



Case Report

Sarcomatoid carcinoma of the thyroid: Report of a subtype of anaplastic thyroid carcinoma

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ABSTRACT

Thyroid sarcomatoid carcinoma is a rare malignancy. It manifests as a rapidly growing neck mass in the elderly. The histologic findings are characterized by an admixture of anaplastic epithelial and mesenchymal components. It can be easily confused with other thyroid malignancies, such as sarcoma, lymphoma, and medullary carcinoma, both clinically and cytopathologically. Immunohistochemical stain studies are essential to confirm the diagnosis. The prognosis is poor and survival beyond 1 year is very rare. We report a man aged 79 years with a rapidly growing large thyroid mass.

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1. Introduction

The majority of sarcomatoid carcinomas of the thyroid are anaplastic carcinomas. Anaplastic thyroid carcinomas are divided into three histologic subtypes according to the predominant cell type: spindle cell, giant cell, and squamoid. Sarcomatoid carcinoma resembles spindle cells, which are sarcoma-like, and characterized by an admixture of anaplastic epithelial and mesenchymal components. Immunohistochemical stain (IHS) studies are essential to confirm the diagnosis. As with all anaplastic thyroid carcinomas, sarcomatoid carcinoma is extremely aggressive and tends to invade the surrounding tissues, similar to true sarcoma.

2. Case report

A man who was 79 years of age and a Buddhist monk presented with shortness of breath and palpitations for 1 month. He had no evidence of cardiac or pulmonary dysfunction to account for his symptoms, but he had a notable goiter/mass particularly on the left side of his neck. He stated that he had the goiter for 3 years, but it had rapidly enlarged around the time his symptoms started.

On examination, there was a goose-egg-sized firm, fixed, oval mass superimposed on the enlarged left lobe of the thyroid. There was no palpable lymph node enlargement but trachea deviation to the right was evident.

Thyroid echography confirmed a large (7.7 × 7.0 × 5.8 cm) cystic lesion with heterogeneous intensity. Magnetic resonance imaging of the neck (Fig. 1) confirmed the mass that forced the trachea deviation.

The initial laboratory studies revealed thyroid-stimulating hormone 2.56 uIU/mL (normal range, 0.35–5.5 uIU/mL); free thyroxine 0.94 ng/dL (normal range, 0.89–1.81 ng/dL) and triiodothyronine 145.9 ng/dL (normal range, 60–189 ng/dL). Serum calcium was 2.17 mmol/L and phosphate was 0.9 mmol/L. Blood liver enzymes and creatinine were all normal as were the results of the complete blood count except for a hemoglobin of 10.6 mg/dL.

Fine needle aspiration cytology from the thyroid mass using a 21-G needle was performed. Papanicolaou stains were done. The aspirates revealed anisonucleosic follicular cells with hyperchromatic nuclei and spindle-shaped cells (Fig. 2). He underwent a total thyroidectomy. The pathology showed sarcomatoid-like spindle cell proliferation around the thyroid follicles or in a fascicular or storiform pattern. Prominent vascularization and tumor cells invading the venous wall were also noted (Figs. 3 and 4). The section from the left lymph nodes showed extrathyroid tumor extension in the soft tissue. The results of IHS revealed a diffuse cytokeratin and focal positivity with vimentin, but epithelial membrane antigen (EMA) and thyroglobulin were not detected.

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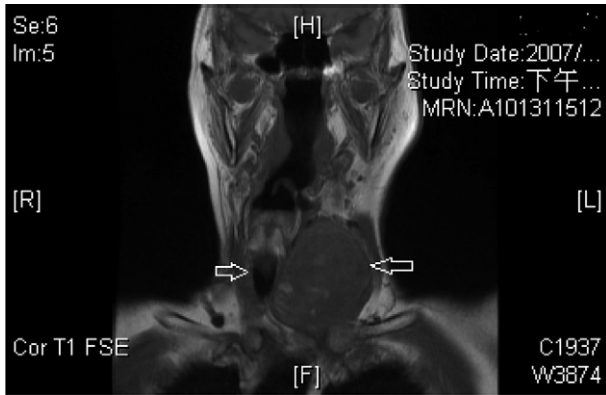


Fig. 1. Large thyroid tumor (\blacktriangleleft) causing trachea (\blacktriangleleft) deviation.

The patient did not receive radiotherapy or chemotherapy upon his request and died 1 year later.

3. Discussion

Anaplastic thyroid carcinoma (ATC) accounts for only 2%–5% of all thyroid cancers [1]. Microscopically, ATC is classified into two major categories [2]. The first is squamoid, resembling non-keratinizing squamous cell carcinoma, without follicles, papillae, or even trabeculae or nests. The second category is sarcomatoid, including spindle cells and giant cells, and may exhibit a fascicular or storiform pattern of growth, heavy neutrophilic infiltration, prominent vascularization and cartilaginous/osseous metaplasia. In the study by Us-Krasovec *et al* [3], the spindle cell population was the most rare and was seen in 7 of 83 patients (8%) with ATC. Cytologic evaluation from thyroid aspirates is very challenging in when differentiating spindle cell clusters from true sarcoma, lymphoma, and medullary carcinoma. However, sarcomatous thyroid carcinoma does not form follicles, papillae, or even trabeculae or nests, but the tumor still retains an unmistakable epithelial appearance on morphology and in the immunohistochemical staining pattern [4]. Immunohistochemically, the most useful marker is keratin, which is expressed in 40%–100% of cases [5]. Vimentin, carcinoembryonic antigen, and EMA may be diffusely or focally distributed. Thyroglobulin is undetectable in truly undifferentiated carcinoma, as in our case.

