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Adenoid Cystic Carcinoma of the Trachea

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ABSTRACT

Background: Primary malignant neoplasms of the trachea are very rare and there is limited information available on this subject. Adenoid cystic carcinoma is a slow-growing malignant tracheal tumor and the best method of treatment is surgical resection. This study was conducted to evaluate patients with adenoid cystic carcinoma of the trachea who underwent surgical treatment.

Materials and Methods: In this descriptive study, 9 patients treated for adenoid cystic carcinoma from 1995 to 2007 at the Mashhad Ghaem Hospital and Tehran Imam Khomeini Hospital were assessed.

Results: There were 9 patients (3 males and 6 females) with a mean age of 56.3 years. Dyspnea and stridor were the most common presenting symptoms (88.8%). All patients underwent rigid bronchoscopy and biopsy. The most common site of involvement was the lower third of trachea (44.4%); 77.7% of patients underwent surgical resection. Death occurred in one patient after tracheal resection due to aspiration pneumonia (14.2%). Postoperative radiotherapy was performed in 28.4% of patients because of positive surgical margin and in 22.2% due to inappropriate location of the tumor after bronchoscopic ablation. During a three-year follow up, one patient (11.1%) had tumor recurrence. Resection with post-operative radiotherapy was performed for him. The three-year survival was 88.8%.

Conclusion: Because of the nature of adenoid cystic carcinoma of the trachea, surgical resection is the best method of treatment. But if surgical margins are positive post-operative radiotherapy will be necessary. In patients who are not candidates for resection, radiotherapy can be an effective alternative treatment. (*Tanaffos* 2008; 7(4): 49-54)

Key words: Adenoid cystic carcinoma, Cylindroma, Trachea, Treatment

INTRODUCTION

Primary tumors of the trachea are rare and often malignant. Adenoid cystic carcinoma (ACC)

is among the primary malignant tumors of trachea which include 25% of tracheal malignancies. ACC is locally invasive and usually amenable to resection. Although late local recurrence after resection is a feature of this tumor, excellent long-term palliation is commonly achieved after both complete and incomplete resections with a small difference in

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survival rates. Long-term control can also be obtained by radiotherapy alone (1). ACC is frequently referred to as cylindroma, but this term should be abandoned because it implies that the tumor is benign. ACC is a slow-growing neoplasm, and the patient usually experiences symptoms for more than a year before the diagnosis is made (2).

The aim of this study was to assess patients with primary tracheal ACC treated at the Mashhad Ghaem Hospital and Tehran Imam Khomeini Hospital from 1995 to 2007.

MATERIALS AND METHODS

In this descriptive (case-series) study, all patients treated for adenoid cystic carcinoma of the trachea at the Mashhad Ghaem Hospital and Tehran Imam Khomeini Hospital from 1995 to 2007 were assessed. Age, sex, clinical signs and symptoms, diagnostic method, site of involvement, method of treatment, complications and the three-year survival were studied in all patients.

RESULTS

Nine patients were studied. The M/F ratio was 1/2. They were between 38-75 years-old and the mean age of patients was 56.3 yrs. The most common presenting symptoms were dyspnea and stridor (88.8%), other symptoms were cough (77.7%), bronchospasm attacks similar to asthma (33.3%) and hemoptysis (11.1%).

Chest x-ray and CT-scan were performed for all patients before surgery; CT-scan showed the site of the tumor and its local extension in 100% of patients, while lateral neck views and chest X-rays demonstrated them in 55.5% of the patients. Figure 1 shows the CXR and Figure 2 demonstrates the CT-scan of a patient with an ACC adjacent to the carina.

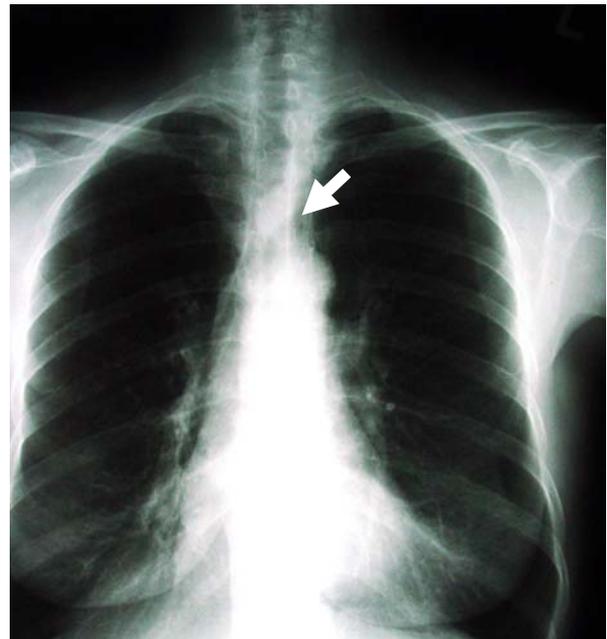


Figure 1. Chest x-ray demonstrated the site of tumor adjacent to the carina (white arrow)

