CASE STUDY

Multiple Mucosal Neuromas in the Larynx as Part of a Multiple Endocrine Neoplasia Type 2B

Neuromas mucosos de localización laríngea en una neoplasia endocrina múltiple 2B

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Clinical Case

We present the case of a 28-year-old female diagnosed with multiple endocrine neoplasia type 2B (MEN 2B) when she was 17, consisting of medullary thyroid cancer, bilateral pheochromocytoma, lingual neuromas and marfanoid appearance. Bilateral adrenalectomy was performed as a first step, with complete thyroidectomy and bilateral ganglion dissection and resection of lingual neuromas later on. The patient is currently under treatment with levothyroxine, fludrocortisone acetate and hydrocortisone.

The women presented with clinical signs and symptoms with a history of 1 year, characterised by intermittent dysphonia that became constant and progressive in the last few months; patient denied having dyspnnea, cough and weight loss. Nasopharyngolaryngoscopy revealed left paramedian vocal fold paralysis, compensated by the contralateral fold, whose mobility is preserved, so adequate glottic closure was achieved. In addition, the patient presented multiple pale pink polypoid masses, smaller than 1 cm, without exudate or mucosal ulceration. These were found on both vocal cords as well as in the oesophageal vestibule (Fig. 1A).

We decided to perform direct laryngoscopy in the operating theatre to take a biopsy and resect the laryngeal masses. The biopsies were sent to pathology; microscope study revealed a proliferation of fusiform cells that formed a nodule typical of laryngeal neuroma under the stratified non-keratinized flat epithelium of the larynx (Fig. 2A and B). The patient had adequate postoperative clinical evolution and complete remission of the laryngeal neuromas was corroborated by laryngeal nasofibroscopy in the follow-up at 2 months and at 6 months (Fig. 1B).

Discussion

Multiple endocrine neoplasia type 2 (MEN 2) occurs in 1:200 000 live births; it is a neuroendocrine neoplasia with an autosomal dominant predisposition and has variable penetration.1

The neoplasia MEN 2B includes only 5% of all MEN 2 cases. However, its clinical course is the most aggressive of the MEN 2. It is a rare disease caused by mutations in the germline
Mucosal neuromas. Its exact incidence is not known. Tollemitt et al.\textsuperscript{9} presented mucosal neuromas in the glottis and supraglottis in a MEN 2B and Shimazaki et al.\textsuperscript{10} described single laryngeal neuromas in a 73-year-old patient without MEN 2B.

Mucosal laryngeal neuromas can show up as lesions that can potentially compromise the airway in patients with MEN 2B. Although they are apparently rare, it should be remembered that this location could be found in patients with this disease. Consequently, examination, treatment and follow-up in the presence of laryngeal neuromas make it possible to keep the airway permeable initially, while also suspecting MEN 2B if such a diagnosis does not exist already; this permits management of this treatable condition, minimising the course of the disease and the mortality rate.\textsuperscript{1,3}

Figure 1  (A) Image obtained through laryngeal nasofibroscopy in which you can see multiple pale pink polypoid masses, smaller than 1 cm, lacking exudate or mucosal ulceration, on both vocal folds. (B) Image obtained through laryngeal nasofibroscopy at 6 months’ follow-up, in which complete remission of the laryngeal neuromas can be seen.

Figure 2  (A) Microscope image in which, under the stratified non-keratinized flat epithelium of the larynx, a proliferation of fusiform cells that form a nodule can be seen. (B) Microscope image at 45× in which fusiform cells, stained with H&E characteristic of the presence of neuromas, can be seen.

in the RET proto-oncogene and codifies tyrosine for a kinase receptor that is expressed in neuroendocrine cells.\textsuperscript{2,3}

MEN 2B is characterised by medullary thyroid carcinoma (MTC), pheochromocytoma, mucosal neuromas, ganglioneuromatosis and marfanoid appearance.\textsuperscript{4} Mucosal neuromas generally develop on the tongue, lips, gastrointestinal tract and palate. Within these neoplasias, mucosal neuromas are the most typical clinical phenotype and represent the earliest clinical sign of MEN 2B.\textsuperscript{5} Other distinctive features of MEN 2B are the presence of an elongated face, thick and turned out eyelids and prominent lips.\textsuperscript{6}

Without surgical intervention, the mean age of survival for MEN 2B is 21 years.\textsuperscript{6} In 95% of the cases of MEN 2B, MTC develops at a relatively early age and, consequently, a more aggressive disease course is exhibited.\textsuperscript{7,8} In this case the diagnosis of MEN 2B and the accompanying MTC took place at the age of 17 years. For that reason, a complete thyroidectomy with bilateral ganglion dissection was performed 3 months after the diagnosis.

Even though mucosal neuromas commonly occur in MEN 2B, these lesions in the larynx are very rare, which is why this is the first case presented in the American continent with the presence of laryngeal neuromas. However, globally there have been a few cases presented in the United Kingdom, Germany and Japan.\textsuperscript{9}

There is only minimal documentation on mucosal laryngeal neuromas. This exact incidence is not known. Tollemitt et al.\textsuperscript{9} presented mucosal neuromas in the glottis and supraglottis in a MEN 2B and Shimazaki et al.\textsuperscript{10} described single laryngeal neuromas in a 73-year-old patient without MEN 2B.
Conflict of Interests

The authors have no conflicts of interests to declare.

References