Granular Cell Tumor- A Rare Laryngeal Tumor

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Abstract

Introduction:
Granular cell tumors (GCTs) are an uncommon neoplasm, originating in any part of the body. The most common sites of origin are the head and neck, while the larynx is a relatively uncommon location.

Case report:
We report an uncommon case of a 27-year-old man who presented persistent hoarseness of voice for 3 months. Endoscopic laryngeal examination revealed a mucosal growth with smooth surface along the right vocal cord. The patient was treated by surgical resection under fine dissection by microlaryngoscopy.

Conclusion:
Our case demonstrates various differential diagnoses possible in laryngeal neoplasms. It also shows that immunocytochemistry plays an important role in differential diagnosis.

Keywords:
Granular cell, Larynx, Neoplasm.

Received date: 16 Oct 2013
Accepted date: 14 Dec 2013

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Introduction
Granular cell tumors (GCTs), also known as Abrikossoff tumors, are benign, slowly growing neoplasms, presumed to be of Schwann cell origin. They may occur anywhere in the body, although 50% occur in the head and neck. The most common site is the tongue, while the larynx is involved in approximately 10% of all cases. GCTs typically develop in the fourth and fifth decades of life, and are relatively rare in children. The vast majority of tumors arise from the posterior aspect of the true vocal folds; approximately half of these extend into the subglottis. These tumors are covered with an intact squamous mucosa and are smooth, polypoid, firm, homogeneous, and well circumscribed with a gray-yellow cut appearance (1).

Case Report
A 27-year-old male presented to our center with a history of hoarseness of the voice for the past 3 months. There was no history of any vocal abuse. The patient was a smoker, but there were no other specific previous or family histories. Endoscopic laryngeal examination revealed a mucosal growth with smooth surface along the right vocal cord arising from the anterior commissure and extending to the posterior commissure. The bilateral vocal cords displayed equal movement. On physical examination, there were no specific findings in the head and neck area.

Microlaryngeal surgery was performed under general anesthesia and revealed a proliferative mucosal growth with smooth surface along the right vocal cord from the anterior to the posterior commissure, with involvement of the undersurface of the vocal cord (Fig. 1). A linear incision was made over the superior margin of the right vocal cord. The lesion was dissected from the entire length of the vocal cord. On excision of the lesion, gelatinous material was seen to be extruding from the lesion. Histopathological examination revealed vocal cord mucosa with an unencapsulated subepithelial tumor nodule. The tumor cells consisted of granular cytoplasm with eosinophilic globules. The nuclei were round to be oval in shape, and there was an absence of atypia. The resected margins were free of tumor tissue (Fig.2,3).

Fig 1: Microlaryngeal examination showing lesion along the right vocal cord

Fig2: Histopathology slide showing tumor consisting of rounded and polygonal cells with indistinct cellular borders

Fig3: Histopathology slide showing small, vesicular and centrally or eccentrically located nuclei and abundant granular cytoplasm
Staining for tumor markers was performed to obtain a confirmative diagnosis. The tumor was positive for S-100 and negative for chromogranin, cytokeratin and synaptophysin (Fig. 4). The diagnosis was confirmed as GCT (myoblastoma) of the right ventricle of the larynx.

The patient was advised to undertake complete voice rest for 10 days and was initiated on speech therapy. He experienced progressive improvement of the voice over the course of the next 2 months, and also stopped smoking. The patients has been undergoing regular follow-up at the outpatient clinic and has reported no post-operative complications. There was no evidence of recurrence at a 2-month follow-up laryngeal endoscopy (Fig.5).

Discussion

In 1926, Abrikosoff first described a GCT which was named myoblastic myoma based on the differentiation of the skeletal muscle. Although much controversy exists concerning the origin of this type of tumor, immunohistochemistry shows positivity for the S-100 protein, neuron-specific enolase and negativity for muscle-related antigens. Additionally, electron microscopy reveals a cluster of squamous cells with a continuous perineurium-like basement membrane, dehydrated axons surrounded by angulated bodies (2-4); supporting Schwann's cell theory.

Laryngeal GCTs appear as small, rounded, firm submucous tumors covered with whitish gray or yellow, flat and normal mucosa, and often resemble vocal fold polyps or granulomas. Their small size (less than 2 cm) and their well-circumscribed, but nonencapsulated featurea are typical (5). These tumors mostly involve the posterior third of the true vocal cords, but are also found on the anterior commissure, arytenoids, the false vocal folds, subglottis, and the post-cricoid region. In contrast, polyps are usually connected to vocal abuse and are gel-like, smaller structures when compared which GCTs which are more fibrotic. Laryngeal local trauma is connected to the etiology of granuloma, whether it is physical or chemical. They are similar to GCTs macroscopically, but are also smaller and usually located in the posterior larynx. They do not evolve so slowly. GCTs tend to be solid and homogenously enhancing on computed tomography (CT) scans, often mimicking a squamous cell carcinoma (6); but in this case we had no specific findings. The diagnosis was made after histologic examination of biopsied material from direct laryngoscopy. In 50–65% of all cases, there is development of 'squamous pseudoepitheliomatous hyperplasia' in the overlying epithelium. An endoscopic biopsy specimen including only the mucosa may result in a misdiagnosis of a well-differentiated squamous cell
carcinoma(7). Other differential diagnosis include paraganglioma and adult-type rhabdomyoma.

Therefore, a biopsy specimen should include sufficient normal tissue adjacent to the tumor. The cytoplasm of a GCT contains ill-defined, spindle-shaped or polygonal, vacuolated nuclei and eosinophilic granules (8). These granules are strongly positive for the Periodic Acid Schiff (PAS) reaction, and immunohistochemistry reveals strong positivity for S-100 protein and neuron-specific enolase. These tumors also stain for vimentin, myelin-associated glycoprotein (Leu-7) and CD 68 (KP-1).

The malignant type of GCT is uncommon and accounts for only 1–2% of all cases. Although criteria for malignant GCTs have not yet been determined, this malignant tumor 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.

Once the diagnosis of a benign granular cell tumor is confirmed, complete surgical resection becomes the appropriate treatment. During removal, securing a negative free margin is mandated. A frozen section is recommended to confirm properly performed surgical resections without leaving any tumorous remains (9). Radiation therapy or chemotherapy is not recommended because it may be less responsive or induce malignant transformation. Even though surgical resection is performed properly, there is an 8–21% recurrence rate, usually at the primary site. Some of these recurrences may be due to a failure to diagnose multiple lesions. Moreover, in cases of inadequate removal with remnants of the tumor at the resection margins, the tumor may recur (10).

Although it is rare for GCTs to occur in the larynx, accurate diagnosis and appropriate treatment and follow-up are mandatory. Regular long-term follow-ups are required using fiber-optic laryngoscope to confirm its recurrence. GCTs grow slowly and evolve on average between 6 and 7 months before patients perceive a notable discomfort and seek medical advice (11).

Conclusion

Laryngopharyngeal GCTs are uncommon tumors that are conventionally treated with surgical resection. Close collaboration with an experienced pathologist is necessary to establish the diagnosis, with immunohistochemical evaluation required to confirm the diagnosis.

References