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Case Report

Laryngeal Paraganglioma

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- Paraganglioma
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Abstract

Introduction: Paragangliomas (PGL) are uncommon tumors originated from the neural crest. They represent only 0.012% of all tumors and 0.6% of tumors in head and neck topography. The goal is to report a rare disease of atypical presentation.

Case Report: A 40- year- old male presented to our service complaining of neck pain for six months with worsening in the last month, foreign body sensation in throat and hoarseness. Direct laryngoscopy was performed and mass of nodular aspect, rich in vascularization was detected in the left aryepiglottic fold. The patient underwent laryngeal microsurgery to remove the lesion with a suspected diagnosis of cyst or granuloma. Twelve hours after the surgical procedure reproach was needed, because the patient had heavy bleeding at the site. The hemorrhage was contained with cauterization of the bleeding site. Histopathology showed the presence of neuroectodermal lineage cells, which required immune histochemistry examination to establish final diagnosis, which revealed PGL. In postoperative follow-up the patient presented with improved clinical picture without neck pain.

Conclusion: PGL are rare tumors, usually benign, rich in vascularization and slow growing. Surgical excision is the treatment of choice.

INTRODUCTION

Paraganglioma (PGL) are uncommon tumors originated from the autonomic nervous system (ANS) throughout the body [1]. So far more than 20 areas of the human body with PGL tissue have been reported. They represent only 0.012% of all tumors and 0.6% of tumors in head and neck topography. Ninety percent of PGL are located in the adrenal gland. When found in that location it is called pheochromocytoma. The most common PGL in the head and neck region are located in the carotid body, followed by the jugular, tympanic and vagal branches. Other rare locations are: larynx, nasal cavity, orbit and trachea. Very few PGL have been described in the larynx; however, some authors suggest that perhaps they are more common than recognized in the literature [2,3].

CASE PRESENTATION

A 40- year- old male presented to our service complaining of neck pain for six months with worsening in the last month, foreign body sensation in throat and hoarseness. No co morbidities were reported. In his physical examination no abnormalities were detected. Direct laryngoscopy was performed and mass of nodular aspect, rich in vascularization was detected in the left aryepiglottic fold. The mass measured 01 centimeter in its largest width and did not obstruct the patient's airway. The presumed diagnosis was cyst or granuloma (Figure 1). The patient received drug therapy with omeprazole and showed no reduction of the mass in a 60-day follow up. Laryngeal microsurgery was performed to remove the lesion. During the excision frozen section pathology was not ordered due to absence of malignant aspect. Twelve hours after the surgical procedure reproach was needed because patient had heavy bleeding at the site. The hemorrhage was contained with cauterization of the bleeding

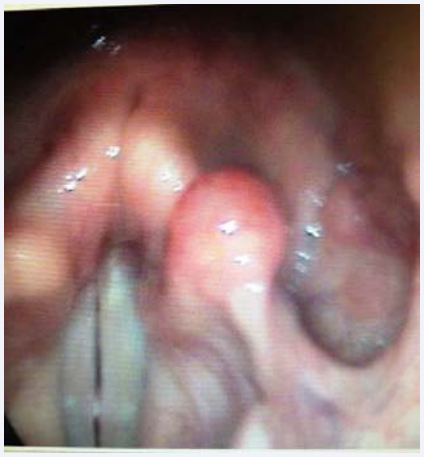

Figure 1 Rigid Videolaryngoscopy – Mass in the left aryepiglottic folds.

site. Histopathological examination showed the presence of neuroectodermal lineage cells, requiring immunohistochemistry to establish definitive diagnosis. The panel was positive for S-100, chromogranin, enolase and SYN, consistent with PGL. The patient continues on postoperative follow up 12 months after the procedure and presents with improvement of the clinical picture with no cervical pain.

