OTORINOLARINGOLOGIA

PUBBLICAZIONE PERIODICA TRIMESTRALE - POSTE ITALIANE S.P.A. - SPED. IN A. P. D.L. 353/2003 (CONV. IN L. 27/02/2004 N° 46) ART. 1, COMMA 1, DCB/CN - ISSN 0392-6621 TAXE PERÇUE

QUARTERLY JOURNAL ON
OTORHINOLARYNGOLOGY,
AUDIOLOGY
PHONATRICS, HEAD AND
NECK SURGERY
MAXILLO-FACIAL SURGERY
PLASTIC RECONSTRUCTIVE
SURGERY
OTONEUROSURGERY

VOL.69 No.4
DECEMBER 2019

EDIZIONI MINERVA MEDICA

Online version at http://www.minervamedica.it

Otorinolaringologia 2019 December;69(4):204-8
DOI: 10.23736/S0392-6621.19.02214-8

CASE REPORT

Multidisciplinary management of a giant and obstructive laryngeal myxoma

Massimo MESOLELLA [1] *, Gerardo PETRUZZI [1], Francesco A. SALZANO [2], Santina LAMARCA [1], Miriam CIERI [3], Daniela RUSSO [3], Maurizio IENGO [1], Massimo MASCOLO [3]

[1]Section of Ear Nose and Throat, Department of Neurosciences, Federico II University, Naples, Italy; [2]Department of Medicine and Surgery, Scuola Medica Salernitana University, Salerno, Italy; [3]Section of Pathology, Department of Advanced Biomedical Sciences, Federico II University, Naples, Italy

*Corresponding author: Massimo Mesolella, Section of Ear Nose and Throat, Department of Neurosciences, Federico II University, Naples, Italy. E-mail: massimo.mesolella@unina.it

ABSTRACT

Myxomas are benign lesions which can arise in any anatomic location but are mainly found in the heart. Exceptionally they interest the head and neck district, where the most frequently involved sites are the mandible and the maxilla. There are only few described cases of myxomas of the larynx. Here we report a new case of a laryngeal myxoma of the left true vocal fold, the second occurred in a female patient, presenting with severe dyspnea, dysphonia and dysphagia. We describe this case for two reasons. First is because it presents intubation which was safely treated through careful planning. A meticulous treatment planning by surgeon and anaesthetist was made mutually carried out prior to micro-laryngoscopic excision in case di large vocal cord polyps causing severe airway obstruction. The second reason is because a laryngeal myxoma is a very rare lesion. We provide a review of the literature, focusing on possible risk factors, and discuss the differential diagnosis and the best treatment options in laryngeal myxoma.

(*Cite this article as:* Mesolella M, Petruzzi G, Salzano FA, Lamarca S, Cieri M, Russo D, *et al*. Multidisciplinary management of a giant and obstructive laryngeal myxoma. Otorinolaringol 2019;69:204-8. DOI: 10.23736/S0392-6621.19.02214-8)

KEY WORDS: Myxoma; Larynx; Gastrointestinal stromal tumors; Airway management.

The definition of myxoma is applied to a group of benign mesenchymal neoplasms, which are mostly hypocellular with low-grade cellular atypia and abundant mucoid stroma composed of sulfated and non-sulfated glycosaminoglycans.[1] Indeed, the term "myxoma" was firstly introduced by Virchow in his first description of this kind of lesions, due to their resemblance to the gelatinous matrix of the umbilical cord.[2] Myxomas typically arise in the atria of the heart, but they have occasionally been described in the head and neck region, more often in mandible and maxilla, as well as in other sites (nasal cavity, parotid, and oral mucosa). Nevertheless, laryngeal myxomas are extremely rare, with a strikingly small number of cases described in literature. Since the report of Chen *et al.* in 1986, the first among those published and still available in PubMed, only 13 cases have been reported.[3-5] To date, yet little is known about the epidemiology and pathophysiology of this lesion, whose recognition might be arduous and troublesome for clinicians. In fact, its differential diagnosis includes some frequently encountered benign lesions, such as vocal cord polyps and ganglions, but also several malignancies, such as myxoid liposarcoma, myxoid leiomyosarcoma, myxoid chondrosarcoma, rhabdomyosarcoma, and other soft tissue tumors with myxoid degeneration.[6]

Herein we describe a new case of a vocal fold myxoma in a 59 years old woman. This case highlights a group of patients which will be very difficult to intubate. Infect the polyp covered more than 90% of glottis area. Above review the literature about the topic, discussing the main clinical and histological features of these rare benign lesions, to better define their diagnostic algorithm, facilitate their prompt recognition and avoid dangerous misdiagnosis.

