Salvage Hemi-Cricoidectomy without Reconstruction of a Locally Aggressive Chondrosarcoma in a 37-Year Old Patient

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Abstract

We report a case of a locally aggressive 3.3 cm chondrosarcoma treated with salvage hemi-cricoidectomy, removing 60% of the cricoid cartilage, without any reconstruction, in an otherwise healthy 37-year old male. The patient was decannulated six weeks post op and is a year later breathing without stridor, fully rehabilitated.

Introduction

Chondrosarcoma (CS) of the larynx was first described in 1935 [1]. Any bone that grows by endochondral ossification is at risk for a chondrosarcoma [2]. These bulky tumors can invade and destroy adjacent bone and/or cartilage and spread into surrounding tissue. CS is more prevalent in men than women and generally occurs in the sixth and seventh decade of life [3]. Less than 1% of laryngeal tumors are represented by CS and most commonly occurs in the cricoid with involvement of the posterior lamina [1,4,5]. Other subsites would include the thyroid cartilage, arytenoids, and epiglottis [1]. For locally aggressive CS primary total laryngectomy is commonly recommended, since the tumors are radio-resistant.

Work up for CS includes CT and/or MRI to demonstrate location, dimensions and the amount of calcification of the tumor [5]. CT scans can define a fine, punctate stippled to coarse "popcorn" calcification within the tumor [2,5]. MRI scans can be beneficial by identifying the extent of possible tumor penetration into the surrounding soft tissue [4].

Fine needle biopsies can be difficult to obtain due to the hard nature of the lesion. Therefore, a final diagnosis is often performed on the surgical specimen [5]. Histological diagnosis of Laryngeal CS is based on criteria by Lichtenstien and Jaffe who in 1943 described and categorized malignant cartilaginous tumors [6]. Criteria includes: (I) cells with plump nuclei, (II) more than an occasional cell with two such nuclei and (III) giant cartilage cells with large single or multiple nuclei or with clumps of chromatin [6]. However, Evans et al., grouped CS into grades I, II and III according to the mitotic rate, nuclear size, and cellularity [7]. In Grade I (low grade), well differentiated, small, densely staining nuclei are seen with two or more nuclei within one lacuna. In addition, in an isolated area there may be a small number of larger pleomorphic nuclei. Furthermore, in Grade I mitotic activity and an increase in cellularity should be absent. In Grade II (moderately differentiated), the mitotic rate is low (less then two mitosis per 10 HPF) and moderate size nuclei and an increase in cellularity are seen towards the periphery, as well as tumor lobules in isolated areas of the tumor. Grade III (poorly differentiated or high-grade) is defined by the identification of two or more mitoses in the most active area of the tumors. Spindle cell pattern is also prevalent in large areas in Grade III tumors, and the mitotic rate is in excess [8].

Conservative surgical treatment is considered the treatment of choice [2]. However, a total laryngectomy is often recommended when laryngeal defects are revealed that cannot be reconstructed. Adjuvant radiotherapy to other sites than larynx have been ineffective, and by extrapolating that experience to laryngeal CS external beam radiation is subsequently in general not considered a viable treatment option. There is very limited amount of data exist regarding chemotherapy for laryngeal CS [4].
Case Presentation

We present the case of a 37-year-old male with history of Grade I chondrosarcoma of the cricoid cartilage diagnosed in 2015. The patient has a strong family history of malignancies including eight brothers now deceased from various cancers. He presented to his primary care physician with a sensation of fullness in his neck without pain, stridor/dyspnea, dysphonia, dysphagia or odynophagia. He was eventually found to have a 3.3 cm cricoid mass near the left crico-arytenoid joint by CT and MRI scans. The mass was confined to the cricoid cartilage and had not invaded the thyroid, esophagus, or vocal cords, but the left vocal cord was partly immobilized, likely due to compression of the crico-arytenoid joint. The tumor engaged roughly 60% of the cricoid. A biopsy from an outside institution confirmed a low grade CS, Grade I. He was initially evaluated at three outside institutions, all of whom recommended total laryngectomy. He refused this and first opted for concurrent chemotherapy and radiation therapy at one of these institutions, despite understanding that it would be unlikely that the tumor would respond to such regimen. He underwent treatment 3/2016 to 4/2016. Post-treatment imaging (Figure 1,2) showed that the mass had neither regressed or progressed significantly from chemo-radiation. He presented to us for a forth opinion, seeking an alternative surgical option in the form of a larynx sparing partial cricoid resection.

