Case Report

Carcinoma of Thyroid with Thymus Like Differentiation: A Diagnostic Challenge on Fine-Needle Aspiration Cytology

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Abstract

Carcinoma of thyroid showing thymus like differentiation is a rare tumor arising from ectopic thymic or branchial pouch remnants. Very few papers describe the cytological features of this uncommon neoplasm. We report a case of 52-year-old woman who presented with swelling in front of the neck since 3 years. Fine-needle aspiration (FNA) showed cell rich smears composed of spindled to polygonal cells with eccentrically placed nuclei and orangophilic cytoplasm. A possibility of medullary carcinoma was suspected. Biopsy showed carcinoma thyroid with thymus like differentiation. Presence of pleomorphic, poorly differentiated cells with individual cell keratinization on FNA helps us to suspect this rare entity.

Keywords: Carcinoma showing thymus-like elements, Fine-needle aspiration cytology, Medullary carcinoma

INTRODUCTION

Carcinoma of the thyroid with thymus like differentiation is a rare tumor that arises from ectopic thymus or the branchial pouch remnant. This entity was first described by Miyauchi *et al.*[1] A decade later, Rosai and Chan used the term "carcinoma showing thymus-like elements (CASTLE)" while describing its morphological features.[2] CASTLE is considered to be a slow growing indolent tumor though it can involve lymph nodes.[3] This entity is categorized as an independent type of thyroid tumor in the WHO classification. Though it resembles squamous cell carcinoma (SCC), clinically has a good prognosis than SCC.[4]

CASE REPORT

A 52-year-old woman presented with a slow growing swelling in the front of the neck since 3 years, which moved with deglutition. She also complained of difficulty in breathing associated with dry intermittent cough since a year. No hoarseness/change in voice were noticed. There was no significant loss of weight/appetite. Contrast computed tomography showed a thyroid nodule with retro sternal extension and extension into the right trachea-esophageal grove with lateral displacement of carotid vessels. Vocal cords appeared normal.

Fine-needle aspiration cytology (FNAC) smears were cellular and composed of malignant spindle to polygonal cells arranged in dyscohesive syncytial fragments with eccentric nuclei, scant to moderate cytoplasm imparting a plasmacytoid appearance in a background of lymphocytes and few follicular cells (Figure 1). A possibility of medullary carcinoma was suggested. Total thyroidectomy was performed along with lymph node clearance.

Histopathology

Gross morphology

The right thyroid lobe showed a well-defined, lobulated greyish white to tancolored tumor measuring 4 cm × 2.5 cm, surrounded by a rim of residual thyroid tissue on one side (Figure 2).

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Microscopy showed a lobulated neoplasm composed of syncytial sheets and nests of tumor cells with indistinct cell borders, moderate cytoplasm and vesicular nuclei separated by fibrous septae containing focal lymphoplasmacytic infiltrate. Occasional tumor islands showed central foci of squamoid differentiation imparting a Hassall corpuscle-like appearance (Figure 3). On immunohistochemistry, tumor cells were positive for cytokeratin, CD117 and CD5 (Figure 3).

Treatment and follow-up

Patient was treated with external beam radiotherapy 50 GY for 1 month and was on regular follow-up for 3 years with no detectable recurrence or metastasis.

DISCUSSION

FNAC is a common modality of evaluation of thyroid lesions in routine clinical practice. Cytological features of various common thyroid neoplasms are fairly well-characterized. However, CASTLE is less often considered in the cytologic differential diagnoses owing to its rarity. Characteristic morphological features like lobulation are not evident on cytology. Smears can be mistaken for various neoplasms like medullary carcinoma, lymphoepithelioma or anaplastic carcinoma.[5] Features of CASTLE that can mimic medullary carcinoma include dyscohesive cells, eccentric tumor cell nuclei and presence of interspersed larger cells in a clean background. The serum calcitonin levels may not be available since FNA is usually performed as first-line workup as was seen in the present case. Possibility of lymphoepithelioma may be considered if numerous lymphoid cells are also noted along with along with the tumor cells while foci of squamous differentiation may cause confusion with metastatic SCC. CD5 immunohistochemistry can be used to confirm the diagnosis. It is a surface protein expressed in mature T-cells and is positive in 1 CASTLE, indicating thymic differentiation (Table 1).[5-8]

The various entities that may be considered on cytology is shown in a tabular form Youens *et al*.,[7] have reported the presence of pseudonuclear inclusions and papillary fronds in CASTLE. However, these findings were not noted in our case. In a study of 20 cases by Ito *et al*., one case of CASTLE was diagnosed on cytology whereas rest were categorized as thyroid carcinoma of the unusual type.[9]

