
ENT Department, Centro Hospitalar e Universitário de Coimbra

**Clara Silva**
ENT Department, Centro Hospitalar e Universitário de Coimbra

**Ana Margarida Amorim**
ENT Department, Centro Hospitalar e Universitário de Coimbra

**Luís Silva**
ENT Department, Centro Hospitalar e Universitário de Coimbra

**Correspondência**
António Jorge Ferreira
11391@chuc.min-saude.pt
Praceta Prof. Mota Pinto, 3004-561 Coimbra

Artigo recebido a 29 de Dezembro de 2020. Aceite para publicação a 17 de Janeiro de 2021.

*ABSTRACT*

*A young woman with persistent cough, dysphonia, and progressive dyspnea for 6 months was referred to the pneumologist. Her pulmonary function tests revealed airway obstruction with increased airway resistance and hyperinflation, but with no response to bronchodilators. High resolution CT scan showed a right-sided, lobulated, infraglottic lesion, reducing by 50 to 75% the airway lumen. Video-bronchoscopy confirmed the presence of two vascularized infraglottic lesions and biopsy was performed. Complete surgical excision was achieved via a combined technique (endolaryngeal and medial cervicotomy approaches). The histopathological diagnosis was laryngeal schwannoma. At 6 months follow-up, cough and hoarseness had resolved, and indirect laryngoscopy showed normal vocal cord mobility without tumor recurrence.*

*Keywords: Subglottic, Schwannoma, Larynx*

## INTRODUCTION

Schwannomas are benign neural sheath tumors arising from Schwann's cells surrounding the peripheral nerves.[1,2] Although quite rare in absolute numbers, up to 45% of schwannomas occur in the head and neck region.[2] Schwannomas located in the larynx, especially in the subglottic region, are quite rare, with only a few cases reported in the literature.[3–5] Herein, we present a case of a young woman diagnosed with a laryngeal schwannoma presenting as subglottic mass.

## CASE REPORT

A 36-year-old woman, with an uneventful medical history, presented to the pneumologist with persistent cough, hoarseness and progressive dyspnea for 6 months. Breathlessness was exacerbated by exercise, laughing and crying. Physical examination and laboratory results were unremarkable. Her pulmonary function tests revealed airway obstruction with increased airway resistance and hyperinflation, but with no response to bronchodilators. High resolution CT scan showed a right-sided infraglottic lesion, growing between the inferior border of the cricoid cartilage and the first tracheal ring, measuring 29x16mm in axial plane and reducing by 50 to 75% the airway lumen (Fig.1).

The patient underwent video-bronchoscopy which confirmed the presence of two vascularized infraglottic lesions: one with sessile implantation on the right

**FIGURE 1**
CT of the neck region showing an infraglottic lobulated lesion in the right tracheal wall, with an endoluminal component, extending to the contralateral wall.

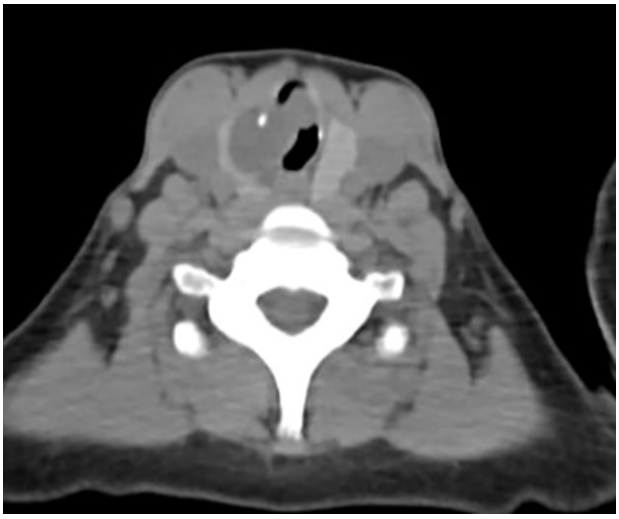
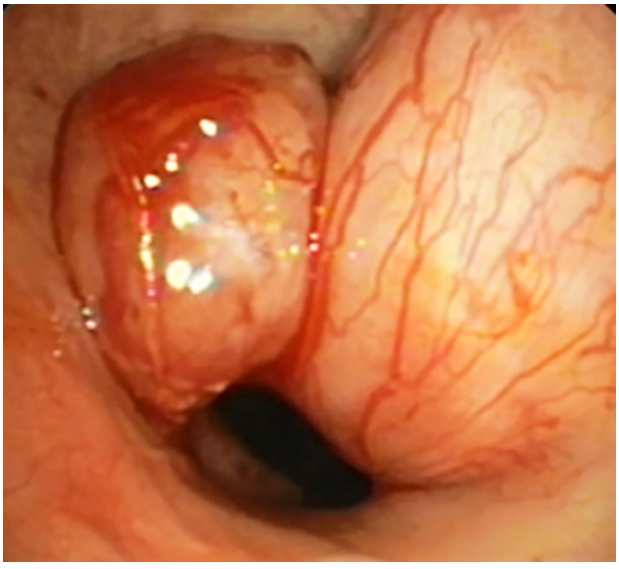