Case report

A 59-year-old female was admitted to our Department of Neurosciences, Section of Ear Nose and Throat, complaining of severe dyspnea with approximately two years of duration, accompanied by increasing hoarseness with progressively worsening dysphonia and dysphagia, especially for solid foods. The patient had a 30-year smoking history (about 10 cigarettes a day).

Flexible fibro laryngoscopy revealed a large mass occupying the entire glottis region, arising from the anterior and medium third of the left true vocal cord, with minimal residual airway at posterior laryngeal inlet (Figure 1). She underwent thyroid ultrasound examination, thorax X-ray, ECG, and screening laboratory routine, which were all unremarkable. Subsequently, the patient was referred to surgery. Based on the discussion with the anesthetist and considering the size and location of the neoplasm, was used a video laryngoscope (Mc Grath, series 5, Aircraft Medical, Edinburgh, UK) with its handle mounted display and angulated blade design. Due to the concerns of a possible ventilation and intubation failure after the induction of anesthesia and as well as the risk to patient's life, a surgeon was on standby for potential emergencies while anesthesia was induced.

The patient's blood pressure (BP: 130/92 mmHg), electrocardiogram (ECG: normal) and oxygen saturation (SpO_2: 96%) were evaluated in the operating theater prior to the start of procedure and was monitored continuously during the procedures.

An attempt using a fiberoptic laryngoscope to guide a flexible endotracheal tube with an internal diameter of 4.0 mm to intubate through the narrow glottis space was successful. The whole procedure of intubation lasted about 3 minutes. The patient received mechanical ventilation and then underwent surgery.

During microlaryngoscopy, under general anesthesia, a microflap incision along the implantation base of the left true vocal fold mass was performed, allowing its removal. Intraoperatively, it appeared as a multinodular, rubbery to firm, greyish polypoid lesion measuring approximately 1.4 cm in diameter (Figure 2, 3, 4). Extubation was uneventful. Her breathing and voice immediately improved. Histology showed a proliferation of cytologically bland, stel-

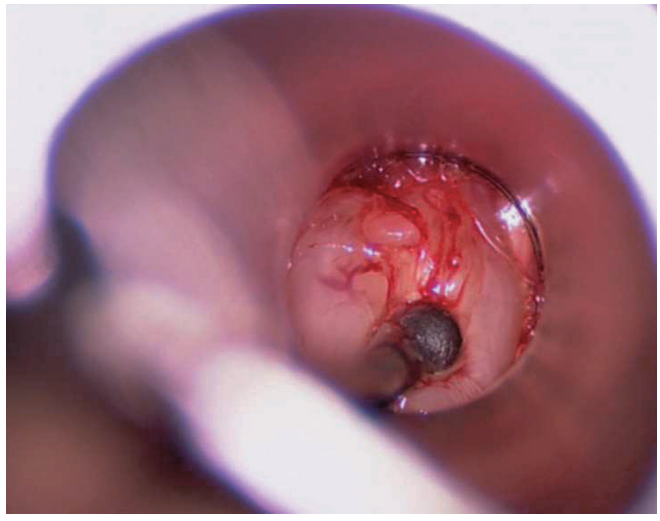

Figure 2.—Appearance of lesion during microdirect laryngoscopy.

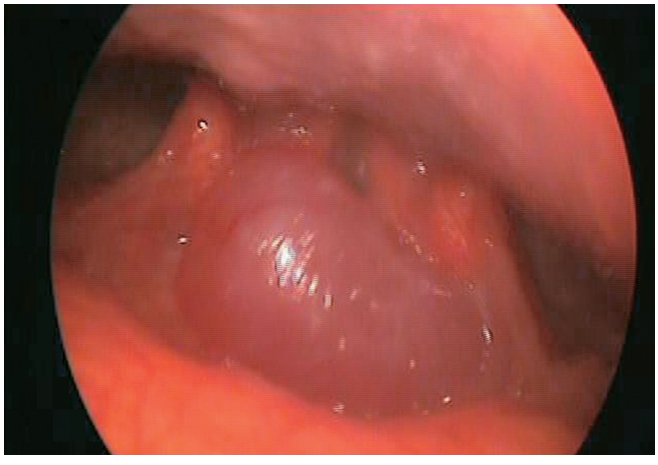

Figure 1.—The lesion of the glottic region, arising from the anterior third of the left true vocal fold.

