Case Report

Clinical Problem solving │ Pathology Laryngeal Tumor

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Abstract

Giant cell tumor (GCT) is mostly a benign bone tumor. Only 2% of GCTs arise in the head and neck region, mostly in cranial bones. Laryngeal GCTs (LGCTs) are relatively rare and present as hoarseness and neck swelling, mostly in middle aged men. LGCTs are usually only locally destructive, and pulmonary metastases and malignant transformation are rarely observed. We present here a rare case of a man in his 50s with LGCT of the larynx originating in the thyroid cartilage lamina. The pathogenesis, physical, imaging and pathologic findings and treatment options of LGCT are discussed.

CASE

A man in his 50’s presented with hoarseness that had progressively worsened over the previous month. His past medical history was significant for acid reflux. He denied a history of smoking. Flexible-fiberoptic laryngoscopy revealed bulging of the left false vocal fold and irregularity and impaired movement of the left true vocal fold. Neck fullness was palpated at the level of the left thyroid cartilage lamina. Computed tomography of the neck demonstrated a space-occupying lesion involving the left larynx and measuring 3.5X3X4 cm, with destruction of the left laryngeal cartilages and penetrating the left thyroid gland lobe. No neck lymphadenopathy was noted. Magnetic resonance imaging of the neck demonstrated a left laryngeal submucosal transglottic mass measuring 2X 3X4 cm causing destruction of the left thyroid cartilage lamina and penetrating the left thyroid gland, but sparing the anterior commissure and the right larynx. The lesion was enhanced with gadolinium. There was no neck lymphadenopathy. Micro-direct laryngoscopy revealed a large submucosal mass covered with normal-appearing mucosa in the left ventricle. In surgery, the mucosa over the mass was elevated, the mass was biopsied and the biopsy was sent for histopathology examination. Hematoxylin and eosin staining (Figure-1) demonstrated a dual population of cells: osteoclastic-like multinucleated giant cells (*) with ill-defined cytoplasm and mononucleated cells (**). A few apoptotic cells (arrowhead) and mitotic divisions (arrow) were also present. Ki67 showed a 20% labeling index, and immunostaining for vimentin was positive. Immunostaining for cytokeratins (CK, CK5/6, CK-AE1, and CK-AE3), desmin, actin, thyroglobulin and thyroid transcription factor 1 (TTF-1) were negative.

What is your diagnosis?

1. Laryngeal poorly differentiated squamous cell carcinoma.
2. Laryngeal giant cell tumor.
3. Laryngeal poorly differentiated sarcoma.
4. Laryngeal brown tumor.

DIAGNOSIS

Laryngeal giant cell tumor

DISCUSSION

Giant cell tumor (GCT) is mostly a benign bone tumor. The common skeletal sites for GCT are the distal end of the femur and the proximal end of the tibia. Only 2% of GCTs arise in the head and neck region, mostly in cranial bones, specifically, the maxilla, base of the skull and mandible, in descending order [1]. Laryngeal GCTs (LGCTs) are relatively rare [2]. The most common laryngeal locations are the thyroid, cricoid, and epiglottis cartilages, in descending order [1,3]. LGCTs are more common in middle-aged women, with an average age of 41 years, while LGCTs are more prevalent in males, with a male-to-female ratio of 9:1 [3]. The close association between the male predilection of a LGCT and the earlier endochondral ossification of the laryngeal cartilaginous skeleton in males suggest that LGCTs may originate from primitive mesenchymal cells in ossifying laryngeal cartilages [3].

The most common clinical presentation of a LGCT is hoarseness and anterior neck swelling. Other symptoms include pain, dysphagia and dyspnea [1]. Preoperative radiological studies do not assist in the differential diagnosis of LGCTs from other laryngeal tumors. However, neck computerized tomography may assist in determining laryngeal and extralaryngeal tumor extension, cartilage destruction, periosteal reaction, tumor...
mineralization, and neck node involvement. Neck magnetic resonance imaging may demonstrate the origin and extension of the tumor [4]. Unlike other benign bone-originating lesions, LGCTs may accumulate FDG [5,6], therefore positron-emitted computed tomography may also assist in the diagnostic process, although its role is still controversial.

The definitive diagnosis of a LGCT is established by tumor biopsy. LGCTs have pleomorphic histologic features. They contain two components: one composed of diffuse osteoclast-like multinucleated giant cells with acidophilic vacuolated cytoplasm, and the other composed of round, oval or polygonal mononuclear stromal cells [7]. The nuclei in the multinucleated giant cells are similar to those of the stromal cells. The rich fibrovascular stroma of the tumor contains numerous thin-walled capillaries, often with small areas of hemorrhages. The pathological differential diagnosis may be challenging because the giant cell component of a LGCT resembles the giant cell subtype of a pleomorphic sarcoma. The differentiation between these two tumors is made according to the characteristic findings on hematoxylin & eosin staining. Immunohistochemistry stains are not usually mandatory to establish the diagnosis of an LGCT, however they may be performed to rule out other tumors in the differential diagnosis, including poorly differentiated squamous cell carcinoma (positive cytokeratins), pleomorphic sarcoma of smooth or striated muscular origin (positive desmin and actin) and poorly differentiated thyroid carcinoma (positive thyroglobulin and TTF-1).

LGCTs are usually only locally destructive, and pulmonary metastases [8] and malignant transformation [9] are rarely observed. As such, the overall prognosis of LGCT can be considered as being favorable.

Treatment of LGCT is controversial. The majority of patients are managed by complete tumor excision (via total or partial laryngectomy). The role of radiotherapy is undetermined. Bell et al. [10] suggested laryngeal preservation with radiotherapy. LGCTs frequently tend to recur despite adequate local excision. Therefore, radiotherapy was also suggested as an adjuvant treatment, but, again, without clear evidence of its efficiency [2]. Adjuvant radiotherapy is highly acceptable in orthopedics, however, there is a high incidence of radiation-induced sarcomas in bones previously harboring GCTs that were primarily treated by radiotherapy [2].

Our patient underwent a fronto-lateral laryngectomy and a left hemithyroidectomy for complete tumor resection. The tumor was seen intra-operatively to originate in the left thyroid cartilage lamina and penetrate the thyroid gland and adjacent striated muscles. Due to the extent of the tumor and its extralaryngeal spread, the patient later received intensity-modulated radiotherapy (62cGy) to the larynx and bilateral necks. At the 4-year follow-up, he was alive with a functional hemi-larynx, mild subglottic stenosis, per-oral nutrition and no evidence of disease.

REFERENCES