Anaplastic Thyroid Carcinoma Encasing the Left Common Carotid Artery: A Case Report

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Abstract

Background: Anaplastic thyroid cancer is a rare but lethal type of thyroid cancer.

Case presentation: Herein, we report a 69-year-old male who was diagnosed with anaplastic thyroid cancer. The patient received neoadjuvant radiation therapy because the mass encased the left common carotid artery and infiltrated the left jugular vein. The tumor decreased in size; thus, surgery was performed. The patient underwent a total thyroidectomy with en bloc excision of the neoplasm and the carotid artery, ligation of the left jugular vein, and excision of the lymph nodes. The placement of a PTFE graft restored the continuity of the left common carotid artery.

Conclusion: The prognosis of anaplastic thyroid cancer is very poor, with a reported 5-year overall survival of 7%. It is managed with a combination of surgery, chemotherapy, and radiotherapy; however, its prognosis is still dire.

Keywords: anaplastic thyroid cancer; BRAF mutation; radiation therapy; chemotherapy; prognosis.

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I. Introduction

Anaplastic thyroid carcinoma is the rarest form of thyroid cancer, but it is the deadliest (1). It comprises less than 2% of all thyroid cancers. Nevertheless, it is responsible for up to 40% of thyroid cancer deaths (2). The median survival is 5-12 months, and the 1-year survival rate ranges between 20% and 40% (3).

All cases of anaplastic thyroid cancer are considered stage IV (4). The discovery of effective therapy for tumors harboring a BRAF V600E mutation has changed the treatment (5). If the tumor is deemed resectable and there is no evidence of distant metastasis, surgery is strongly recommended. Radiation therapy with or without chemotherapy should be administered in these patients postoperatively. Neoadjuvant radiotherapy and/or chemotherapy should be considered if the tumour is initially unresectable due to local extension. If the tumor becomes resectable, surgery is recommended. Unresectable locally advanced tumors or metastatic tumors should be treated with radiotherapy and/or chemotherapy (6).

Herein we describe the case of a 69-year-old patient who was diagnosed with anaplastic thyroid cancer that invades the left common carotid artery and infiltrates the left jugular vein. The patient received neoadjuvant radiation and displayed a good response. Total thyroidectomy with en bloc excision of the carotid and placement of a PTFE graft to restore the continuity of the carotid artery was performed.

II. Case Presentation

A 69-year-old man visited the emergency department with a rapidly growing neck mass associated with hoarseness. Physical examination reveals a firm, fixed mass on the left neck and enlarged lymph nodes. Computed tomography imaging showed a large heterogeneous left thyroid mass with tracheal deviation and lymphadenopathy but no distant metastases. The mass encases the left common carotid artery and infiltrates the left jugular vein. Fine-needle aspiration was consistent with anaplastic thyroid cancer in the primary tumor. The

patient received neoadjuvant radiotherapy (one cycle) and was reevaluated. The mass decreased in size, and therefore surgery was decided as the next step of the treatment plan.

A total thyroidectomy was performed with a cervical incision, lymph nodes' excision, and vagus nerves' recognition at both sides. During the operation, it was not possible to detach the tumor from the enclosed carotid artery. Consequently, an en bloc excision of the neoplasm and the carotid artery and placement of a PTFE graft to restore the continuity of the carotid artery was carried out. The infiltrating left jugular vein was ligated. The patient recovered uneventfully. On the fifth postoperative day, he was transferred to the department of oncology for further treatment.

III. Discussion

Anaplastic thyroid cancer (ATC) is a form of thyroid cancer. It is also known as undifferentiated thyroid cancer as its cells do not look and do not behave like typical thyroid cells. It is the fastest growing and most aggressive cancer of the thyroid gland (1). Fortunately, ATC constitutes less than 2% of all thyroid cancers. Nonetheless, it is responsible for up to 40% of thyroid cancer deaths (2, 7). It carries the worst prognosis among thyroid cancers, with a median survival rate of 5-12 months and a 1-year survival rate of 20-40% (3, 8). In the United States, the reported incidence is slightly more than 1000 new cases annually. Worldwide frequency likely approximates that in the United States (9). ATC usually affects people during the sixth to seventh decades of life with a female predominance (male to female ratio 3:1) (1). It usually occurs in patients with a history of thyroid pathology like preexisting goiter, follicular thyroid cancer, papillary thyroid cancer. Almost 20% of patients with ATC display differentiated thyroid cancer in their thyroid glands (5, 9).

