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Case Report

Supraglottic Paraganglioma in Type IV Paraganglioma Syndrome Treated with CO2 Laser: A Minimally Invasive Approach

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1. Abstract

Paragangliomas are benign tumors derived from extra-adrenal paraganglia. They are rarely asymptomatic and may involve the head and neck region in 3% of cases. Asymptomatic supraglottic paraganglioma associated with paraganglioma syndrome is described in this case report. A 35-year-old woman affected by papillary thyroid cancer, was referred to our Unit for preoperative evaluation for total thyroidectomy. During fibrolaryngoscopy, a massive hypervascular lesion was identified in the supraglottic region, originating from the right aryepiglottic fold. Neck MRI with contrast and total body PET-CT revealed multiple similar lesions in the neck and thorax. The patient underwent embolization of the laryngeal lesion with Histoacryl glue and subsequently endoscopic removal with CO_2 laser with simultaneous total thyroidectomy. Pathology confirmed the diagnosis of supraglottic paraganglioma and papillary thyroid cancer. Her postoperative course was uneventful. Genetic testing was positive for succinate dehydrogenase subunit B (SDHB) mutation thus indicating a Type IV paraganglioma syndrome. This is an unusual case of asymptomatic paraganglioma of the larynx associated with multiple paragangliomas and papillary thyroid cancer. CO_2 laser surgery could be a safe and minimally invasive treatment for this kind of lesion.

2. Key words

Paraganglioma; Multiple paraganglioma; Supraglottic paraganglioma; CO_2 laser; Papillary thyroid carcinoma

3. Introduction

Paragangliomas (PGL) are rare benign slow-growing tumors arising from extra-adrenal paraganglia [1]. Head and neck PGL represent 3% of all cases [2]. In the literature, only 80 cases of laryngeal involvement are described, often concerning the supraglottic region, arising from paraganglia of the superior laryngeal artery and nerve [1]. Only a few of them are associated with multiple PGL.

The most common symptom of laryngeal PGL is dysphonia, but dysphagia, stridor and foreign body sensation may also occur [1]. Macroscopically, PGL are submucosal, highly vascularized, red-bluish lesions, covered by intact mucosa [1]. Since they bleed easily, surgical treatment is usually preceded by embolization [3]. To date, the most frequently reported surgical removal is excision by a cervicotomic approach [1], whereas carbon dioxide (CO_2) laser has been used in association with partial epiglottectomy [4].

4. Case report

A 35-year-old woman had a positive family history of papillary thyroid carcinoma (PTC), thus she underwent blood analysis which showed elevated levels of thyroid-stimulating hormone (TSH) and anti-thyroid peroxidase antibodies. Neck ultrasonography revealed an isthmus nodule which was analyzed with fine-needle aspiration cytology, yielding PTC TIR5 (according to the Italian consensus for the classification and reporting of thyroid cytology) [5].

As preoperative evaluation for total thyroidectomy, fiberoptic laryngoscopy was performed and it revealed a hyper-vascularized, red-purplish lesion of the right aryepiglottic fold, expanding towards the glottis. It was covered by intact mucosa, without reduction of vocal fold mobility. Cervical enhanced magnetic resonance imaging (MRI) revealed an enhancing solid supraglottic lesion (10x27x30 mm) originating from the posterior-lateral region of the hypopharynx. Two similar lesions were diagnosed at the right jugular foramen and left carotid bifurcation. 18 (^{18}F) dihydroxyphenylalanine (DOPA) whole-body positron emission tomogra-

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phy (PET) /computed tomography (CT) (Figure 1) confirmed enhancement of the neck lesions, but others were diagnosed in the right jugular region, superior mediastinum and aorto-pulmonary area. No abdominal lesions or suspect lymph nodes were present. Urine metanephrine levels were normal. The surgical plan was discussed and we proposed endoscopic CO_2 laser removal of the supraglottic lesion, because of the possibility of future dyspnea and dysphagia associated with a simultaneous total thyroidectomy.

