

Case Report

Atypical Granular Cell Tumor of the Larynx: Report of an Unusually Aggressive Tumor

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Abstract

Introduction: Granular cell tumors are uncommon benign neoplasms. The head and neck is the most common site, usually localized to the anterior tongue. The larynx is rarely involved, representing 1.6% to 3.7%.

Case report: We present the case of a 31-year-old, non-smoking female, with a history of recurrent cysts in the pyriform sinus. She consulted for dyspnea. By physical exam, there were no specific findings in head and neck area. Cervical computed tomography demonstrated a tissular mass in the laryngeal area. Laryngoscopy showed a submucosal tumor of the aryepiglottic fold. A biopsy was performed. Histopathological examination revealed round or oval-shaped tumor cells with abundant eosinophilic granular cytoplasm. The nuclei are sometimes small and regular and sometimes hyperchromatic and pleomorphic. The tumor cells showed immunoreactivity for S100 protein and vimentin. The tumor was diagnosed as an atypical granular cell tumor.

Conclusion: Pathologists should be aware of this exceptional lesion. So, that the patient could have appropriate treatment.

Keywords: Larynx; Granular cell tumor; Histology

Introduction

Granular cell tumor (Abrikosov's tumor) of the larynx is a very uncommon lesion. The exact origin of granular cell tumor is still unclear. However, most authors believe that the neoplastic process derived from neuroectoderm. This tumor can develop in any part of the body and 50% occur in the head and neck area, especially in the tongue. Laryngeal granular cell tumors (GCTs) represent 3% to 10% of all cases. In this report we present the clinico-pathological characteristics of laryngeal granular cell tumor and we discuss the differential diagnosis of this particular entity in this rare location.

Case Report

We present the case of a 31-year-old non-smoking female with a history of recurrent cysts in the pyriform sinus excised in 2010 and 2012. She consulted for dyspnea. By physical examination, there were no specific findings in head and neck area. Laryngoscopy revealed a mass of the left piriform sinus and the laryngeal wall partially obstructing the epiglottis. Cervical computed tomography (CT) demonstrated a tissular mass of $48 \times 28 \times 22$ mm protruding in the laryngeal light. This mass depended from the left pyriform fossa. There was no cervical lymphadenopathy. Direct laryngoscopy showed a submucosal tumor of the aryepiglottic fold flowing the pharyngolaryngeal wall and the left pyriform sinus. A biopsy was performed. Histopathological examination revealed round, oval-shaped or spindle cells with focally marqued cellularity. The tumor cells had abundant eosinophilic granular cytoplasm (Figures 1 and 2). The nuclei were sometimes small and regular; and sometimes hyperchromatic and pleomorphic. The tumor cells showed immunoreactivity for S 100 protein and vimentin (Figure 3). There was no immunoreactivity for cytokeratin, CD45, HMB45, thyroglobulin and GPAP. The presence of cellular pleomorphism, moderate atypia without important mitosis and positivity for S 100 protein advocated for the diagnosis of atypical granular cell tumor. Therefore, the patient had a partial laryngectomy. Histological examination showed the same aspect as the biopsy.



Figure 1: Tumor cells with different shapes: round, oval-or spindle (H&E, ×200)

Discussion

In 1926, Abrikossof was the first who described a granular cell tumor [1]. Its histogenesis is controversial and laryngeal involvement is uncommon. Despite the preferential involvement of the posterior larynx [2], tumors may also be found in the anterior larynx, vocal folds and arytenoids [3]. A slight female predominance exists for GCTs. The female-to-male ratio is estimated at approximately 3:2. They

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Figure 2: oval-shaped tumor cells with abundant eosinophilic granular cytoplasm. The nuclei present moderate pleomorphism without mitotic features or necrosis (H&E, ×200).