Typically, patients report a rapidly growing neck mass appearing, associated with progressive local symptoms such as dysphagia, dyspnea, cough, neck pain, and infiltration of large neck vessels (5). History and clinical examination are the first steps in the initial evaluation of patients. Physical examination reveals a dominant neck mass, while 40% of the patients have lymph node enlargement (3). There is no time to lose, and diagnosis must be urgent. Imaging modalities such as thyroid ultrasound, computed tomography, magnetic resonance imaging, and positron emission tomography scan (PET scan) are fundamental to determine whether the tumor is resectable or not. Fine-needle aspiration (FNA) often provides enough cytologic information to allow diagnosis (10).

All cases of ATC are considered stage IV, reflecting the poor prognosis for people with this type of cancer. Stage IV is divided into IVA when ATC is confined to the thyroid, IVB when cancer has spread to nearby lymph nodes, strap muscles around the thyroid, or nearby tissues of the neck such as the larynx, trachea, esophagus, nerve to the larynx or back toward the spine, and large blood vessels, and IVC when there are distant metastases (4). At the time of diagnosis, almost 50% of the patients present with metastatic disease, 40% with extrathyroidal extension or lymph node involvement, and only 10% with intrathyroidal participation (3). The most typical sites of distant metastases encompass, in descending order, the lung, bone, and brain (11).

Treatment of ATC has changed dramatically during the last decade due to improved outcomes with the discovery of effective therapy for tumors harboring a BRAF V600E mutation. The initial management of ATC is comprised of four steps:

- 1. Assessment of the stability of the airway
- 2. Extensive metastatic workup
- 3. Evaluation of the resectability of the primary tumor
- 4. A rapid test to determine whether the tumor harbors a BRAF V600E mutation

The preferred approach for ATC should be the complete resection of the mass. The resectability is determined by assessing both the tumor burden and the extent of invasion of the structures involved. Surgery, radiotherapy, and chemotherapy are commonly used in ATC. Intensity-modulated radiation therapy (IMRT) is the standard of radiotherapy as it limits the damage to the surrounding normal structures. Chemotherapy encompasses the use of either conventional chemotherapy with a combination of taxanes, anthracyclines, and platinum-based cytotoxic agents or combined BRAF/MEK inhibitors. Conventional chemotherapy displays no clear evidence for improvement in quality of life or survival. The reported response rate is suboptimal, typically less than 15% (5, 12, 13).

For patients with the confined disease (stage IVA and IVB) in whom R0 (negative microscopic margins) or R1 (positive microscopic margins) is expected, surgical resection is strongly recommended. Stage IVA tumors should be treated by total thyroidectomy. Stage IVB tumors total thyroidectomy with prophylactic/therapeutic central and lateral neck lymph node compartments should be carried out. Unfortunately, ATC might invade vessels, nerves, muscles, esophagus, trachea, and larynx. Therefore, complete resection without sacrificing vital structures can be performed only in well-selected patients. Postoperatively, IMRT with concurrent systemic therapy should be offered in these patients. If a stage IVB tumor is deemed unresectable, neoadjuvant radiotherapy and/or chemotherapy should be considered. If cancer becomes

resectable, surgical resection should be reconsidered. For patients with unresectable locally advanced tumors or metastatic disease, the treatment options include radiotherapy and/or chemotherapy (5, 6).

Even though it is the rarest type of thyroid cancer, ATC is the deadliest, with a poor prognosis (5). The reported 5-year survival rate is 7% regardless of the stage. The 5-year survival rate based on the stage is as follow: stage IVA 31%, stage IVB 10%, and stage IVC 3% (14). Factors demonstrating worse prognosis encompass age older than 70 years, acute symptoms (duration of severe complaints such as dysphonia, dysphagia, dyspnea, and rapid growth of the tumor < 1 month), leukocytosis (white blood cell count $\geq 10,000/\text{mm3}$, tumor size >5 cm, and distant metastasis (15).

IV. Conclusion

Anaplastic thyroid cancer is a rare but fatal type of thyroid cancer. Patients with a rapidly growing neck mass require immediate investigation. Initial management encompasses assessing the stability of the airway, an extensive metastatic workup, the evaluation of resectability of the primary tumor, and a rapid test to determine whether the tumor harbors a BRAF V600E mutation. Resectable tumors with no evidence of metastatic disease should be removed. Unresectable locally advanced tumors or metastatic tumors should be treated with radiotherapy and/or chemotherapy. The reported 5-year survival rate ranges between 3-31%, indicating the poor prognosis of such tumors.

Abbreviations:

ATC: Anaplastic Thyroid Cancer

IMRT: intensity-modulated radiation therapy

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