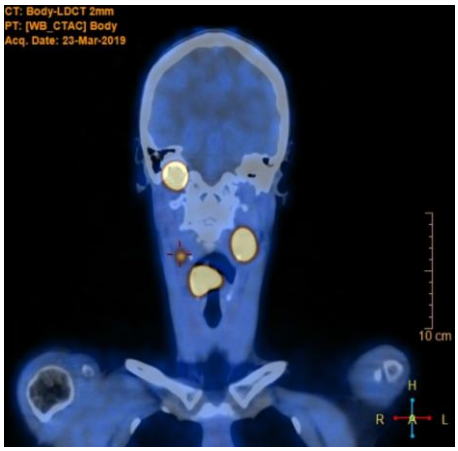

Figure 1: PET-CT scan with 18F-DOPA. Multiple head and neck sites of high uptake intensity are shown. Two other lesions with high DOPA uptake (not shown in this image) were encountered in the superior mediastinal region behind the trachea and in the aorto-pulmonary window.

Angiography and selective embolization of the right superior and inferior thyroid arteries were performed with Histoacryl glue (Figure 2). Then the patient underwent endoscopic CO_2 laser en-bloc removal of the supraglottic lesion with jet ventilation anesthesia. After removal, an orotracheal tube was inserted and total thyroidectomy was performed. Prophylactic micro-tracheostomy was accomplished. No complications occurred peri-operatively and the patient was discharged after tracheostomy closure on the 6th post-operative day.

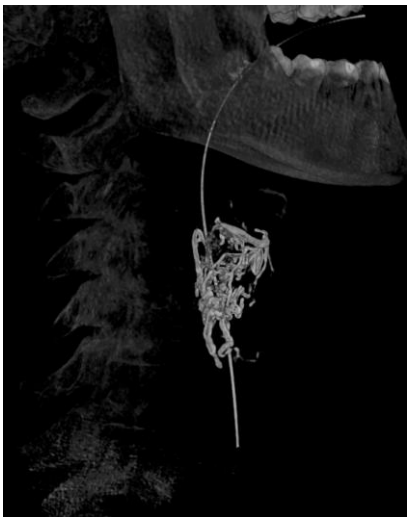

Figure 2: 3D maximum intensity projection (MIP) reconstruction, sagittal view, post-embolization procedure for preoperative devascularization of laryngeal glomus tumor showing the cast of Histoacryl glue utilized.

Pathologic examination confirmed the diagnosis of laryngeal PGL and PTC (staged pT1bN0 [6]) associated with Hashimoto's thyroiditis. No regional lymph node metastases were encountered. After selective embolization, the patient experienced right vocal fold palsy; she was referred to early speech therapy [7] with complete mobility restoration (Figure 3). Fiberoptic laryngoscopy and 68Ga-DOTA-Phe(1)-Tyr(3)-octreotide (^{68}Ga-DOTATOC) - PET/CT did not reveal any pathological findings (Figure 4). After 6 months, she underwent a genetic test, which was positive for succinate dehydrogenase subunit B (SDHB) mutation (Exon 7, variant c.701T>C p.Leu234Pro) related to type IV paraganglioma syndrome [8-9]. Her father and two sisters were also positive. At one-year follow-up, the patient is alive and in good health, takes oral replacement therapy and has started radiometabolic therapy.

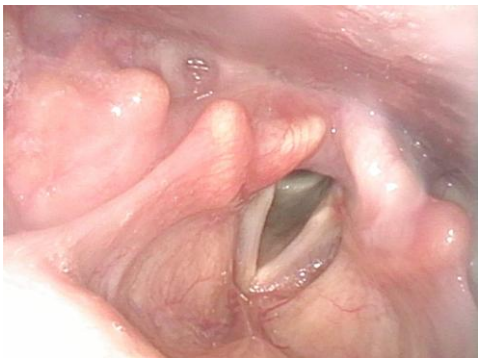

Figure 3: Post-operative fiberoptic laryngoscopy showing complete recovery of the right vocal cord palsy after speech therapy.