The patient was again informed that standard of care is a total laryngectomy, but he again refused. After careful examination and thorough discussions about risks and benefits we decided to offer him a left hemi-cricoidectomy and temporary tracheal stent with the possibility of a low tracheostomy which he elected to receive on 1/3/2017. He was carefully informed that a salvage total laryngectomy could be the next step if the larynx would become non-functional or if negative margins could not be achieved. He was also informed that a trachea-esophageal fistula could develop. The initial partial cricoidecetomy was performed under general anesthesia with the patient intubated by endotracheal tube, no 6. We made an incision through the cricothyroid membrane and resected 80% of the tumor (Figure 3) to the cricoid lamina, leaving the remaining posterior 20% to protect the esophagus, and since it was hard to distinguish how much to resect intraoperatively. The resection spared the left vocal cord and did not extend into the arytenoid. A 6-cm long tracheal stent was then placed by a thoracic surgeon using guide wires and a fiber optic endoscope, in order to prevent collapse of the airway. The stent was meant to stay in for 3 weeks until the lateral airway would be stable.

The patient's hospital course was complicated by an intense immediate postoperative cough leading to significant subcutaneous emphysema, involving the upper torso, upper arms, neck and lower face, as noted in PACU. He also developed a small right-sided pneumothorax which later that night required a chest tube placement for less than 24 hours. His lung function improved rapidly and his swallowing function was normal throughout the postoperative phase, with no sign of trachea-esophageal fistulation. He could rapidly be advanced from liquids to soft mechanical food, and never had a feeding tube placed. The subcutaneous edema subsided within 5 days and he was discharged home in stable condition on 1/9/2017. A postoperative follow up CT neck confirmed that there was still tumor remaining in the cricoid lamina (Figure 4).

Final pathology report confirmed the diagnosis of a low grade chondrosarcoma with positive posterior margins at the cricoid lamina (Figure 5). On 1/27/2016, the patient returned for removal of the stent and resection of residual tumor. At this time we decided to perform a low tracheostomy after removal of the tracheal stent to increase the access to the posterior cricoid and to avoid the need for another stent, which most likely had been the reason for the violent cough.
postoperative cough he had experienced after the first operation. At this time we were able to resect the posterior left cricoid lamina down to the tracheo-esophageal membrane and all the way up to the inferior aspect of the left arytenoid. It was now clear that the left lateral tracheal wall, which previously had been resected, had not collapsed.

His subsequent hospital stay was uncomplicated. There was some minor subglottic edema which prompted us to start IV dexamethasone post-operatively.

On 3/8/2017, 10 weeks post operatively from the initial procedure and 6 weeks after the salvage resection and tracheostomy. He had a slightly weak voice and on fiber optic examination the left vocal cord showed slightly decreased movements but no significant edema and only some mild granulation of the anterior and lateral wall (Figure 6). He was subsequently decannulated.

On 4/11/2017 he reported he was running 4 miles a day without any shortness of breath or stridor. Laryngoscopy demonstrated a normal right vocal cord but a sluggish left vocal cord. The left sided subglottic granulation tissue was stable.

On 7/18/2017 he returned for a 1 month follow up visit. He reported developing an upper respiratory infection with a productive cough but he denied any change in voice or stridor, only mild shortness of breath. He was treated with antibiotics and steroids with overall improvement of symptoms. On 9/12/2017, laryngoscopy demonstrated complete immobility of the left vocal cord, but with completely healed granulation tissue. CT demonstrated no sign of recurrent CS (Figure 7). On examination 12/5/2017 he had developed minimal inspiratory stridor on heavy inhalation, which did not restrict him on a daily basis, and also inter mittent mild hoarseness (Figure 8). On 3/13/17, a year after completion salvage hemi-cricoidectomy his condition is stable and he is fully rehabilitated.

Discussion

Chondrosarcoma (CS) of the larynx is a rare pathological condition. It most commonly occurs in the cricoid cartilage, mostly involving the posterior lamina, and the standard of care of locally aggressive tumors is total laryngectomy. We report a case of a 37-year old male with a biopsy verified low grade CS that involved more than half of the cricoid. In a two-stage procedure, we resected about 60% of the cricoid, decannulated him within a month and a year later he is recurrence-free and fully rehabilitated. We conclude that the risk of tracheal collapse, tracheo-esophageal fistula formation and need for reconstructions might be exaggerated, even for significant cricoid resections.

References
