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An Unusual Case of Thyroid Hurtle Cell Carcinoma with Direct Extension to the Right Brachiocephalic Vein, Right Auricle, and Right Atrium: Case Report

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ABSTRACT

The clinical behavior of various types of thryroid tumor have been much studied during the past several decades, and the histologic features, surgical management, and prognostic factors of follicular and papillary tumors in particular have been clarified to a considerable degree. On the other hand, there is still controversy concerning management of Hurtle cell tumor (HCT) of the thyroid. HCT is not a common disease, making it impossible to obtain sufficient clinical data at a single institution. It has been reported that all HCT are aggressive and should be treated as malignant tumors. It is believed that an accurate differential diagnosis can be made between cancer and adenoma on the basis of pathological studies. We describe a patient with HCT of the thyroid extending into the right atrium. To our knowledge, after a Medline search, this is the first such case in the medical literature. The interesting features are described, and the relevant literature is briefly reviewed.

INTRODUCTION

Specific thyroid tumor with Hurtle cells represents a rare clinical form of thyroid neoplasia and has a particular biological behavior. Tumors included in this category are those in which more than half of the cell population is made up of Hurtle cells. We present a case of a 69-year-old man with extension of a thyroid Hurtle cell carcinoma to the right atrial auricle and the right atrium via great cephalic vessels, and successful treatment with surgical excision during cardiopulmonary bypass. Pathological examination using hematoxylin-eosin, van Gieson, and argyrophilic nucleolar organizer region stains, positive immunoreaction for thyroglobulin labeled streptavidin avidin biotin, and ultrastructural examination established the definitive diagnosis. Surgical treatment is the main therapeutic measure and must be performed under oncological curative principles, with total thyroidectomy for malignant tumors. The frequency of thyroid carcinoma metastasis to the heart is very low, and the absence of early symptoms makes the clinical diagnosis difficult.

CASE REPORT

A 69-year-old man was admitted to the Hacettepe University Department of Otorhinolaryngology because of a mass on the right side of his neck and weight loss. Results of advanced examinations showed that the mass originated from the thyroid gland. Laboratory results revealed normal thyroid hormone levels. Incisional biopsy of the mass for pathologic diagnosis was performed under local anesthesia, because the mass was fixed to the surrounding structures and overlying skin. Pathological examination was consistent with Hurtle cell tumor (HCT) of the thyroid, so total thyroidectomy and radical neck dissection were performed. Macroscopically there was an infiltrative tumor with replacement of the right lobe of the thyroid. The tumor was tan to pink on the cut surface and had small foci of hemorrhage. On histologic examination the tumor was found to be composed of anaplastic cells with abundant granular eosinophilic cytoplasm and round to oval nuclei. There was an infiltration to the adjacent thyroid parenchyma and extension beyond the capsule as well as vascular invasion (Figure 1). Immunohistochemically the tumor cells were positive for thyroglobulin (Figure 2). Histopathologic diagnosis was confirmed as Hurtle cell thyroid carcinoma. Whole-body computerized tomography (CT) was performed and unexpectedly revealed a tumor thrombus that filled the superior vena cava and the right atrial auricle (Figure 3). The patient's thyroglobulin hormone level was high (>300 ng/mL) (normal range is 0-55 ng/mL), and the antithyroglobulin antibody was negative, although his thyroid-stimulating hormone level was normal. A tumoral mass and cavo-atrial thrombus were identified in the right atrium and superior vena cava by means of transthoracic echocardiography. This mass was accepted as a primary tumor extension into intracardiac cavities via great cervical and cardiac veins. The patient had no symptoms related to tumoral extension, such as superior vena cava syndrome or

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Figure 1. Infiltration of the tumor to the adjacent thyroid parenchyma (hematoxylin-eosin, original magnification × 115).