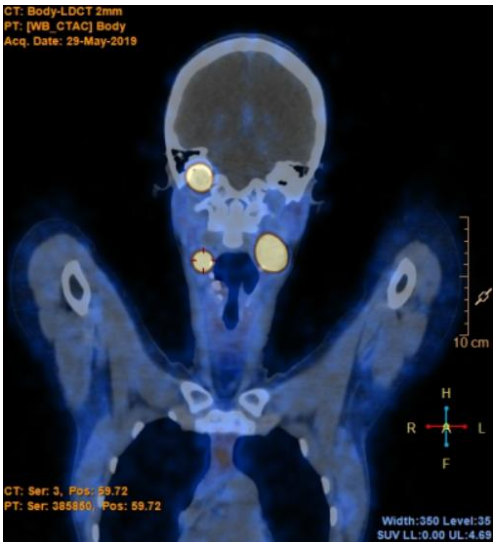

Figure 4: Post-operative ^{68}Ga-DOTATOC-PET/CT showing no further uptake in the right supraglottic region. The other head and neck lesions are unchanged.

5. Discussion

To date, about 80 cases of laryngeal PGL [1] have been described in the literature. Overall, all patients complained of at least one symptom, such as dysphonia or dysphagia, or had lateral cervical masses [1]. Our case represents a very rare finding, even though the size

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of the lesion was quite large. Moreover, it was associated with PTC without pheochromocytoma, thus multiple endocrine neoplasia type 2 (MEN2) and PGL/PTC/pheochromocytoma syndrome were excluded [10]. Only three cases with an association between PTC and PGL, without pheochromocytoma, have been described [10-12], and PGL always originated from a carotid glomus. To the best of our knowledge, ours is the first case of laryngeal PGL associated with PTC and multiple other localizations in the setting of type IV paraganglioma syndrome.

Although a CT scan has been suggested for any submucosal laryngeal mass, MRI represents the gold standard to detect PGL: it can recognize primary lesions, and characterize vascular structures and adjacent tissues [1,13]. Moreover, DOTATOC is a somatostatin analogue, and because of the common expression of somatostatin receptors in PGL, ^{68}Ga-DOTATOC-PET/CT has a higher sensibility and specificity to detect neuroendocrine tumors, compared with a CT-scan [14].

In selected cases, surgery can be performed with a cervicotomic approach or under microlaryngoscopic view with a CO_2 laser [1]. Intra-operative bleeding control is more difficult during the latter approach, especially if closure of major feeding vessels is required. In our case, CO2 laser removal was feasible thanks to effective preoperative embolization, which provided less blood loss and better exposure of the tumor during the surgical procedure [3]. Sesterhenn et al. [4] described the only case treated with this approach, but they needed partial epiglottectomy to achieve better exposure of the tumor. That patient also experienced vocal fold paralysis, which recovered with speech therapy.

In our case, a multidisciplinary consultation was necessary to plan treatment. We decided to perform endoscopic removal of the supraglottic PGL with CO_2 laser in jet ventilation and, subsequently, to proceed to an easier orotracheal intubation to perform a safe total thyroidectomy. A temporary prophylactic tracheostomy was carried out due to the extensive laryngeal laser procedure together with an anterior cervicotomy. After pathological confirmation of the benign PGL and well differentiated PTC, a genetic test was performed. A very rare SDHB mutation was found, thus revealing type IV paraganglioma syndrome which can be associated with malignant paraganglioma [8-9]. Due to the anatomical localization of the other PGL and the related high risk of cranial nerve damage during surgery, the patient was enrolled in a clinical trial and has been treated with radiometabolic therapy. A wait-and-scan approach can be a good option in PGL due to their benign and generally slow-growing characteristics [2,9]. In this case, radiometabolic therapy with somatostatin analogue was proposed due to the possibility of malignancy and multiple locations of PGL [9,15].

6. Conclusions

Head and neck localization of multiple PGL syndrome is very rare. Association with PTC in a non-MEN context is even rarer. Laryngeal PGL is rarely asymptomatic but surgical removal is always indicated. In this scenario, a minimally invasive endoscopic CO2 laser removal can be a reliable and safe procedure with a low morbidity and complication rate.

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