Figure 3: Immunohistochemical staining showing positive staining for PS100 (IHC ×200).

occur between the third and fifth decades of life. However Laryngeal involvement is more observed in younger men, with 36 years median age [4]. Rare pediatric cases have been described in the literature [5]. GCTs grow slowly during 6 to 7 months before patients seek medical advice. Symptoms depend on the localization and size of tumor. The most common symptoms for laryngeal GCTs are horseness, cough, hemoptysis, stridor, otalgia and dysphagia [6]. On laryngoscopic exam, GCTs appear to be firm, fixed or pediculated. Ulceration is not common and borders are usually well defined. Encapsulation is also not frequent but the presence of infiltrative borders simulate invasion observed in malignant GCT. Their size usually ranges between 0.3 cm and 3 cm [7]. GTC diagnosis is confirmed by pathological exam. These tumors typically have an infiltrative growth pattern and a histiocytoid-type appearance. The cytoplasm of a granular cell tumor contains ill-defined, spindle-shaped or polygonal vacuolated nuclei and eosinophilic granules. These granules are strongly positive for PAS reaction. Immunohistochemistry reveals strong positivities for S-100 protein and neuron-specific enolase [8]. Nuclear pleomorphism and mitotic figures are not usually seen in benign granular cell tumors. Pseudoepitheliomatous hyperplasia of the overlying mucosa may be present in 50% of cases. Rare cases can have moderate degree of epithelial atypia. The pseudoepitheliomatous hyperplasia can be a

J Cancer Sci Ther, an open access journal ISSN: 1948-5956 clue, on superficial biopsy specimens, that one may be dealing with a granular cell tumor. These tumors also stain for vimentin, myelinassociated glycoprotein (Leu-7) and CD68 [9]. The malignant type of granular cell tumor is uncommon and accounts for only 1% to 2% of all cases [10]. Although, criteria for malignant GCTs have not yet been determined; malignancy is suspected in cases which have nuclear pleomorphism, frequent mitoses and an increase in the nucleus/ cytoplasm ratio, spindle-shaped cellularity, necrosis and vacuolated nucleus with large nucleoli. It is also clinically suspected in cases where the tumor size is ≥ 4 cm and where the tumor grows rapidly and shows recurrence or infiltration into the adjacent tissue. Metastases when present are to the cervical lymph nodes and lungs. It has been shown that mutations that occurred in the lamin B receptor coding gene are able to induce an altered nuclear morphology in granulocytes [11]. In addition, by visualizing the fate of lamina associated domains in single cells, recent observation revealed that G9a functions as a regulator of nuclear lamina contacts [12]. These findings taken together indicated that epigenetic mechanisms might be involved in the regulation of nuclei shape, since the main role of G9a is establishing and maintenance of H3K9me1/2 in the mammalian cells [13]. Interestingly, as recent study illustrated that G9a also protects DNA methylation at specific loci [14], aberrantly gain and lose of DNA methylation by alternation of chromatin loading of G9a might also affect the shape of the nuclei. Therefore, it is interestingly to study the expression level and chromatin distribution of G9a and its associated epigenetic markers in the cells that display abnormal nuclei shape. The treatment procedures of GCT depend on the site and extension of the lesion. Surgical excision is the treatment of choice. Small tumours are managed by endoscopic resection or by thyrotomy with total resection of the tumour. Laryngofissure and partial laryngectomy are used for larger tumours. Radiation therapy is a poor treatment option as these tumors are radio resistant.

Despite adequate resection, tumors recurrence rate is 8% to 21%, and usually occurred at the primary site [7]. Furthermore, the tumor may recur in a different aspect from the primary tumor. The long term natural history is still unknown because of the rarity of this lesion. As such, patients should still be followed for recurrence with laryngeal examination after mass resection particularly when a subtotal resection has been performed. Pathologists should be aware of this diagnosis that we come across by coincidence, keeping in mind that it may be among the posteriorly located glottis pathologies. Moreover, a tight cooperation should be made with the expert so that the patient could have adequate treatment.

Conflict of interest

None

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