Anaplastic thyroid carcinoma, a report on 10 cases

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Abstract

Anaplastic thyroid carcinomas are undifferentiated and highly aggressive tumors of the thyroid follicular epithelium, with a disease-specific mortality approaching 100 percent. This study reports ten cases with advanced anaplastic thyroid carcinoma with ineffective surgical interventions. Between 2001-2006, we treated 10 patients with anaplastic carcinoma of the thyroid. All patients presented with advanced phase and were documented by pathological study. The surgical intervention was thyroidectomy in 3 cases, debunking surgery in other three, just tracheostomy in three cases, and only biopsy in one patient. All of our patients died in 4 days to 8 months after surgery. Surgical intervention had no benefit in treatment of advanced anaplastic carcinoma of the thyroid and the suitable surgical intervention was to maintain an open airway. Early recognition of the disease is essential to allow prompt initiation of therapy. There is no cure for advanced situations, and surgery, radiotherapy, and chemotherapy play no important role in the treatment.

Keywords: Anaplastic thyroid carcinoma; Surgical intervention; Cancer

Introduction

Cancer of the thyroid gland is an uncommon tumor and account for about %1 of all cancers in USA; and approximately 92% of thyroid cancers (80% papillary, 10% follicular, and 2% Hurthle cell carcinomas) have a good to excellent prognosis. The remaining 8% of thyroid malignancies including anaplastic (undifferentiated) carcinomas, medullary thyroid carcinomas, thyroid lymphoma, and metastatic tumors having dismal prognosis. Most anaplastic carcinomas of the thyroid appear to arise in patients with nodular goiters or differentiated carcinomas of the thyroid. Some anaplastic cancers present with cervical lymphadenopathy or lung, bone, liver or brain metastases.1-5

Nearly all patients with anaplastic carcinoma exhibit a thyroid mass. However, regional or distant spread is present at the time of initial diagnosis in 90% of cases.3,9 Distant metastases are found at initial disease presentation in 15-50% of patients.1-10 The lungs are the most common site of distant metastasis and involve up to 90% of patients with distant disease.10,11 The diagnosis of anaplastic carcinoma is usually established by cytological examination of the materials obtained by fine-needle aspiration biopsy or of tissue obtained by large needle or surgical biopsies. Diagnostic imaging is useful for defining the extent of disease, treatment planning, and monitoring the response to treatment. Computed tumography (CT) of the neck and mediastinum can accurately delineate the extent of the thyroid tumor and identify tumor invasion of the great vessels and upper aerodigestive tract.12 Sonography of the neck also can accurately identify the size of tumor and detect extra thyroidal invasion and the involvement of local and regional lymph nodes. In addition, both sonography and CT can help to guide fine-needle aspiration of solid, non-necrotic portions of the tumor for diagnosis.13,14 Anaplastic cancer of the thyroid is too difficult to treat, because most patients seek medical advise when the tumors are in advanced stage.2-4 However, if mass is confined only to thyroid, surgery with adjuvant therapy will be moderately beneficial. Relatively favor-
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Prognostic factors include unilateral tumor, diameter of the mass less than 5 cm, and absence of extrathyroidal invasion, without involvement of cervical lymph nodes. Thyroid lobectomy involving wide margin of adjacent soft tissue on the side of tumor is virtually an aggressive surgical approach. There is no effective therapy for advanced or metastatic anaplastic thyroid carcinoma and the disease is uniformly fatal. The average survival time from diagnosis ranges from three to six months. Death is usually attributable to upper airway obstruction.

Materials and Methods

In a retrospective study performed between 2001-2006, and based on clinical records, we identified ten patients with anaplastic thyroid carcinoma admitted to our hospital. In all cases, the diagnosis was confirmed by pathological findings.

Results

We treated 10 patients for anaplastic carcinoma of the thyroid (Table 1). Seven patients were female with an age range of 32-71 years and a mean of 57 years. The three remaining male patients aged from 62 to 71 years and their mean age was 66 years. Of three patients who underwent previous thyroid surgery, two had papillary thyroid carcinoma and one nodular goiter. FNA was performed on all patients and anaplastic carcinoma of the thyroid was documented in 9 patients. One patient (32 years old female) exhibited suspected cells in FNA.

Table 1: Characteristics of the patients under study

<table>
<thead>
<tr>
<th>Patients</th>
<th>Sex</th>
<th>Age</th>
<th>Previous thyroid pathology</th>
<th>Size of tumor</th>
<th>Distant metastasis</th>
<th>Treatment</th>
<th>Survival</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>71</td>
<td>-</td>
<td>10x9</td>
<td>No</td>
<td>Trach</td>
<td>4 days</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>67</td>
<td>Tx, Papillary carcinoma</td>
<td>4x3</td>
<td>No</td>
<td>Tx</td>
<td>1 month</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>62</td>
<td>-</td>
<td>8x6</td>
<td>No</td>
<td>Debulking</td>
<td>1 month</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>65</td>
<td>-</td>
<td>5x5</td>
<td>No</td>
<td>Trach</td>
<td>7 days</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>57</td>
<td>-</td>
<td>8x5</td>
<td>No</td>
<td>Debulking</td>
<td>20 days</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>55</td>
<td>Tx, Papillary carcinoma</td>
<td>7x4</td>
<td>No</td>
<td>Debulking</td>
<td>1.5 months</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>32</td>
<td>-</td>
<td>6x5</td>
<td>No</td>
<td>Tx</td>
<td>3 weeks</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>71</td>
<td>-</td>
<td>7x7</td>
<td>Lungs</td>
<td>Biopsy</td>
<td>1.5 months</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>70</td>
<td>Tx, Nodular goiter</td>
<td>6x4</td>
<td>No</td>
<td>Tx,Rx</td>
<td>8 months</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>45</td>
<td>Goiter</td>
<td>8x6</td>
<td>No</td>
<td>Trach</td>
<td>11 days</td>
</tr>
</tbody>
</table>