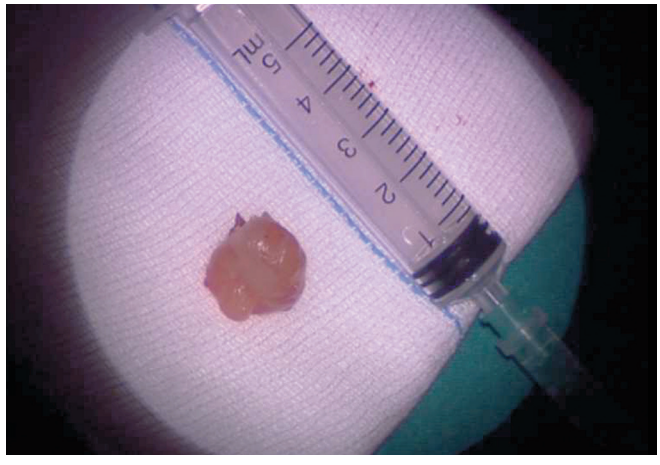

Figure 3.—Macroscopically, the greyish polypoid lesion of 1 cm in diameter.

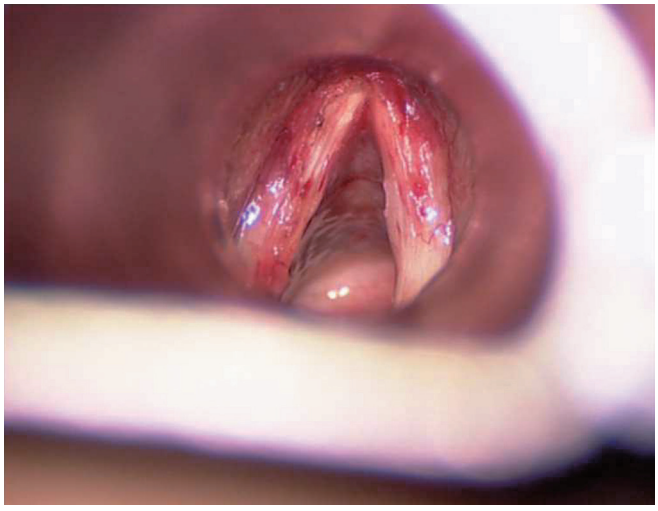

Figure 4.—At the end of the operation, laryngoscopy showed complete exeresis of the neoplasm.

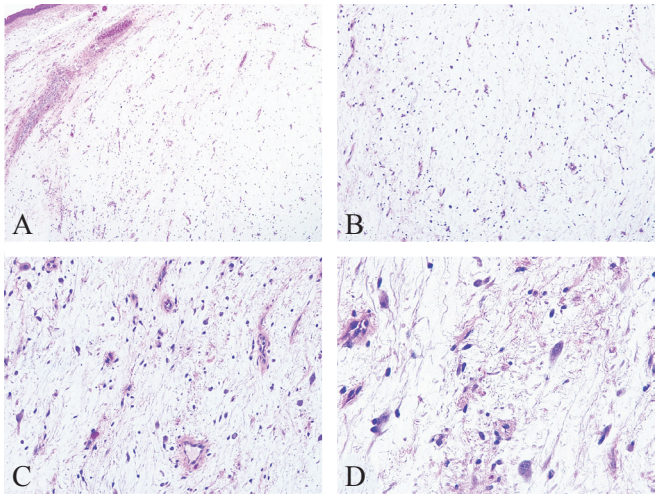

Figure 5.—Histological findings of the case: A, B) cytologically bland stellate proliferation cells within a loose myxoid stroma diffusely involving the corion (H&E staining; magnification 50× in A and 100× in B); C) absence of an evident vascular component (H&E staining; magnification 200×); D) neither cellular pleomorphism nor mitoses were seen (H&E staining; magnification 400×).

late cells within a loose myxoid stroma diffusely involving the corion, in absence of an evident vascular component (Figure 5). The overlying squamous epithelium appeared to be thickened with parakeratosis. Neither pleomorphism nor mitoses were seen. On immunohistochemical analysis, the cellular component was negative to all tested markers, including smooth muscle actin, desmin, protein S100 and CD34; the Ki-67 proliferation index was quite low. Based on clinical, instrumental, morphological and immunohistochemical features a diagnosis of vocal cord myxoma was carried out.

