ABSTRACT

Objective: (1) To review the epidemiology, pathophysiology, and treatment of giant cell tumor of the larynx (2)
Method: A case of giant cell tumor of the larynx from a tertiary care university teaching hospital is presented, including the patient’s clinical course and subsequent treatment. The case is discussed with emphasis on the treatment options for such a lesion.
Results: The patient underwent hemilaryngectomy with radial forearm free flap reconstruction and fifteen of cycles of chemotherapy. The patient is disease free at 34-months follow-up. The patient was decannulated and continues to have a good voice with excellent quality of life to this day.

Conclusion: Our experience demonstrates that giant cell tumor of the larynx may present as a malignant neoplasm without adversely affecting the patient’s prognosis when treated aggressively with surgical resection and adjacent chemotherapy.

INTRODUCTION

Neoplasms arising from the supporting laryngeal cartilages are uncommon [2,3,6]. These tumors represent less than 2% of all primary laryngeal neoplasms, and include chondromas, chondroblastosomas, osteoblastomas, giant cell reparative granuloma, brown tumors of hyperparathyroidism, aneurysmal bone cyst, and osteosarcoma, and spindle cell or sarcomatoid carcinomas [2,6,8,9]. Giant cell tumors (GCTs) are predominantly benign neoplasms of the long bones, most often occurring at the distal femur or proximal tibia [2,3,6,7]. Although rare in the head and neck, GCTs have a predilection for areas of endochondral ossification in the base of skull, namely the sphenoid, ethmoid and temporal bones [2,3,6,9]. GCTs of the larynx, as first reported by Wessely in 1940, are considered to be locally aggressive, but benign and are exceedingly rare [2,6,7,8,9,10]. Fewer than 30 cases have been described in the literature, one of which was reported to be malignant [8,12]. Upon further review, several authors have questioned the validity of the malignant designation given to this case [6,9]. We present a case of a young man with laryngeal GCT marked by malignant osteosarcomatous transformation. He was treated with a partial laryngectomy and free-flap reconstruction resulting in excellent postoperative function. To our knowledge this represents the first reported case of a malignant GCT of the larynx.

CASE PRESENTATION

A 34-year-old man presented with a three month history of an asymptomatic right-sided anterior neck mass. Ultrasonography and fine-needle aspiration were performed at an outside institution. Ultrasonography revealed a 3 x 4 cm mass in the anterior portion of the right thyroid lobe. A fine-needle aspiration specimen was obtained. The aspirate was non-diagnostic for malignancy and the patient was referred for further management. Two weeks prior to presentation the patient underwent thyroidectomy for a thyroid nodule that was subsequently found to be a reactive lesion. The patient denied any symptoms of voice alterations.

One week later the patient underwent right laryngobronchoscopy, right horizontal hemilaryngectomy, and left radical far scream free flap reconstruction. In-office biopsy revealed the ipsilateral palatoglossal muscles, thyroid cartilage up to the midline, ventricular cartilage, lateral portion of the cricoid cartilage and hyoid bone, and lateral pharyngeal wall including the entire piriform sinus. Microscopic and deep muscle margins were negative for tumor involvement. The bulk of the tumor was the larynx was submucosal with significant involvement of the thyroglottis. Microscopically, the tumor appeared to arise from the thyroid cartilage, and was composed of osteoclast-like multinucleated giant cells diffusely distributed among atypical mononuclear spindle cells. The nuclei of the osteoclast-like giant cells were indistinguishable from those of the multinuclear stromal cells (Figure 2). There were significant atypia including cellular pleomorphism, increased mitotic activity with atypical mitosis, necrosis, and foci of lacy-like malignant osteosarcoma present throughout the tumor (Figure 3). The cellular proliferation was associated with an arborizing vascular network, and frank vascular invasion was identified. The final diagnosis was malignant osteosarcoma with osteoclast-like multinucleated giant cell transformation. A bone scan and positron emission tomography (PET) scan failed to reveal a primary osseous site of origin, and thus ruled out the possibility of metastases to the larynx.

