■ CASE STUDIES

Laryngeal Paraganglioma: Diagnosis and Treatment. A Propos of a Case

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Paragangliomas of the larynx (LPG) are unusual neuroendocrine tumours that are seen as a vascular submucosal mass, with a female predilection. We add another case of LPG to the 77 previously reported in the literature. We present the clinical, radiological, and surgical features of a supraglottic LPG seen in a 40-year-old woman with an 8-month history of slowly progressive hoarseness. The tumour was removed approaching a lateral thyrotomy. The patient remains disease free 18 months after surgery. Preoperative CT, angiography, and embolization are useful in making the diagnosis and reducing perisurgical bleeding of LPG before surgical intervention. Complete excision through an external mucosa-sparing approach is the treatment of choice.

Key words: Laryngeal paraganglioma. Submucosal mass. Neuroendocrine tumours. Angiography. Embolization.

Paranglioma laríngeo, diagnóstico y tratamiento. A propósito de un caso

Los paragangliomas laríngeos (PGL) son tumores poco frecuentes, derivados del sistema neuroendocrino, que suelen iniciarse como una masa submucosa endolaríngea, con predilección por el sexo femenino. Aportamos un nuevo caso a los 77 publicados. Presentamos las características clínicas, radiológicas y quirúrgicas de un PGL supraglótico de una mujer de 40 años de edad, con clínica de 8 meses de evolución de ronquera. El tumor fue resecado mediante tirotomía lateral, previa embolización selectiva. La paciente permanece libre de enfermedad después de 18 meses tras la cirugía. La tomografía computarizada, la angiografía y la embolización preoperatorias son útiles en el diagnóstico y en la reducción del sangrado periquirúrgico. La exéresis completa vía externa con conservación mucosa es el tratamiento de elección.

Palabras clave: Paraganglioma laríngeo. Masas submucosas. Tumores neuroendocrinos. Angiografía. Embolización.

INTRODUCTION

LPG constitutes an unusual clinical condition with a mere 77 cases reported in the literature.[1] It appears predominantly in females and requires a differential diagnosis with other endolaryngeal submucosal masses, such as carcinoid tumours, small cell tumours, or medullary carcinoma of the thyroid[2-4] in order to establish prognosis.[2] We contribute a new case to the literature that was treated by our department.

CLINICAL REPORT

Forty-year old female with a history of childhood cerebral palsy due to encephalitis, epilepsy, and congenital cardiopathy. She presented with an 8-9 month history of having the feeling of a foreign body in the pharynx, odynophagia, hoarseness, and slight dyspnoea when placed in the supine position. Examination revealed a submucosal lesion in the right arytenoepiglottic fold that decreased the glottic lumen (Figure 1). A computerized tomography (CT) of the neck was ordered and was reported to depict a solid, homogenous supraglottic mass partially occupying the endolaryngeal lumen (Figure 1). A microlaryngoscopic exploration was performed under general anaesthesia revealing a soft tumouration; scant haematic material was extracted in the biopsy that was spread onto a Petri dish as an extemporaneous cytology and was negative for malignant cells. One week following surgery, the patient presented bleeding in the upper airway which remitted spontaneously, as well as increased dyspnoea that dictated the need for tracheostomy.

When faced with a vascular neck lesion and considering an external surgical approach, an arteriography was performed that revealed vascular tumouration dependent on the right superior thyroid artery, which was embolized (Figure 2). The lesion was excised externally by means of thyroidotomy, with opening of the right lobe of the thyroid in order to reach the endolarynx. The final pathology report determined that it was an LPG.

DISCUSSION

The finding of a submucosal supraglottic mass calls for a differential diagnosis of LPG among other malignant

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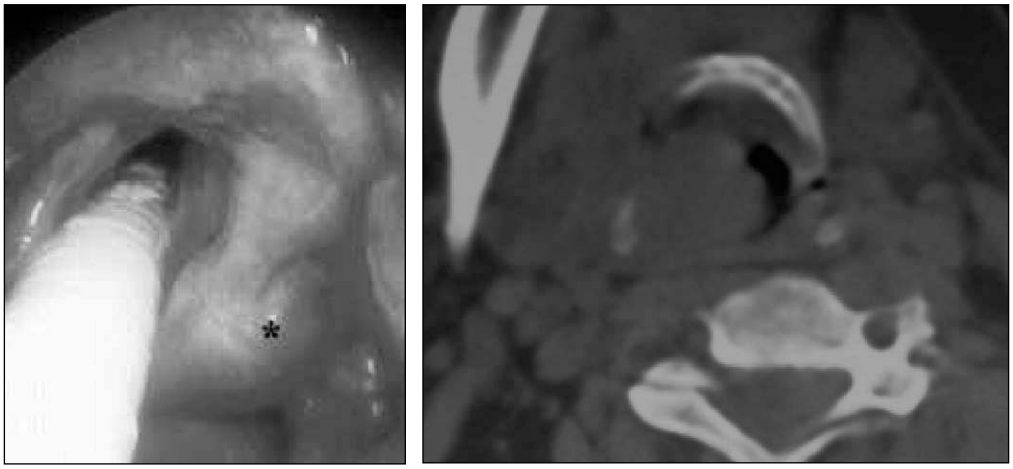