DISCUSSION

PGL derive from neural crest cells associated with the parasympathetic nervous system and are located in the proximity of arteries and cranial nerves [4]. PGL represent a distinct entity among neuroendocrine tumors originating from the neural crest and are located next to the sympathetic and parasympathetic nerves. They have the capacity to produce a wide variety of neuroendocrine products. Head and neck PGL are associated with the vagus nerve and its branches, such as the superior laryngeal nerve. Laryngeal PGL are rare and most often located in the supraglottic region, comprising 2.77% of all head and neck PGL [5,6]. They are most frequently benign tumors, although metastasis and invasive growth into surrounding tissues have been reported in less than 10% of all cases [2,4].

PGLs are three times more common in women than men, in contrast to literature we present a case in which a man was affected. Most PGL occur in the supraglottic region, mainly in the right aryepiglottic folds (82%) and less commonly the glottis and subglottis (15% and 3.5%, respectively) [5,6]. Hoarseness, pain, dysphagia and neck mass are the cardinal symptoms of this tumor [2]. Our patient presented with three of these symptoms, we believe neck mass was not present due to the location and size of the tumor. Supraglottic PGL can present clinically with hoarseness and difficulty breathing because of the mass effect causing airway obstruction (Figure 2). The topography of the disease can affect the mobility of the vocal cords, causing breathy voice. It's usually seen as a small submucosal, red-colored mass located in aryepiglottic fold. The nature of these highly vascular tumors often result in hemoptysis and in some cases the onset of heavy bleeding during the biopsy [5]. Endoscopic biopsy is still controversal due to the risk of uncontainable bleeding and eventual aspiration.

Imaging studies should be performed if PGL diagnosis is suspected. They determine the location and extent of PGL providing essential data to establish the surgical treatment of the tumor. Careful study of the anatomical relations, vascularization and adjacent nerves, help anticipate potential surgical risks [7]. Doppler ultrasounds may be useful for carotid body tumors, but for laryngeal tumors its use is limited. Magnetic resonance imaging can provide better tissue characterization and detailed evaluation of the tumor, including its location, size and vascularization. A "salt and pepper" appearance is expected in PGL. Digital subtraction angiography is a preoperative vascular mapping tool and not frequently used for diagnostic purpose. It also allows preoperative embolization, reducing the risk of intraoperative hemorrhage [8].

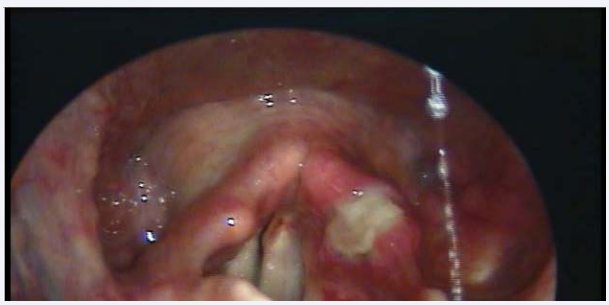

Figure 2 Rigid Videolaryngoscopy – 15 days after surgical procedure.

Definite diagnosis is achieved after adequate pathology studies. The identification of a laryngeal neuroendocrine tumor requires a combination of light microscopy, immunohistochemistry markers and electron microscopy [4]. The immunohistochemistry panel may include CD56, chromogranin A, SYN, P63, S100, viment, CD34, CD31, calponin, cytokeratins, actin, SMA, HMB45, melan-A and IV collagen [9]. The anti-cytokeratin antibody and chromogranin markers are most useful in differentiating PGL from neuroendocrine carcinomas. The most emblematic histopathological finding is the Zellballen pattern, which consists of chief cell nests surrounded by sustentation cells [10]. Zellballen patter was present as well as three immunohistochemistry markers in the presented case, including chromogranin.

Post-operative complications include dysphonia and dysphagia which in most cases resolve spontaneously in about 4 to 6 months [11]. Our patient had an uneventful late postoperative period in a 12 month follow up with no complications reported.

PGL are uncommon tumors, usually benign and slow growing. In the laryngeal region they are well defined and rich in vascularization. Surgical excision is the treatment of choice.

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