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

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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 groove 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 with eccentrically placed nuclei and orangophilic cytoplasm imparting a plasmacytoid appearance in a background of lymphocytes and few follicular cells (Figure 1). 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.

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).

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

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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. 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 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).

The various entities that may be considered on cytology is shown in a tabular form Youens et al., 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.
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 immunocytochemistry 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.

REFERENCES


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