Figure 1. Image of the endolaryngeal examination in which a mass can be seen bulging in the right aryteno-epiglottic fold and a computerized tomographic image in which a space-occupying mass is seen at the right supraglottic level with a very small lateral lumen.

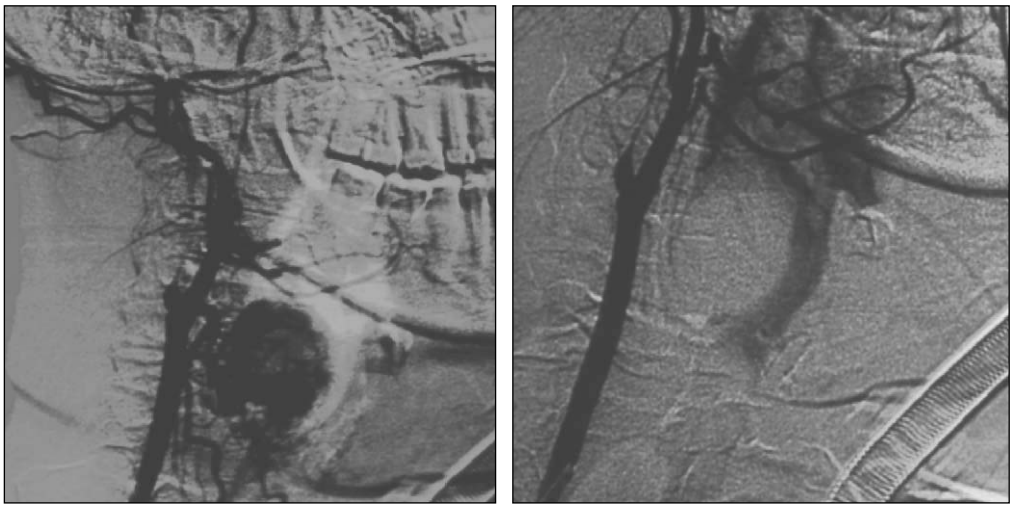

Figure 2. Arteriography showing vascular tumouration dependent on the superior thyroid artery and arteriography following embolization.

lesions,[2-4] using appropriate tests to visualize the lesions with the minimum risk possible for the patient as well as the maximum information for the physician. CT and magnetic resonance (MR) are the imaging techniques used to describe it.[2-4] As a complementary test procedure, fine needle aspiration biopsy (FNAB) is controversial, although some centres perform this procedure as a means of obtaining cytological material from deeper planes of the lesion.[5] Techniques involving radionuclides (indium or iodomethylbenzylguanidine tagged pentetreotide) are reserved for cases in which the diagnosis is doubtful, synchronic lesions, and monitoring the course of the lesion.[2] Despite the fact that they do not tend to present hormonal activity, their vascular characteristics, their mass effect and closeness to large vessels require proper pre-surgical management and preparation. Arteriography will not only define the extension of the tumour, but also the vascular involvement and the degree of collateral circulation.[4,5]

Biopsy has traditionally been contraindicated given the risk of haemorrhage,[1] although with present-day microsurgical techniques, it can be considered with a higher degree of safety when malignancy is suspected.[5] Pre-operative embolization reduces intraoperative bleeding and allows for more meticulous dissection.[4,5] External surgery is the treatment of choice by larynogfissure, thyroidotomy, or pharyngotomy,[1,2,4] or an endolaryngeal approach with excision by laser.[5] Radiotherapy is considered when surgery is contraindicated, in an attempt to halt tumour growth.[3]

We contribute another case of LPG to the literature, the diagnostic suspicion of which must be complemented with non-invasive imaging studies, although the advances being made in endolaryngeal microsurgery make it easier to take samples for biopsy when malignancy is suspected. Arteriography is needed to provide more precise surgical planning and to make the surgery itself easier, given that pre-operative embolization decreases perioperative bleeding. Endolaryngeal surgery and laser resection, whether external or via microsurgery is the treatment of choice when there are no contraindications.

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