**FIGURE 2**
Video-bronchoscopy revealed two vascularized lesions reducing the tracheal lumen by 75%.



side, and a smaller one, pediculate and mobile, on the left side. (Fig.2). Biopsy was performed and histopathological examination suggested a probable neurofibroma. Positron emission tomography revealed a hypermetabolic lesion with a maximum SUV of 4.3.

Surgical treatment was performed using a combined technique, via endolaryngeal and medial cervicotomy approach. The pathologic lesion appeared has a white-yellow, fibroelastic mass with well-defined contours (Fig.3). Its extraluminal component ran in the right tracheoesophageal groove.

The tumor was completely excised, and the two combined fragments (endoluminal and extraluminal) measured about 3cm long (Fig 4.).

**FIGURE 3**
Medial cervicotomy – Extraluminal component

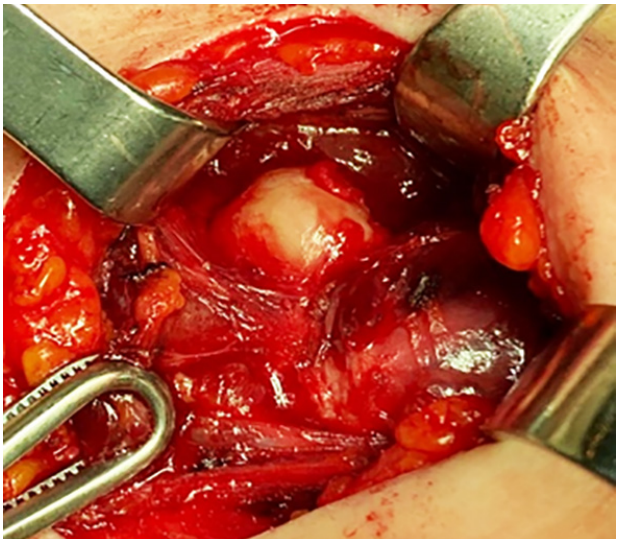
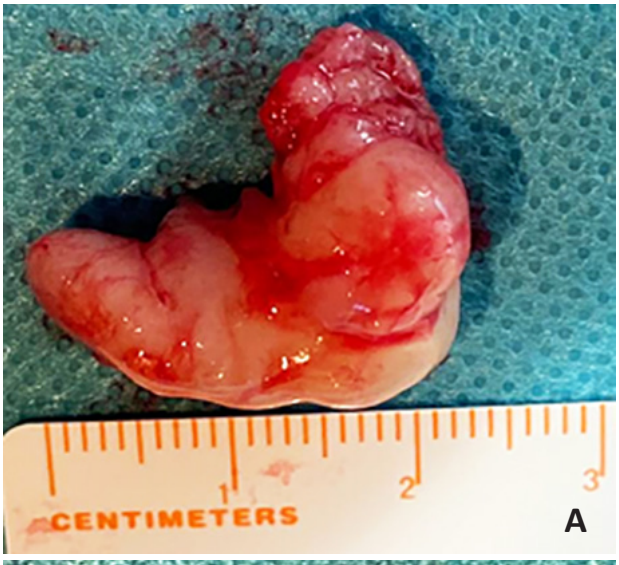


**FIGURE 4**
The resected tumor was a well-defined encapsulated lesion, with a 2cm extraluminal component (A) and a 1cm endoluminal component (B).