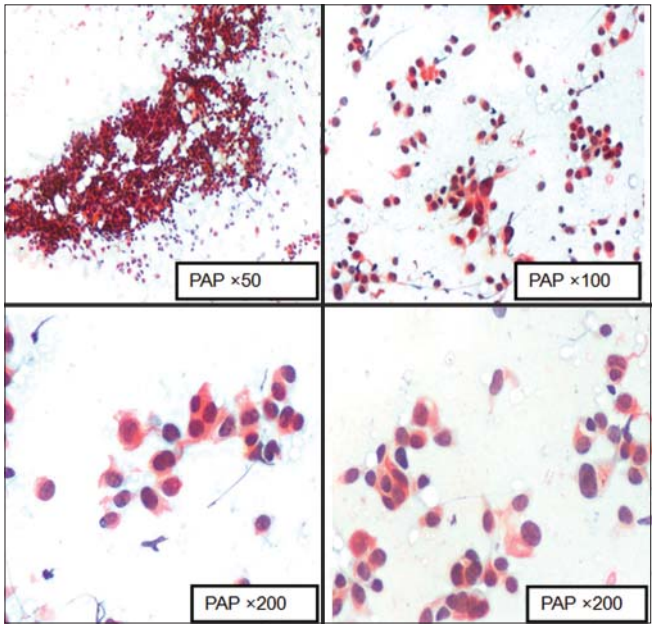

Figure 1: Cytology smears showed dyscohesivepolygonal to ovoid malignant cells with eccentrically placed nuclei (Papanicoloau stain)

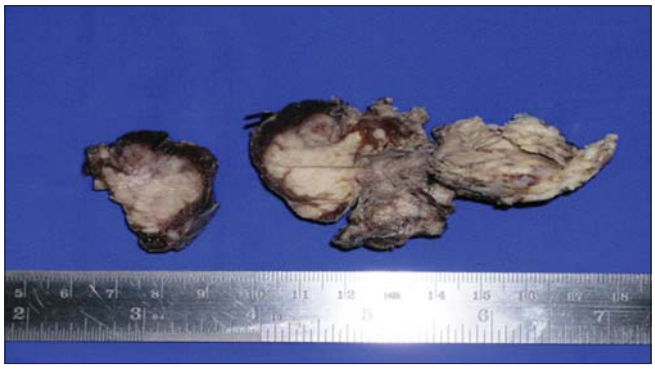

Figure 2: A well-defined lobulated grey white to tan colored thyroid tumor

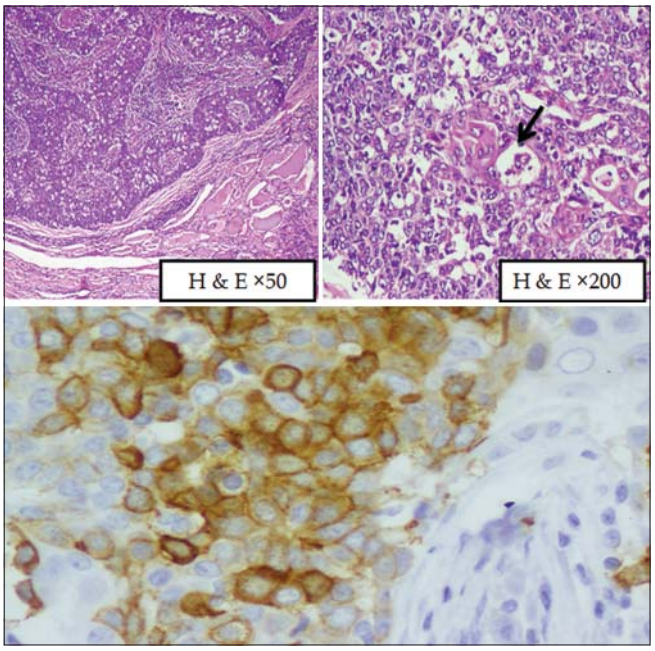

Figure 3: Lobulated tumor with central focus of squamoid differentiation (H and E). Immunohistochemistry: CD5 positivity in tumor cells

Table 1: Cytological differential diagnosis of CASTLE[7,8]

Tumour	Characteristic features
Medullary carcinoma	Wide age group. Family history in familial cases. Raised serum calcitonin. Cytology: Variable cell types, eccentric tumor cell nuclei and amyloid in the background
Anaplastic carcinoma	Older subjects (>70 years), rapid enlargement of thyroid. Local compression symptoms. Cytology: Spindle, giant and bizarre cells in a necrotic background
SCC	Malignant squamous cells, dyskeratoticforms, necrotic background±secondary cystic degenerative changes. History of primary elsewhere
Lymphoepithelioma	Lymphocyte rich smears, poorly differentiated tumor cells. May mimic lymphoma
Papillary carcinoma with squamous metaplasia	Characteristic features of papillary thyroid carcinoma in addition to squamoid cells

CASTLE: Carcinoma showing thymus-like elements, SCC: Squamous cell carcinoma

CONCLUSION

CASTLE, a very rare thyroid tumor is difficult to diagnose pre-operatively. Clinical clues to the diagnosis include relative long duration of swelling, lobulated contour, lack of calcification and extra thyroidal spread. On cytology, characteristic features of other more common thyroid tumors like papillary and follicular neoplasms are absent. Presence of squamoid differentiation and lymphoid population in the background can serve as useful clues when present. Further, cell block or immunocytoto chemistry with CD5 when available, is useful to confirm the diagnosis.

To conclude, the possibility of this uncommon neoplasm must be considered whenever a thyroid tumor with poorly differentiated tumor cells is encountered.

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