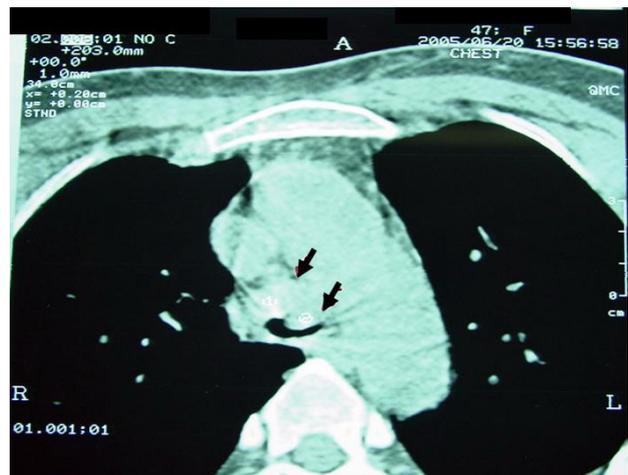


Figure 2. CT-scan of the same patient which shows the site of tumor (black arrows).

In patients who underwent tracheal resection, extubation was done in the operating room and no complication occurred after surgery. But one patient died one month after surgery because of aspiration pneumonia. The mortality rate of surgery was 14.2%.

In 2 out of 9 patients, surgery was not performed due to the location of tumor; in one patient the tumor

was 1 cm below the vocal cords requiring laryngectomy, but the patient did not consent to surgery. In the other patient surgery was not performed because of involvement of the carina and left main bronchus. Complete ablation of the tumor by rigid bronchoscopy was performed for these patients and they were referred to an oncologist for radiotherapy.

Among patients who underwent surgery (7/9), 2 required oncology consultation for postoperative radiotherapy treatment because of positive microscopic surgical margin of resection (28.4% of resected cases) which was done. We did not refer patients with free margin of resection for radiotherapy.

During the 3-year follow-up, 1 patient died one month after surgery and the remaining were all alive (88.8%) and in 1 patient the tumor recurred 2 years after primary surgery. He underwent re-resection and post-operative radiotherapy and there has been no recurrence to date.

Decisions were made for treatment based on pathologic results and location of tumor. Location of tumor, in 2 patients (22.2%) was in the upper third, in 3 patients (33.3%) in the middle third and in 4 patients (44.4%) in the lower third of trachea. The most common site of involvement was in the lower third of the trachea.

After rigid bronchoscopy and biopsy, 7(77.7%) patients underwent tracheal resection, in 3 of them tracheal resection was performed after the result of frozen section. But, in the remaining 4 patients resection was performed later.

Due to the site of involvement, in 1 patient (14.2%) cervical approach, in 3 patients (42.8%) cervico- mediastinal approach and in 3 patients (42.8%) posterolateral thoracotomy approach was used for surgery.

Figure 3 shows intraoperative resection (specimen) of lower third ACC and Figure 4 shows the method

of anastomosis in the same patient.

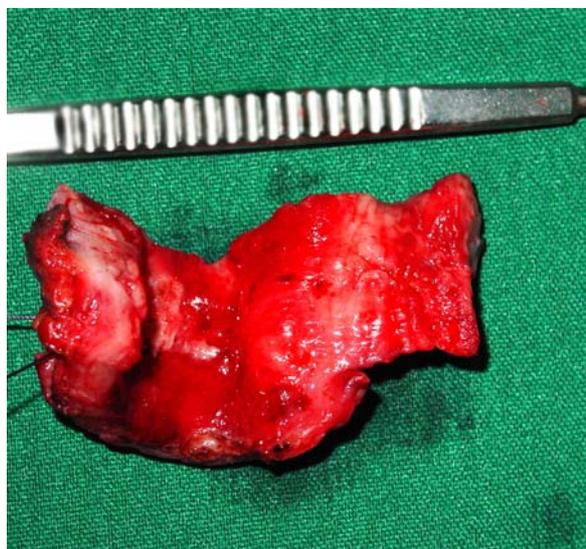


Figure 3. Tumor specimen resected from the lower third of the trachea.



Figure 4. Technique of anastomosis of the distal and proximal ends of the trachea after resection of the tumor via a posterolateral thoracotomy approach.

DISCUSSION

Tracheal malignancy accounts for less than 0.2% of all malignancies of the respiratory tract. The most frequent malignant primary tracheal tumors in adults are squamous cell carcinoma and adenoid cystic carcinoma (3). These tumors are predominant in men with mean age of 57.3 years (4).

In our study, M/F ratio was 1/2 and the mean age

of patients was 56.3 years.

This tumor arises from the bronchial gland and is histologically identical to those that arise from the salivary glands.