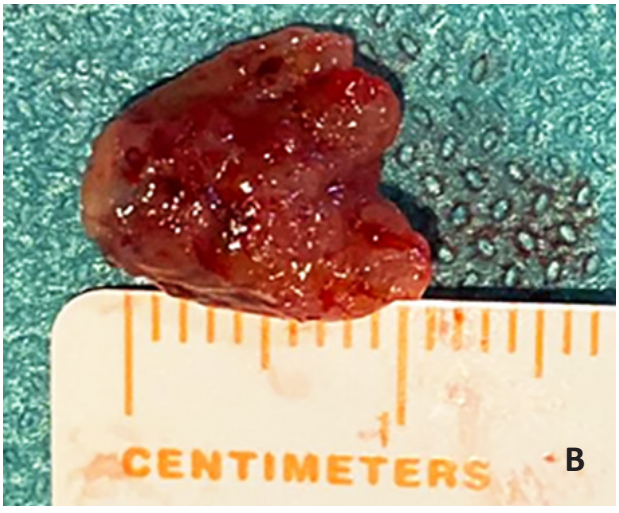
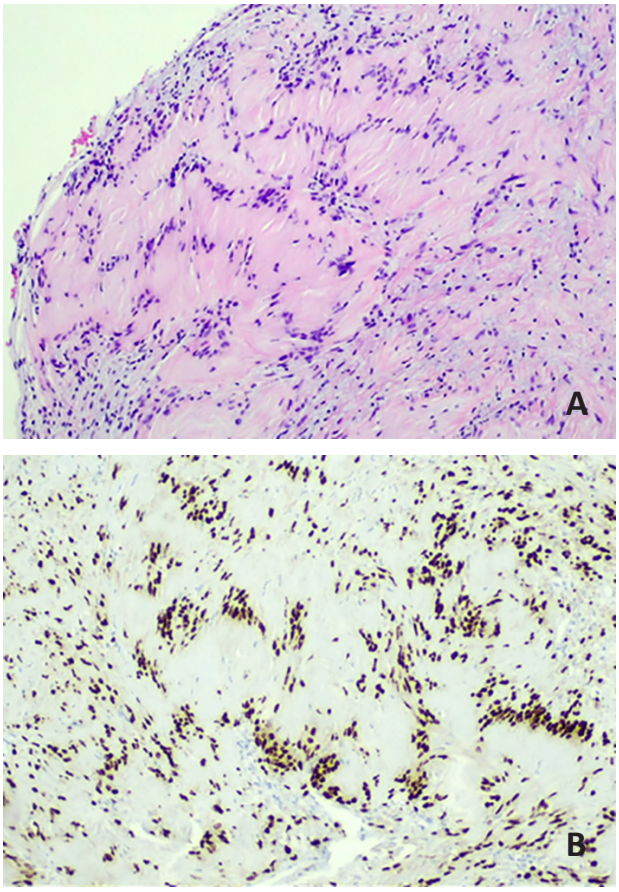



**FIGURE 5**
Histopathology: Hematoxylin and eosin stain (100x) – Verocay bodies in Antoni A areas (A) and immunopositivity for SOX10 (100x) (B)




Histological examination identified hypercellular Antoni A areas alternated with hypocellular Antoni B areas. Verocay bodies were present in the hypercellular component (Fig.5). Immunohistochemistry staining was positive for vimentin, pS100 and SOX-10. The pathological diagnosis was laryngeal schwannoma.
Postoperative recovery was straightforward, and patient was discharge on the fifth postoperative day, with eupneic breathing. At 6 months follow-up, cough and hoarseness had resolved, and indirect laryngoscopy showed normal vocal cord mobility without tumor recurrence.

## DISCUSSION

Laryngeal schwannoma is a very rare entity, representing between 0.1% and 1.5 % of all head and neck schwannomas.[3–5] These tumors may occur at all ages without sex preference.[4–6] Almost all laryngeal schwannomas arise from the aryepiglottic fold or arytenoids, with only three cases reported in the subglottic region.[3–5] Due to its slow growing nature, almost all laryngeal schwannomas present insidiously[6,7] Symptoms (e.g. cough, globus sensation, dysphonia and dyspnea) may arise from compression of neighboring structures, including the nerve fibers, or obstruction of the airway lumen.[4,5] Subglottic schwannoma may also mimic bronchial asthma, with increased breathlessness on laughing and crying. These symptoms occur because of the ball valve mechanism created by the tumor. Pulmonary function tests, however, will show an obstructive pattern without response to bronchodilators.[3] Imaging modalities, such CT and MRI, are often use to establish the size and extent of the mass, as well as to investigate its anatomical relations[8] CT scan usually show a well defined, hypodense mass without signs of cartilage destruction. Contrast enhancement often demonstrate central areas of low attenuation surrounded by a peripheral enhancing ring[7,9] On MRI, schwannomas typically show T2-weighted hyperintensity and strong enhancement following gadolinium injection.[7,9] However, CT and MRI features are not diagnostic. PET has limited value for identifying benign versus malignant peripheral nerve sheath tumors, as there are schwannomas with an elevated FDG uptake.[10] Therefore, only histopathology can confirm the diagnosis.[3,7,8] Schwannomas are encapsulated tumors, with alternating areas of compact spindle cells (Antoni A) and hypocellular less orderly areas (Antoni B). Verocay bodies are formed by alternating rows of palisading nuclei and intervening nuclei free stroma. A strong diffuse S-100 immunoreactivity is a defining feature.[3,7] Complete surgical resection is needed to avoid recurrence.[1] If feasible, an intracapsular enucleation with preservation of nerve fibers should be attempted. However, it´s often difficult to completely resect the tumor by peeling it from the nerve, because of its unclear borders.[1,8] Therefore, in most cases both the tumor and the adjacent nerve fibers are resected.[1,8] In the reported case, the patient had complete reversal of hoarseness without vocal cord paresis following surgery. This supports that tumor origin wasn´t the main trunk of the recurrent laryngeal nerve, rather the internal branch the superior laryngeal nerve.