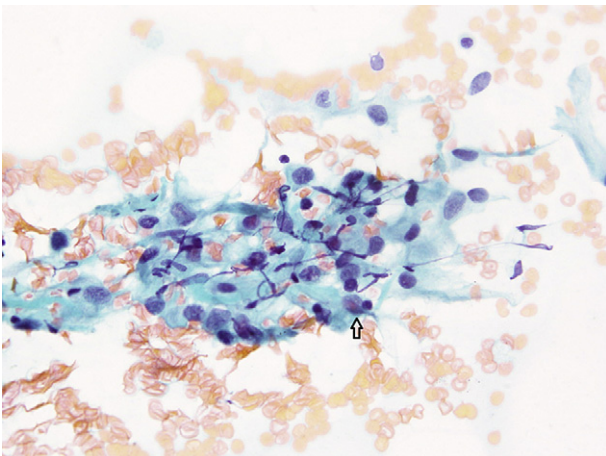


Fig. 2. Anisonucleotic follicular cells with hyperchromatic nuclei (\blacktriangleleft) and spindle-shaped cells.

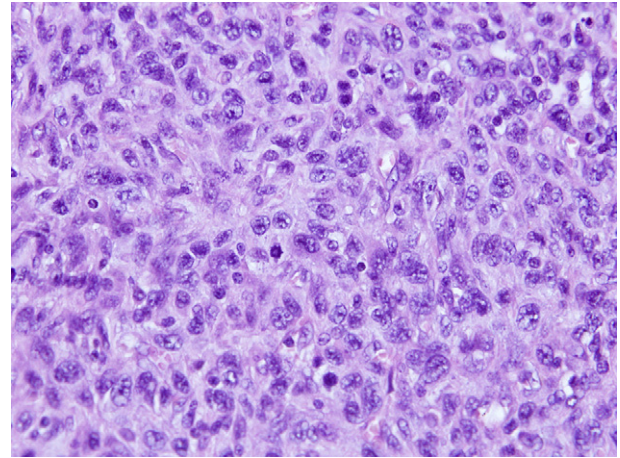


Fig. 3. Sarcomatoid-like spindle cell proliferation around the thyroid follicles or in a fascicular or storiform pattern.

Sarcomatoid thyroid carcinoma is a disease of the elderly and like other ATC typically occurs after age 60 years [6]. The characteristic clinical presentation is a rapidly growing firm mass on top of a long-standing goiter. Most patients have neck masses larger than 5 cm, often associated with one or more symptoms such as hoarseness, dyspnea, and dysphagia [7].

The mortality rate of sarcomatous thyroid malignancy is over 95%, and the mean survival is less than 6 months [8]. Patients with a large tumor (> 5 cm in diameter), acute symptoms (duration of complaints \leq 1 month), distant metastasis at presentation, and marked leukocytosis (white blood cells \geq 10,000/mm³) have a significantly shorter survival time than patients without these symptoms [7]. Cuboidal cell, small cell compact, and mixed cell anaplastic carcinoma of the thyroid may have a slightly better prognosis than giant spindle cell or small cell diffuse anaplastic carcinoma. Surgical debulking of the tumor mass to relieve local obstruction or compression of the vital organs combined with adjuvant hyperfractionated external beam radiation has been shown to improve survival even when distant disease is present [9]. Some authors believe that surgery, complete radiotherapy, and, particularly, doxorubicin-based chemotherapy offer the best possibility of survival [10,11]. However, the prognosis is poor and survival beyond 1 year is very rare due to local recurrent or metastatic disease and involvement of vital structures in the neck.

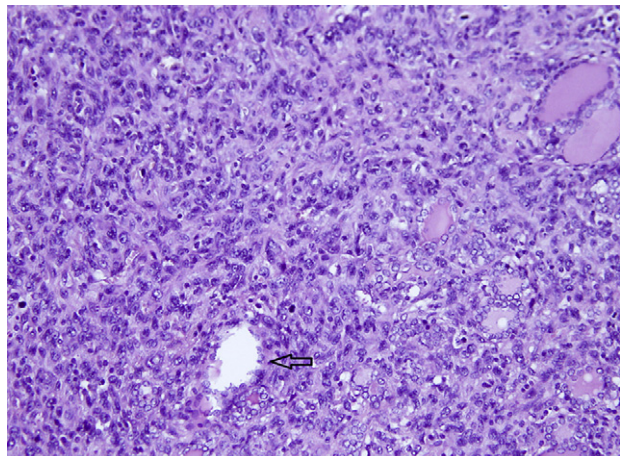


Fig. 4. Note the prominent vascularization and tumor cells invading the venous wall (\blacktriangleleft).

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