Figure 1. Axial computed tomography (CT) scan without contrast demonstrating a large heterogeneous 10.6 x 8 x 4 cm mass, involving the right thyroid lobe with destruction of the right thyroid cartilage and pharyngeal extension.

Figure 2. Osteoclast-type giant cells are diffusely admixed with oval and spindle atypical cells with morphologically similar nuclei. Mitotic figures are readily identified. (hematoxylin and eosin stain, 40X original magnification)

Figure 3. Lacy-like malignant osteosarcoma was present surrounding microvascular and osteoclast-type giant cells. (hematoxylin and eosin stain, 20X original magnification)

DISCUSSION

Giant cell tumor (GCT) represents 5% of all primary bone neoplasms [2,3,6,7]. Although only 2-4% of all GCTs occur in the head and neck [2,3,6,7], the most common orthopedic location for GCT is the proximal femur [1,2,3,6,7,9]. In the head and neck, the bones of the skull base are most frequently involved [2,3,6,8,9]. GCT of the larynx has been reported to occur in the hyoid bone, epiglottis, and cricoid cartilage, but most often involves the thyroid cartilage [2,3,6,7]. Although GCT has been treated successfully with GCT, our patient demonstrated vascular invasion and significant cellular pleomorphism associated with the presence of malignant osteosarcoma. Despite surgical therapy, radiotherapy and/or chemotherapy have all been reported to be viable options for the treatment of laryngeal GCT, the literature demonstrates a preference for conservative, but complete surgical resection, which parallels the treatment model that is advocated in the orthopedic literature [1,2,3,6,7,8,9,10]. Partial laryngectomy with a goal to preserve voice and swallowing function appears to be the standard [2,3,8]. As in our case, reconstruction with a radial forearm free-flap provides an effective means of filling the anatomical defect, while preserving speech and deglutition. Although complete laryngectomy with a goal to preserve voice and swallowing function appears to be the standard [2,3,8]. As in our case, reconstruction with a radial forearm free-flap provides an effective means of filling the anatomical defect, while preserving speech and deglutition. Although complete laryngectomy with a goal to preserve voice and swallowing function appears to be the standard [2,3,8]. As in our case, reconstruction with a radial forearm free-flap provides an effective means of filling the anatomical defect, while preserving speech and deglutition. As in our case, reconstruction with a radial forearm free-flap provides an effective means of filling the anatomical defect, while preserving speech and deglutition.

Regardless of treatment modality, the prognosis for laryngeal GCT is excellent with no reports in the literature of recurrence, metastases or death related to the tumor [2,3,6,8,9,10]. This differs significantly from the pattern exhibited by GCT of the long bones [1,2,6,10]. Of note, however, is one report of a questionable malignant laryngeal GCT by Coyas et al. in 1974 [12]. It is an unusual case that persisted despite numerous attempts at local excision and eventually involved the overlying skin [6,9,12]. The case was initially described as malignant, but has subsequently been reinterpreted as a benign soft tissue lesion [6,9]. Furthermore, an osteocartilaginous origin was not confirmed, which calls into question the pathological diagnosis [6,9]. As of the four-year follow-up, there was no evidence of recurrence after the involved skin was excised. If Coya’s case were accepted as tumor seeding rather than a true malignancy, our patient represents the first reported case of a malignant GCT of the larynx. Our patient remains free of disease 42 months postoperatively, and 34 months following last cycle of chemotherapy. Definitive comments cannot yet be made with regard to likelihood of cure given this relative short follow-up interval.

CONCLUSION

Laryngeal GCT is an uncommon neoplasm that arises from the supporting laryngeal cartilage, and it generally presents as a benign lesion. Although GCT can present as a benign neoplasm, it may display an aggressive and infiltrative growth pattern. Tissue biopsy demonstrates histopathologic findings similar to GCT of the long bones. Conservative, but complete surgical resection without adjuvant therapy offers an excellent prognosis. As with their counterpart in the long bones, laryngeal GCT may degenerate and become atypical. We present the first case of a clearly malignant laryngeal GCT managed by complete surgical excision and adjuvant chemotherapy without evidence of recurrence 34 months following completion of all therapy.

REFERENCES