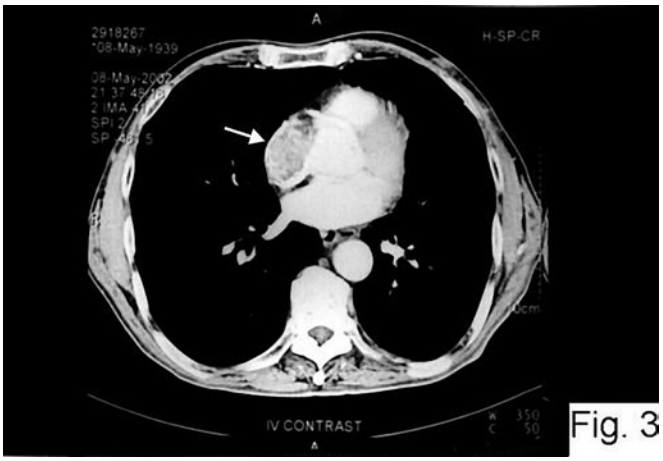

Figure 3. Extension of the tumor thrombus into the right atrium, visible on the thoracal computed tomographic examination.

cardiac arrhythmia, although the tumor filled the superior vena cava and the right atrial auricle.

During the operation a standard midline sternotomy was performed. The right subclavian vein, superior vena cava, and right atrium were filled with the tumor thrombus, but innominate vein invasion was absent. Standard aortic and selective venous cannulation were performed from the inferior vena cava and the innominate vein. Cardiopulmonary bypass (CPB) was instituted. After systemic and topical hypothermia, antegrade crystalloid cardioplegic solution was administered. Following right atriotomy, a superior vena cavotomy was made 1 cm away from the right atrial junction. The thrombus formation, which was approximately 4 cm in diameter, grey-white in color, quite soft, gelatinous, and adhesive to the atrium wall, without a pedicle, was seen (Figure 4). Thrombus formation extended to the superior vena cava and the right subclavian vein. The tumor thrombus was removed from the superior vena cava and the right atrial auricle, and a Fogarthy embolectomy catheter was introduced to the subclavian vein to remove the remaining tumor thrombus. The patient was weaned from CPB. Physical examination revealed no problems in the patient during the postoperative period. Thyroid hormone levels were low following the early postoperative period. The histopathologic diagnosis of the tumor thrombus was Hurtle cell carcinoma (Figure 5). The patient was discharged on the 10th postoperative day, and radioactive I^{131} treatment was applied after surgery. Physical examination results 2 years postoperatively were normal.

DISCUSSION

The forms of malignant disease that most frequently metastasize to the heart are malignant melanoma, carcinoma of the lung, lymphoma, and carcinoma of the breast [Abraham 1990], but almost all types of malignant tumors have been found to metastasize to the heart. Hematogenous and lymphatic spread are considered to be mechanisms responsi-

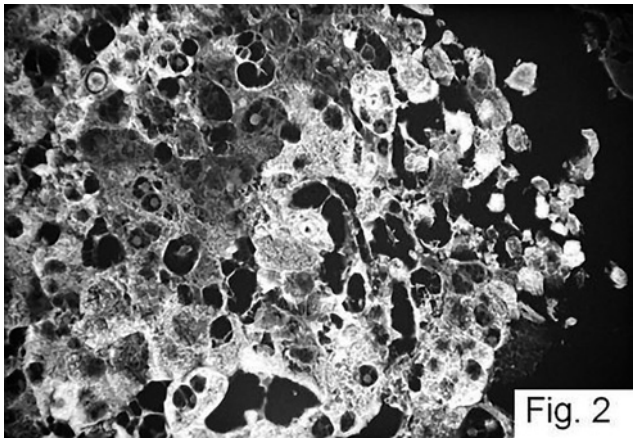

Figure 2. Thyroglobulin positivity in the cytoplasm (immunohistochemistry, original magnification × 460).

