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CASE REPORT

Granular cell tumor on the larynx

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Abstract

Granular cell tumors (GCTs) are neoplastic lesions rarely seen in adults. GCTs are most commonly found in the head and neck region, and exceptionally on the larynx. Laryngeal GCTs symptoms and signs may manifest as hoarseness, dysphagia, coughing, hemoptysis, and stridor. We report a case of a 36-year-old woman, who complained of dysphonia. Her clinical history, pathological findings, and treatment are addressed in this article.

Keywords: granular cell tumor; larynx; dysphonia.

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Introduction

Abrikossoff was the first author to describe granular cell tumors in 1926. It was believed that the neoplasm derived from skeletal muscle cells[1]. In 1974, Sobel and Marquet supported the theory that the tumor originated from undifferentiated mesenchymal cells or Schwann cells using immunohistochemical staining technique[2].

Although controversies have arisen on the origin of GCTs, the immunohistochemical characterization shows positivity to S-100 and neuron-specific enolase (NSE) and negativity to muscle antigens. Additionally, electron microscopy reveals an ultrastructural similarity to the Schwann cells. Thus, the neural origin of GCTs is currently accepted[3].

The pathogenesis of laryngeal granular cell tumors is not well established. Inflammatory, degenerative, regenerative, and congenital etiologies have been proposed. Hwang researched patients with GCTs and found smoking (20 pack-years) as a common habit. Although the immunohistochemical investigation for p53 is one of the most well-known oncogenes involved in smoking-related head and neck cancers, it was negative[1].

Case report

A 36-year-old black woman attended outpatient care for a one-year-long dysphonia. There was no history of dyspnea, odynophagia, dysphagia, weight loss, and alcoholism. She was a long-term smoker (21 pack-years). On her physical exam, no cervical lymphadenopathy was found. And oroscopy and rhinoscopy showed no abnormalities. A video laryngoscopy was carried out and

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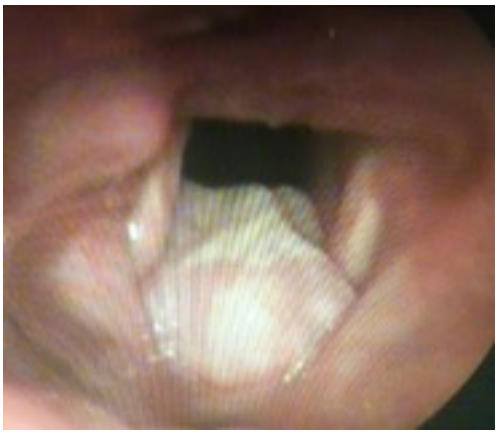

Figure 1. A vegetating tumor in the anterior commissure affecting both of the vocal cords.

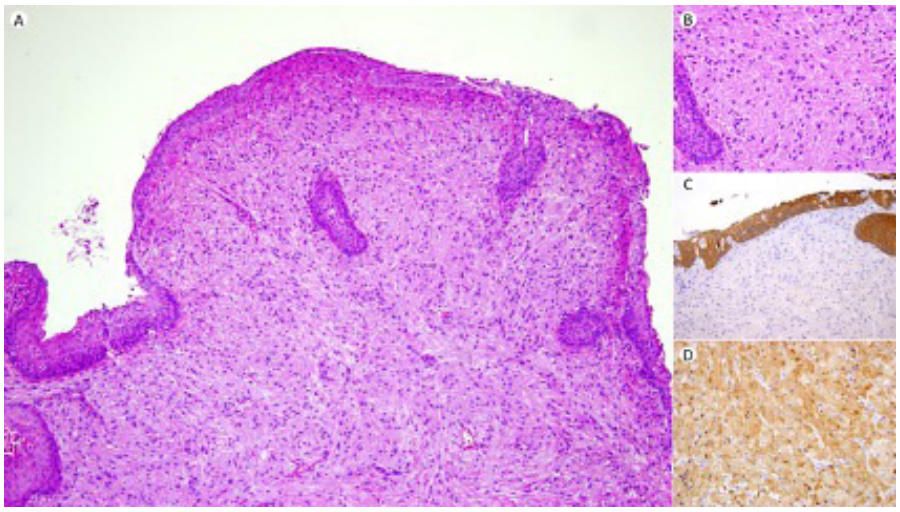

Figure 2. Laryngeal granular cell tumor. **A** - A sheet of uniform epithelioid cells interdigitating with the epithelium (HE). **B** - Granular eosinophilic cytoplasm and small round nuclei (HE). Immunohistochemical reactions: **C** - no immunoexpression for cytokeratins (AE1AE3); and, **D** - positivity for S-100 antibody.