The patient was discharged the day after the surgical procedure. At the first follow-up visit, one month after surgery, flexible laryngoscopy showed complete remission with a good vocal outcome.

Discussion

Myxomas are rare benign tumors, commonly arising in the heart;[5] regarding the head and neck region the most frequently involved sites are maxilla and mandibula.[3] Laryngeal myxomas are very rare.[6-12] The most common location for a laryngeal myxoma is the vocal cord, followed by the vocal fold, aryepiglotic fold and the epiglottis.[13-16] Supraglottic myxomas tend to present later than those in vocal folds because they can grow larger before they become symptomatic, whereas vocal fold myxomas are detected earlier because they can cause disturbance of vocal fold vibrations.[9] Macroscopically they appear as grey-white, usually well circumscribed masses with a jelly cut surface; microscopically the gelatinous component predominates and constitutes the characteristic loose myxoid stroma in which a small number of stellate cells are dispersed in absence of atypia, necrosis or any vascular component.[1] Laryngeal myxomas are exceedingly rare, with only 13 cases described in literature to date, the current being the 14th. The main epidemiologic and clinical features of all reported cases retrieved by searching in PubMed for the key words “laryngeal” and “myxoma” are summarized in Table I.[6-18]

The prevalence of laryngeal myxomas is higher in the sixth decade (N.=5),[8, 9, 15, 17, 18] followed by the fifth decade (N.=4),[10, 12, 14] with only a minor number of cases widely distributed from the third to the seventh decade.[6, 7, 11, 13, 16] Conformingly, our patient was 59 years old at the time of the diagnosis. In consideration of the dramatic male predilection of laryngeal myxomas, our case stands out, representing the second report of occurrence in a female patient.[6, 15, 17]

Regarding tumor size, a strong correlation can be envisaged between tumor dimension and symptomatology. Tumors smaller than 1 cm in diameter mainly cause voice disorders (hoarseness and dysphonia), while masses of conspicuous dimensions provoke harsher symptoms (dyspnea and dysphagia), varying in relation to the involved anatomic sites (vocal cords, supraglottic and epiglottic region).

TABLE I.—*Main features of the reported cases.*[6-18]

Study	Age (years)/sex	Anatomic site (region)	Size (cm)	Symptoms	Smoking history
Chen[6]	38/M	Epiglottis	5.6×4.3×2.4	Dysphagia, dysphonia	NR
Sena[7]	70/M	Aryepiglottic fold	6.5×5.0×2.5	Hoarseness	Yes
Hadley[8]	64/M	Left vocal fold	1.0×0.6×0.2	Dysphonia	Yes
Tsunoda[10]	57/M	Right vocal fold	0.7×0.7×0.7	Hoarseness	Yes
Kim[9]	62/M	Right vocal fold	2.5×2.5×1.5	Dyspnea	Yes
Orliaguet[11]	42/M	Epiglottis, between vocal folds	NR	Dyspnea, snoring	NR
Baruah[12]	57/M	Aryepiglottic fold, epiglottis	6.5×5.0×1.0	Dysphagia, dysphonia	NR
Idrees[13]	46/M	Right vocal fold	0.8	Hoarseness	Yes
Leu[14]	53/M	Right vocal fold	0.5	Hoarseness	No
Nakamura[15]	64/F	Right vocal fold	0.4	Hoarseness	Yes
Ritchie[16]	77/M	Left vocal fold	0.8×0.3×0.2	Hoarseness	Yes
Tang[17]	60/M	Right vocal fold	1.0	Hoarseness	Yes
Williams[18]	60/M	Right vocal fold	NR	Dry cough, dysphonia, sensation of lump in the throat	No
Present case	59/F	Left vocal fold	1.4	Hoarseness, dysphonia, dysphagia	Yes

NR: not reported; IHC: immunohistochemistry.

Of notice, the great majority of patients, including ours, had a documented smoking history (10 of 12 patients, 83%; data not reported in two cases). Therefore, a relevant role for smoking in the pathogenesis of laryngeal myxomas could be proposed and should be studied and confirmed by further investigation.

