Osteosarcomas of the Larynx

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Objectives/Hypothesis: The objective was to review the etiology, presentation, treatment, and prognosis in patients with osteosarcoma of the larynx using an illustrative case example. Study Design: Case report and literature review. Methods: A case is reported, and a review of all available published cases of osteosarcoma of the larynx is presented. Results: Osteosarcoma of the larynx may be a difficult clinical diagnosis. To be able to reach the correct diagnosis, a high index of suspicion and due diligence in obtaining deep biopsy specimens are important. Treatment is primarily surgical. Outcomes for this disease are generally poor, with most patients dying of the disease as a result of pulmonary metastasis. Conclusion: Osteosarcoma of the larynx is a rare entity, with only 14 cases reported to date. Pathological confirmation of osteoid is required for diagnosis. It is likely that aggressive surgical intervention directed at complete tumor extirpation is the treatment of choice, although long-term prognosis is poor. Key Words: Larynx cancer, osteoid, osteosarcoma.

Laryngoscope, 115:74–77, 2005

INTRODUCTION

Sarcomas of the larynx are relatively rare, non-epithelial derived neoplasms constituting only 0.32% to 2% of all laryngeal neoplasms, of which 50% are malignant. Osteosarcoma represents the rarest of these soft tissue neoplasms. To date, only 13 cases of primary osteosarcoma of the larynx have been reported (Table 1). In the present study, we review this rare neoplasm with an illustrative case example.

CASE REPORT

A 69-year-old Caucasian female with a history of smoking had been treated with partial resection followed by chemoradiation therapy for a supraglottic squamous cell carcinoma 4.5 years previously. During the past 3 years, she had had chronic aspiration and progressive dyspnea. Multiple biopsies at two outside institutions were performed. The biopsy findings were negative for malignancy and were consistent with chondroradionecrosis. It was at this time that the patient presented to our institution.

Examination revealed the laryngeal framework to be larger than the normal. Fiberoptic laryngoscopy revealed diminished glottic opening attributable to cartilaginous overgrowth. Computed tomography (CT) scan demonstrated dystrophic calcification outside of the confines of the larynx (Fig. 1). As a result of these findings and the patient's progressive dyspnea, an elective tracheotomy was performed. Biopsies of the larynx revealed dystrophic calcification, but no frank evidence of malignancy. Because of a high clinical index of suspicion of chondrosarcoma, as well as the lack of functional value to her larynx from the standpoint of either respiration or phonation, consent was obtained for a total laryngectomy.

On dissection through the neck, the patient was found to have a firm mass involving the larynx, strap muscles, and thyroid gland with circumferential involvement of the esophagus. Intraoperative frozen-section specimens revealed osteosarcoma. The patient underwent total laryngectomy, total thyroidectomy, total pharyngectomy, and total esophagectomy with gastric pull-up. The patient's postoperative course was uneventful. However, the patient developed pulmonary metastases and died of disease 10 months later.

Final pathological findings revealed osteosarcoma of grade 4 (Fig. 2). Moderate calcification of neoplastic osteoid islands separated by large cells with irregular, hyperchromatic nuclei were observed. The tumor formed a band of closely spaced cells with large nuclei and scanty cytoplasm.

DISCUSSION

Osteosarcoma of the larynx was first described in 1942. Malignant sarcoma of the larynx is a rare entity, making up 0.5% to 1% of laryngeal neoplasms. To date, 13 cases of osteosarcoma of the larynx have been described. Of these 13 cases, 12 cases were in men, and 1 of these cases presented following radiation therapy for other neoplasm.

To be classified as osteosarcoma, ostoid must be produced in the tumor. Osteoid does not necessarily have to be calcified and may be noncalcified, lace-like, and trabecular in appearance. Production of ostoid is important in differentiating osteosarcoma from anaplastic chondrosarcoma. The most common presenting complaints in osteosarcoma of the larynx are hoarseness, dyspnea, and acute airway obstruction. In our review, we noted that the mean duration of symptoms was approximately 6 months before diagnosis. Diagnosis is often made following multiple biopsies, because of the difficulty in obtaining a confirmatory specimen.