## CONCLUSION

Laryngeal schwannoma is a rare entity, that can only be confirmed by histopathological examination. Nonetheless, CT and MRI are important pre-surgically to establish anatomical relations and outline surgical approach. PET CT is of limited value in differentiating benign versus malignant processes. Complete surgical excision remains the treatment of choice. Whenever possible, intracapsular enucleation should be attempted in the eventuality of the schwannoma originates in a motor branch of the laryngeal nerve.

**Acknowledgments**
We would like to thank Dr. Vânia Almeida, for providing the histopathological images and for her comments that greatly improved the manuscript.

**Conflicts of Interest**
The authors declare that they have no conflict of interests.

**Data Confidentiality**
The authors declare that data confidentiality was ensured.

**Animals and Peoples Protection**
All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation and with the Helsinki Declaration.

**Privacy policy, Informed consent, and ethical committee authorization**
Written consent to the submission of the case report was given by the patient.

**Funding**
The authors received no funding for this work.

**Data Availability**
There is no publicly available data of this article.

**References**
1. Ijichi K, Muto M, Masaki A, Murakami S. Recurrent plexiform schwannoma involving the carotid canal. Auris Nasus Larynx. 2018 Apr;45(2):358-361. doi: 10.1016/j.anl.2017.04.002.
2. Knulst R, Bosman W-M, Ritchie ED, da Costa A. Cystic schwannoma of the recurrent laryngeal nerve: a rare finding posing diagnostic difficulties. BMJ Case Rep. 2014 Apr 25;2014:bcr2014203873. doi: 10.1136/bcr-2014-203873.
3. Sofi FA, Mir MH, Bagdadi FS, Mehmood K. Hidden diagnosis in the subglottic larynx: Schwannoma mimicking as bronchial asthma. N Am J Med Sci. 2012 Jul;4(7):325-7. doi: 10.4103/1947-2714.98596.
4. Takeda K, Horiuchi M, Nakaya M, Yamaguchi K, Fujikawa A. Schwannoma of the trachea; a new resection technique. Auris Nasus Larynx. 2003 Dec;30(4):425-7. doi:10.1016/s0385-8146(03)00088-9.
5. Murata T, Shino M, Yasuoka Y, Chikamatsu K. Subglottic Schwannoma: A report of a rare case that was treated with medial thyrotomy. Am J Otolaryngol [Internet]. 2013 Sep [cited 2020 Jun 11];34(5):569–73. Available from: https://linkinghub.elsevier.com/retrieve/pii/S0196070913001075.
6. De Heer LM, Van Berkhout FT, Priesterbach-Ackley LP, Buijsrogge MP. Schwannoma of the Recurrent Laryngeal Nerve: A Rare Entity. Innovations (Phila). Jan/Feb 2017;12(1):64-66. doi: 10.1097/IMI.0000000000000334.
7. Wong BLK, Bathala S, Grant D. Laryngeal schwannoma: a systematic review. Eur Arch Otorhinolaryngol. 2017 Jan;274(1):25-34. doi: 10.1007/s00405-016-4013-6.
8. Xu X-Q, Hong T, Zheng C-J. Schwannoma originating from the recurrent laryngeal nerve in a thyroid cancer patient: A case report and review of the literature. World J Clin Cases. 2018 Dec 26;6(16):1202-5. doi: 10.12998/wjcc.v6.i16.1202.
9. Plantet MM, Hagay C, De Maulmont C, Mahe E, Banal A, Gentile A. et al. Laryngeal schwannomas. Eur J Radiol. 1995 Nov;21(1):61-6. doi: 10.1016/0720-048x(95)00655-a.
10. Beaulieu S, Rubin B, Djang D, Conrad E, Turcotte E, Eary JF. Positron Emission Tomography of Schwannomas: Emphasizing Its Potential in Preoperative Planning. AJR Am J Roentgenol. 2004 Apr;182(4):971-4. doi: 10.2214/ajr.182.4.1820971.