The main difficulties in the diagnosis of these tumors lay in their rarity and in the existence of common pitfalls. Myxomas and vocal cords polyps are morphologically very similar, regarding dimension, color and presence of an implant base. Histologically the absence of a vascular component is an essential clue to differentiate a true laryngeal myxoma from myxoid modification of a vocal polyp.[6] Moreover, several malignant entities must be considered in the differential diagnosis of laryngeal myxomas, including myxoid liposarcoma, myxoid chondrosarcoma and angiolipoma.[18-22] In this scenario, immunohistochemistry is a very easily accessible and helpful tool. Myxoma cells are negative for desmin, actin and S100 protein; the demonstration of immunopositivity to one or more of these markers favors alternative diagnoses

Given the anatomical site involved, the infiltrative growth pattern and the recurrence propensity, misdiagnosis could be very dangerous for the patient. Consequently, their treatment gold standard consists in a wide and complete resection with free surgical margins. On the other hand, to obtain an optimal postoperative vocal outcome, during surgical procedure maximum effort should be put in preserving the vocal cord epithelium, the superficial lamina propria and the vocal ligament in the true vocal fold. Inadequate excision results in a high recurrence risk, while overtreatment with demolitive surgery may cause loss of vocal function. So, nowadays, the achievement of the best therapeutic results relies on avantgarde instrumental techniques, as video laryngoscopy, which allow has a double effectiveness in follow-up process, and in performing an excision as much as possible conservative, ensuring the absence of disease on surgical margins, to prevent its reappearance.[23, 24]

Treatment of laryngeal myxoma includes excision in microlaryngoscopy because of its encapsulation, although external incision may sometimes be required.[7] The recurrence rates of head and neck myxomas can be as high as 28% others have advocated for *en-bloc*, wide excision of these tumors to reduce the risk of recurrence.

There are a few key issues that are significant in this case report. Careful planning is essential in trying to intubate a patient with such a large glottis obstruction. In our Institute, in many complicated cases (such this reported), the patient has been seen before the day of surgery by an anesthetist. So, the adequate planning for procedure was made after discussions between the otorhinolaryngologists and a consultant anesthetist and only then preceded with attempting intubation. The use of a video laryngoscopes-assisted endotracheal intubation was essential in this case. We considered other methods, including fiberoptic bronchoscopy, but these methods were not necessary in this case. Endotracheal intubation can be impossible and even dangerous because it can cause complete airway obstruction, especially in patients with large glottic lesions, as it was in our case. In cases of difficult endotracheal intubation due to a severe obstruction, the ability to control the airway is crucial to the management. In general, awake fiber optic intubation should be selected for patients with a difficult airway. The safe way to anesthetize these patients is to maintain spontaneous respiration until the airway is secured.

Tracheostomy under local anesthesia is the safest way to manage patients with upper airway obstruction. However, she refused to accept elective tracheotomy.

Conclusions

Laryngeal myxomas are very rare lesions and, to the best of our knowledge, this is the second described case arisen in a female patient.[15] Although considered benign tumors, it is important to remind that the lack of a true capsule and the infiltrative growth pattern hinder their complete surgical removal. Furthermore, considering the anatomic sites involved, they could sporadically be unexpectedly threatening for patient life, by causing acute upper airway obstruction requiring emergency surgery, as tracheotomy, or, more often, can lead to the development of disabling and permanent surgery-related side effects, such as vocal hypofunctionality.[10]

Generally, a diagnosis of laryngeal myxoma should be performed by integrating clinical, imaging and morphological features; immunohistochemistry has a valuable role in the differential diagnosis.

Meticulous treatment planning by surgeon and anesthetist should be mutually carried out prior to microlaryngoscopic excision in cases of large vocal cord mass causing airway obstruction.

Hopefully, our review of the literature may contribute to better define the main characteristics of this quite uncommon lesion of the head and neck district.

References

1. Graadt van Roggen JF, Hogendoorn PC, Fletcher CD. Myxoid tumours of soft tissue. Histopathology 1999;35:291–312.

2. Virchow RL. Die Cellularpathologie in ihrer Begründung auf physiologische and pathologische Gewebelehre. Berlin: Verlag von August Hirschwald; 1871:593.