Debulking=Debulking surgery; Tx=Thyroidectomy; Trach=Tracheostomy; Rx=radiotherapy

Discussion

The age-adjusted annual incidence of anaplastic carcinoma is about two per million persons. Patients with anaplastic carcinoma of the thyroid are older than those with differentiated carcinoma of the thyroid; the mean age at diagnosis is 65 years and fewer than 10% of cases are younger than 50 years. In our study, except two patients who were younger than 50 years, most subjects were of older age. Sixty to 70
percent of tumors are known to occur in women; and in our study, 7 patients were female. Approximately 20% of patients with anaplastic thyroid cancer have a history of differentiated thyroid cancer. In our report, two patients had a previous operation of combined lobectomy and isthmectomy for papillary carcinoma with 20-30% having a coexistent differentiated carcinoma; the percentage may be even higher with extensive sectioning of the thyroid gland. The majority of synchronous thyroid tumor are papillary carcinoma, but coexisting follicular carcinoma have also been reported. Nearly 10% of patients with hurthle-cell carcinoma have foci of anaplastic carcinoma. Transformation from differentiated to anaplastic carcinoma has been described in patients followed through serial biopsies of the thyroid. These findings lend support to the hypothesis that anaplastic carcinoma develops from more differentiated tumors arising from one or moreDifferentiating events. The primary symptom of anaplastic carcinoma is a rapidly enlarging neck mass, occurring in about 85% of patients. The enlarging thyroid tumor may cause neck pain and tenderness and compression (or invasion) of the upper aero digestive tract, resulting in dysphasia (%30), hoarseness (%25), cough (sometimes hemoptysis, 25%). Less common symptoms are chest pain, bone ache, headache, confusion, or abdominal pain due to metastases. All of our patients presented with rapidly enlarging cervical masses which were grossly visible and hard on palpation. One patient presented with clinical features of subacute thyroiditis. The thyroid function tests were within normal limits. Rarely, rapid growth of the tumor may destroy the thyroid gland, by causing thyroiditis resulting in thyrotoxicosis and severe neck pain and tenderness.

On physical examination, most patients have bilateral albeit asymmetric thyroid enlargement and a dominant nodule is often present. Some nodule may be softer and fluctuant, indicating focal tumor necrosis. A few patients have solitary nodule or nodular goiter. The goiter is often fixed to the surrounding structure and does not move with swallowing. By the time of presentation, the primary tumor is usually greater than 5 cm in diameter, but precise measurement are often difficult to achieve, because of indistinct borders of the tumor. An estimated 50% of patients have enlarged cervical lymph node. None of our patients had any documented cervical lymph node enlargement. Other findings of local extension of the disease include stridor, tracheal deviation and vocal cord paralysis due to compression or invasion of the trachea, and venous dilatation and superior vena cava syndrome due to retrosternal tumor growth. The skin overlying the tumor may be erythematous, or even ulcerated. In our study, one case presented with skin inflammation and one patient exhibited skin ulcer over the neck mass. There may be occasional metastasis in the skin of the chest and abdomen. Two patients had lung metastasis, focal neurological symptoms or signs suggestive of brain metastasis. The diagnosis of anaplastic carcinoma is usually established by cytological examination of cells obtained by fine-needle or tissue obtained by large needle or surgical biopsy.

Prognostic factor is related to extent of the disease, and presence or absence of local and regional metastases. Tumor size also appears to be important. In one study, the two years survival was 25 and 3 to 15% in patients with tumor less and larger than 6 cm respectively. Variables that may predict a worse prognosis include older age at diagnosis, maleness and dyspnea as a presenting symptom. Patients who were previously treated for differentiated carcinoma and subsequently developed anaplastic carcinoma had outcome similar to those without an antecedent thyroid cancer. Except in one patient, the survival time for all subjects under study was less than two months, a period too short to allow chemotherapy or radiotherapy. The longest surviving patient who underwent radiotherapy and chemotherapy after thyroidectomy, died after 8 months due to airway obstruction.

In conclusion, anaplastic carcinoma of thyroid is an aggressive cancer which leads to imminent death. Only those patients, whose pathology is confined to thyroid with small tumor size, can benefit from treatment but can have a short survival time. Total thyroidectomy followed by radiotherapy and chemotherapy is mostly indicated to prolong patient’s survival whenever tumor size is small, otherwise any surgical treatment other than those for opening the airway, would deteriorate the patient’s condition.
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References