registered a vegetating lesion in the anterior commissure. The mass extended up to 1/3 of the middle glottis and compressed the vocal cords bilaterally (Figure 1). A CT scan was performed and demonstrated a laryngeal norm position, vocal cords symmetry maintaining the lumen, and preserved glottal space configuration. No pharynx and infraglottic region abnormalities were detected. Tongue projection and mouth floor were normal. The histologic and immunohistochemical studies showed cell positivity for S-100 and vimentin antibodies, and negativity for cytokeratins. Therefore, the diagnosis of a laryngeal granular cell tumor was established (Figure 2). The patient was submitted to a type 5 cordectomy and showed no pain or postoperative dyspnea. Pathology examination detected positive margins, and a new approach was taken for the complete granular cell tumor removal. There have been no signs of recurrence in a two-year follow-up with a videolaryngoscopy

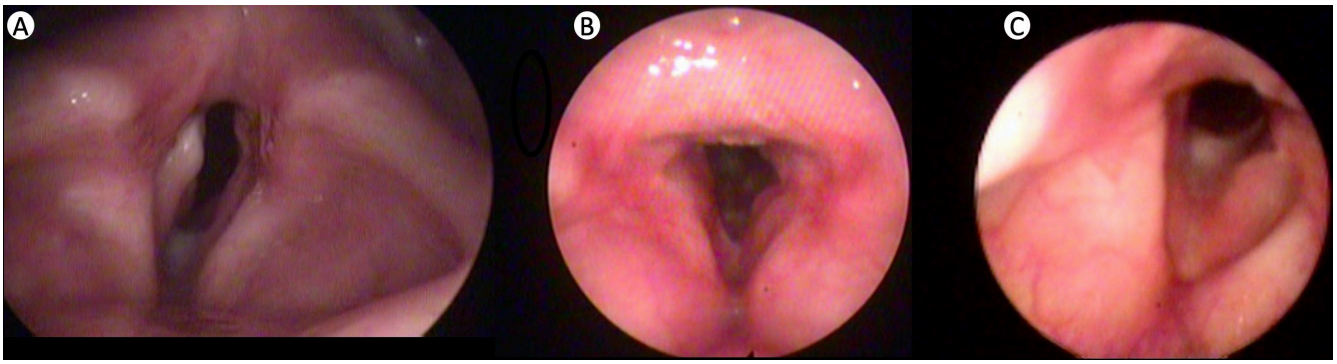

Figure 3. Post-operative endoscopic view of the larynx. **A** - 45 days post-operative. **B** - 1 year post-operative. **C** - 1 year and 9 months post-operative.

every three months (Figure 3). The patient has been under speech therapy to improve vocal patterns.

Discussion

Granular cell tumors are benign neoplasm derived from the Schwann cells[2,3]. Half of the GCTs occur in the head and neck region, however, less than 10% are located on the larynx[1]. The peak incidence is between 30 and 60 years and the mean age of 34 years[1].

The most common symptoms and signs are hoarseness, coughing, dysphagia, odynophagia[4]. Other rare symptoms include stridor, otalgia and hemoptysis[1]. Most of these GCTs cases are a surprising diagnosis to the surgeon, made through the pathological examination only. The differential diagnosis list includes carcinomas, papillomas, polyps, granulomas, cysts, neuromas and neurofibromas[1].

In the laryngoscopy, the GCTs appear as a small, well-circumscribed, submucosal whitish or yellowish mass. GCTs frequently mimic vocal cord polyps or granulomas grossly. The tumor is a solid lesion and can be mistaken for a squamous cell carcinoma on a tomography scan. Laryngeal GCTs mainly involve the vocal cords, but they are reported in the anterior commissure, arytenoids, vestibular folds, subglottis, and postcricoid region[5].

The GCTs cells are polygonal slightly elongated. The abundant cytoplasm contains eosinophilic granules, strongly Periodic Acid Schiff stained. The nucleus of GCTs cells is small. Mitosis figures are rare or absent. Immunohistochemically, the cells are positive for S-100, vimentin and specific neuronal enolase antibodies[2,5].

According to the literature, malignant laryngeal GCTs are rare. The rate of malignant onset was reported as 0.6%[1].

The treatment of choice for laryngeal GCTs is surgical excision with negative margins, and it can be proceeded by direct endoscopy, laryngofissure, and laryngectomy. GCTs do not respond to radiotherapy. When surgical margins are negative, the rate of recurrence is around 8%. On the other hand, when surgical margins are positive, the recurrence increases by up to 50%. And the

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recurrence is multicentric in 16% of the uncompleted excisions[4]. According to the localization, planning is crucial for the total or subtotal tumor excision to keep a functional larynx[4].

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