3. Singaraju S, Wanjari SP, Parwani RN. Odontogenic myxoma of the maxilla: A report of a rare case and review of the literature. J Oral Maxillofac Pathol 2010;14:19–23.

4. Shah A, Lone P, Latoo S, Ahmed I, Malik A, Hassan S, *et al.* Odontogenic myxoma of the maxilla: A report of a rare case and review on histogenetic and diagnostic concepts. Natl J Maxillofac Surg 2011;2:189–95.

5. Reynen K. Cardiac myxomas. N Engl J Med 1995;333:1610–7.

6. Chen KT, Ballecer RA. Laryngeal myxoma. Am J Otolaryngol 1986;7:58–9.

7. Sena T, Brady MS, Huvos AG, Spiro RH. Laryngeal myxoma. Arch Otolaryngol Head Neck Surg 1991;117:430–2.

8. Hadley J, Gardiner Q, Dilkes M, Boyle M. Myxoma of the larynx: a case report and a review of the literature. J Laryngol Otol 1994;108:811–2.

9. Kim KM, Kim SC, Jeong HJ, Kie JH. Myxoma: life-threatening benign nonepithelial tumor of the larynx. Yonsei Med J 1997;38:187–9.

10. Tsunoda K, Nosaka K, Housui M, Murano E, Ishikawa M, Imamura Y. A rare case of laryngeal myxoma. J Laryngol Otol 1997;111:271–3.

11. Orliaguet O, Pépin JL, Veale D, Kelkel E, Pinel N, Lévy P. Hunter's syndrome and associated sleep apnoea cured by CPAP and surgery. Eur Respir J 1999;13:1195–7.

12. Baruah P, Jha DN, Karak AK, Kumar R. Laryngeal myxoma. J Laryngol Otol 2001;115:231–2.

13. Idrees MT, Hessler R, Terris D, Mixson C, Wang BY. Unusual polypoid laryngeal myxoma. Mt Sinai J Med 2005;72:282–4.

14. Leu G, Klein AM, Deyrup AT, Johns MM 3rd. Pathology quiz case 1. Laryngeal myxoma. Arch Otolaryngol Head Neck Surg 2007;133:94–6, 96.

15. Nakamura A, Iguchi H, Kusuki M, Yamane H, Matsuda M, Osako S. Laryngeal myxoma. Acta Otolaryngol 2008;128:110–2.

16. Ritchie A, Youngerman J, Fantasia JE, Kahn LB, Cocker RS. Laryngeal myxoma: a case report and review of the literature. Head Neck Pathol 2014;8:204–8.

17. Tang CG, Monin DL, Puligandla B, Cruz RM. Glottic myxoma presenting as chronic dysphonia: a case report and review of the literature. Ear Nose Throat J 2015;94:E30–3.

18. Williams L, Allen C. A Submucosal True Vocal Fold Mass. JAMA Otolaryngol Head Neck Surg 2015;141:1025–6.

19. Testa D, Motta S, Marcuccio G, Paccone M, Rocca A, Ilardi G, *et al.* Our experience in the treatment of Malignant Fibrous Hystiocytoma of the larynx: clinical diagnosis, therapeutic approach and review of literature. Open Med (Wars) 2016;11:208–14.

20. Tiseo D, Tosone G, Conte MC, Scordino F, Mansueto G, Mesolella M, *et al.* Isolated laryngeal leishmaniasis in an immunocompetent patient: a case report. Infez Med 2008;16:233–5.

21. Mesolella M, Di Martino M, Laguardia M, Galera F, Galli V. Angiolipoma of the larynx. Otolaryngol Head Neck Surg 2007;136:142–3.

22. Mesolella M, Motta G, Galli V. Chondrosarcoma of the epiglottis: report of a case treated with CO2 laser epiglottectomy. Acta Otorhinolaryngol Belg 2004;58:73–8.

23. Mesolella M, Motta G, Laguardia M, Galli V. Papillomatosis of the larynx: treatment with CO2 laser. B-ENT 2006;2:51–4.

24. Motta S, Cesari U, Mesolella M, Motta G. Functional vocal results after CO2 laser endoscopic surgery for glottic tumours. J Laryngol Otol 2008;122:948–51.

Conflicts of interest.—The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

Manuscript accepted: April 8, 2019. - Manuscript revised: March 25, 2019. - Manuscript received: October 31, 2018.