When the tracheal lumen is reduced to one-third of its normal size (cross-sectionally) patients suffer from cough, dyspnea, exertional dyspnea (85%) and stridor (80%). Other symptoms include wheezing (similar to asthma symptoms), hemoptysis (20%) and recurrent pneumonia (5%). The mean interval between the early symptoms and diagnosis is 12 months (5).

In our study, 88.8% of patients had dyspnea and stridor, 77.7% had cough, 33.3% had bronchospasm attack similar to asthma, and 11.1% had hemoptysis.

A careful inspection of the tracheal air column on lateral neck radiographs and hyperextensions reveal the presence of tumor but do not provide specific information required for planned resection and reconstruction. Tracheal tomograms are not sufficient for detection of extra-luminal extension of the tumor or lymph node invasion and this technique is no longer available in most medical centers (7).

A three-dimensional (helical) CT-scan of the central airway can provide precise anatomic information for endobronchial and surgical procedures (8).

In our study, lateral neck x-ray, CXR, and CT-scans were performed in all patients before surgery. CT-scan showed the site of tumor and its local extension in 100% of the patients but lateral neck views and CXR demonstrated the tumor in 55.5% of the patients.

Grillo et al. showed that a bronchoscopic examination is always necessary for diagnosis in the operating room where ventilating, rigid bronchoscopes and biopsy forceps are available and a trained anesthesiologist is nearby. When the indication for primary tracheal resection is clear-cut, bronchoscopy can be deferred until the time of the

operative procedure and frozen section can be used to determine the histology (9).

In our study, after imaging, all patients underwent rigid bronchoscopy and biopsy. Seven patients (77.7%) underwent tracheal resection out of which 3 underwent frozen section with tracheal resection and in 4, resection was performed later.

Tracheal resection and end-to-end anastomosis (using various releasing techniques to minimize the tension) are the primary form of therapy for ACC of the trachea.

Gaissert et al. suggested that resection of the trachea or carina in ACC patients was associated with long-term survival superior to palliative therapy particularly for cases with complete resection and negative airway margins (10).

In a study conducted by Grillo et al. on 198 patients with primary tracheal tumors (SCC and ACC), 147 tumors were excised (74%) and resection and primary reconstruction was performed in 132 patients (66%). Surgical mortality was 5%. Comparison of survival of patients with tracheal tumors who are alive and well without the disease with those who died with carcinoma supports surgical treatment (9).

In another study, Gaissert et al. concluded that laryngotracheal resection and immediate reconstruction for subglottic ACC can be done with good preservation of voice, low morbidity and no compromise of long-term survival (11).

These tumors are more prevalent in trachea than the main-stem bronchus and spread along both submucosal and perineural planes. This pathologic feature accounts for an increased likelihood of positive microscopic surgical margin at the time of tracheal resection. Regional lymph node metastasis is reported in 10% of patients and remote metastasis to lungs, bones, and brain has also been reported. Despite these malignant features, ACC often follows a prolonged course. A slow and insidious

progression, often over several years, is a characteristic of non-treated cases (6).

In a review of 38 patients with ACC of the upper airway, Maziak et al. noted that lymphatic metastases were relatively uncommon and hematogenous metastases occurred in 17 out of 38 patients; pulmonary metastases occurred in 13 patients (1).

In our study, none of the patients showed signs of distant metastasis in the follow-up period. Local recurrence occurred only in 1 patient. Absence of distant metastasis in our patients might be due to the short follow-up period.

Pearson et al. concluded that in patients with ACC of trachea resection may offer excellent and long-term palliation even when the resection was incomplete or pulmonary metastasis was present (12).

Grillo et al. in his study compared patients who underwent resection therapy with those who underwent irradiation alone and reported that significantly better results were achieved by resection and post operative irradiation (9).

Regnard et al. in his study reported that post-operative radiotherapy increased survival. In case of incomplete resection long-term prognosis was worsened by the occurrence of pulmonary metastases in patients with ACC (13). In patients who do not meet the criteria for resection, radiation therapy can be an alternative.

Maziak et al. reported 6 patients with ACC of trachea who underwent irradiation as the primary therapy with a mean survival of 6.2 years (1).

Muller et al. reported a case of ACC of the distal trachea with involvement of the carina and both main stem bronchi cured with total dose of 66 Gy radiation in 2-Gy fractions over a week. Clinically and histologically complete remission was achieved without evidence of disease during 27-month follow-up (14).

In our study radiotherapy performed as adjuvant therapy in patients whom underwent tracheal resection with positive microscopic involvement of the surgical margins.

Because of the nature of adenoid cystic carcinoma of the trachea, surgical resection is the best method of treatment. But if surgical margins are positive, post-operative radiotherapy will be necessary. In patients who are not candidates for resection, radiotherapy can be an effective alternative.

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