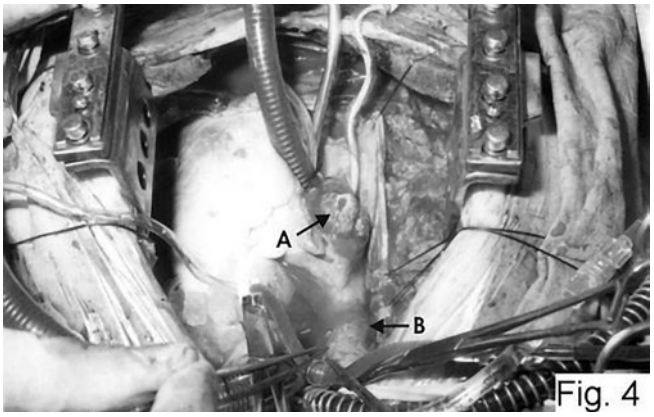

Figure 4. A, Tumor thrombus in the right atrium and right auricle. B, Tumor thrombus fills the whole of the superior vena cava.

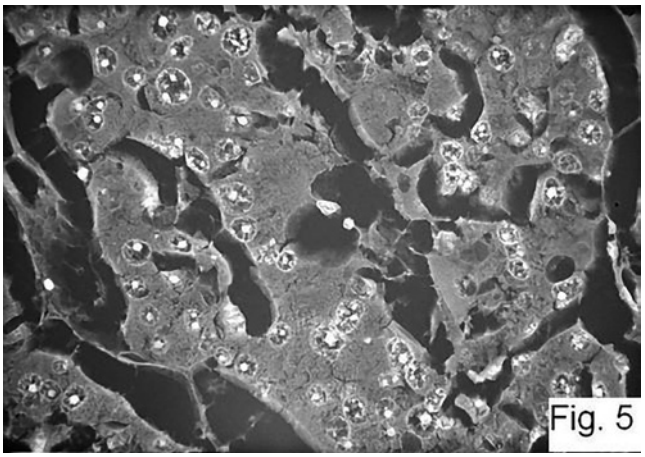

Figure 5. Anaplastic cells with abundant granular eosinophilic cytoplasm are visible in the tumor thrombus removed from the superior vena cava and the right auricle (hematoxylin-eosin, original magnification × 460).

ble for myocardial and pericardial tumor involvement. Endocardial metastasis can also result from extension of the tumor through the vena cava. Cardiac metastases from malignant thyroid carcinoma are uncommon. A case of intracardiac metastasis of thyroid cancer has been reported in the literature [Kim 2000]. The reported incidence of cardiac metastasis from thyroid carcinoma varies from 0% to 2% among autopsy series [Abraham 1990]. The incidence of cardiac metastasis by all carcinomas cited in necropsy studies varies from 3.4% to 19.1% [Prichard 1951]. The access to the heart is most commonly embolic, via the coronary arteries, inferior or superior vena cava, or other small veins [Scott 1939, Lefkovits 1948].

Generally, cardiac metastases of many types of neoplasms are diagnosed on autopsy or coincidently during antemortem investigations looking for distant metastases. Surgical treatment of cardiac metastases has been recommended. Shindo et al [1983] recommend open-heart surgery in case of intracavitary tumors both primary and metastatic to rescue emergency cases with circulatory obstruction secondary to the tumor and also in cases in which surgery will enhance the effects of radiation or chemotherapy. Treatment of cardiac metastasis with systemic chemotherapy and radiation therapy is of limited benefit. Surgery is performed to make tissue diagnosis and alleviate the cardiac symptoms (such as pericardial tamponade and coronary emboli). It is believed that an accurate differential diagnosis can be made between cancer and adenoma on the basis of pathological studies.

In this report a case of thyroid Hurtle cell carcinoma is described in which there was direct extension of the tumor into the thyroid veins, jugular veins, superior vena cava, and right atrial auricle. Interestingly our patient had no cardiac symptoms, although the tumor had filled the whole lumen of the superior vena cava. Cardiac metastasis of this tumor was found incidentally on the whole-body CT scan and transthoracic echocardiography. A scintigraphic examination performed during the preoperative period showed no distant tumor metastasis. On the thoracal CT examination for the investigation of distant metastasis of the tumor, a mass filling the lumen of superior vena cava and the right auricle was found. Cardiac examination of this patient revealed no abnormalities, and none were reported based on electrocardiography results. A right atrial thrombus was revealed in the patient's transthoracic echocardiography.

Although a great deal has been reported about the pathologic diagnosis and surgical management of HCT, controversy still exists and is attributable to the fact that HCT is a rather uncommon tumor, so there is little experience with the disease at any single institution. HCT is sometimes grouped with follicular tumors. It is generally regarded as behaving aggressively, and the differential diagnosis between adenoma and cancer is difficult, as it is for ordinary follicular thyroid tumors. Partial lobectomy was often employed to treat HCT in the past. Recently, total lobectomy has been performed as initial surgery for all HCT. If the pathologic diagnosis is cancer, total thyroidectomy is usually performed.

Total thyroidectomy for all HCT has been advocated because recurrence occurred in 3 of 26 patients with histologically benign adenomas, and recurrence can be lethal. However, several studies have shown that HCT is not particularly aggressive, and conventional pathology can distinguish adenoma from cancer. Bondeson et al [1983] and Gosain and Clark [1984] advocated total lobectomy for HCT and recommended completion of total thyroidectomy only if the postoperative pathologic diagnosis is cancer.

Several groups have reported the clinical features of cancer and adenoma and the differences between them. It has been reported that the size of the primary tumor had predictive value as a criterion for malignancy. Carcangiu et al [2003] reported that patients with cancer had larger sized primary tumors.

HCT has occurred more often in women than men, but the cancer ratio was higher in men. Preoperative diagnosis of cancer is not easy, as for ordinary follicular tumors. Hurtle cells are often observed in hyperplastic nodular lesions, and the differential diagnosis by fine-needle aspiration biopsy cytology (FNABC) between adenomatous goiter and HCT is sometimes difficult. Although several studies have shown that HCT can be reliably diagnosed by FNABC, the differential diagnosis between adenoma and cancer cannot be made by FNABC. Some have reported that intraoperative frozen section analysis is a useful diagnostic tool for the delineation between adenoma and cancer, whereas others have reported that it is not reliable. In the present study we performed FNABC.

Total thyroidectomy facilitates the use of radioiodine and thyroglobin as markers for recurrence [Sugino 2001]; however, Hurtle cell cancer does not accumulate radioiodine, and total thyroidectomy puts the recurrent laryngeal nerve and parathyroid glands at risk to some extent. Thus total thyroidectomy for all HCT at initial surgery is unnecessary. Hemithyroidectomy for all HCT at initial surgery is the treatment of choice, and when the postoperative pathologic diagnosis is HCT, completion of total thyroidectomy should be considered if the patient is at high risk. But in cases such as ours, with wide intravascular involvement, total thy-

roidectomy and postoperative radioactive iodine treatment is preferred.

Several authors have reported tachycardia, atrial fibrillation, and flutter as common findings in cardiac metastasis of tumors [Lefkovits 1948]. Most authors advise that if patients with widely metastatic tumors manifest new cardiac abnormalities such as those cited, there should be suspicion of cardiac involvement of the tumor. A chest x-ray showing an increase in the cardiac silhouette suggests pericardial effusion or cardiac failure secondary to tumor involvement. In this case chest x-ray results were normal. As is shown here, 2-dimensional echocardiography is a well-established tool to aid in the diagnosis of endocardial, myocardial, or pericardial tumor involvement [Johnson 1983]. Other diagnostic tests such as computerized axial tomography with contrast material and magnetic resonance imaging are also helpful in diagnosis and evaluation of cardiac metastases. The diagnosis can be made with I^{131} scanning, and therapy with this isotope is certainly indicated. The T4 hormone as supplementation and suppressive therapy should be implemented. Two-dimensional echocardiography and thoracal CT scan with contrast material may be helpful, and thyroglobulin to be useful as a marker for recurrence [Carcangiu 2003].

In conclusion, thyroid Hurtle cell carcinoma can be an aggressive cancer with a dismal prognosis and should be taken into consideration as a source of cardiac metastasis and a cause of sudden cardiac death. If the patient has a diagnosis of Hurtle cell carcinoma, transthoracic echocardiography or CT may considered for routine use as noninvasive and cost-effective methods for the detection of the cardiac metastasis.

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