Spindle Cell Variant of Medullary Carcinoma Thyroid- A Case Report

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Abstract

A 14-year-old girl presented with a solitary thyroid nodule since 1 year. Fine needle aspiration cytology (FNAC) from the thyroid swelling revealed discretely arranged spindle cells with scant cytoplasm. Because of diagnostic dilemma, a provisional diagnosis of spindle cell lesion of thyroid was given as distinction of amyloid from collagen and basement-like material was difficult on cytology smears. Also, spindle cell might be seen in anaplastic carcinoma, melanoma, soft tissue sarcomas, etc. Tumor cells were positive for cytokeratin and chromogranin in cell block preparation. Thus, the diagnosis of spindle cell variant of medullary carcinoma thyroid was given.

Keywords: Medullary thyroid carcinoma, fine needle aspiration cytology, Spindle cell variant.

Introduction

Medullary carcinoma of the thyroid (MCT) is relatively a rare tumor, comprising 5-10% of all thyroid malignancies.[1] Nonetheless, its high association with other neuroendocrine Tumors[2] makes an early but accurate diagnosis imperative. Pre-operative diagnosis is important for treatment as well as screening family members for associated multiple endocrine neoplasia (MEN) syndrome. Fine needle aspiration cytology (FNAC) has been a sensitive and specific technique for pre-operative evaluation of thyroid tumors.[3] However, MCT is known to exhibit a wide spectrum of growth pattern; as a consequence, cytodiagnosis becomes more difficult.[4]

Case Details

A 14-year-old girl presented with a solitary thyroid nodule in right lobe measuring 2×1.5 cm since 1 year. She was anemic and in hyperthyroid state. There was family history of thyroid surgery in mother at the age of 29 years but no details were available.

Fine needle aspiration smears showed several small and large clusters of elongated spindle-shaped cells with scant cytoplasm and hyperchromatic, mildly pleomorphic nuclei (Figs. 1 and 2). Scant, glassy pink material was present in the background. A provisional diagnosis of spindle cell lesion of thyroid was given. The amorphous material in background showed crystal violet positivity. Tumor cells were positive for cytokeratin and chromogranin (Figs. 3 and 4) and negative for vimentin in cell block preparation. The diagnosis of spindle cell variant of medullary carcinoma thyroid was given**.**

Discussion

Medullary carcinoma of the thyroid (MCT) was first described by Horn,[5] but he failed to separate it from undifferentiated thyroid carcinoma. In 1959, Hazard et al.[6] adopted the term “medullary carcinoma” and recognized the characteristic amyloid stroma. Later, it was identified as the neuroendocrine tumor arising from the parafollicular cells and capable of producing calcitonin.[7,8]

Sporadic and familial forms occur, with the sporadic form accounting for 70% of cases and familial form 10–20%. Hereditary MCT is transmitted as an autosomal-dominant trait either alone as familial medullary carcinoma thyroid (FMCT) or as part of multiple endocrine neoplasia (MEN) type 2A or 2B.The gene involved in the development of this tumor is the RET gene, located on chromosome 10q11.2.[9]

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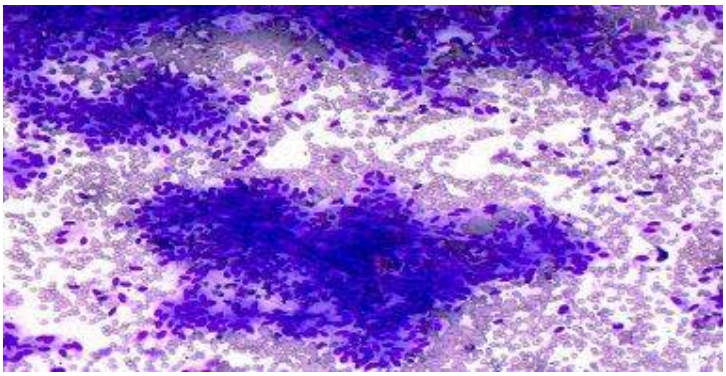

Figure 1.Giemsa Stained Smear Is Highly Cellular Showing Dispersed and Clustered Spindle Cells with Elongated Pale Nuclei and Indistinct Attenuated Cytoplasm

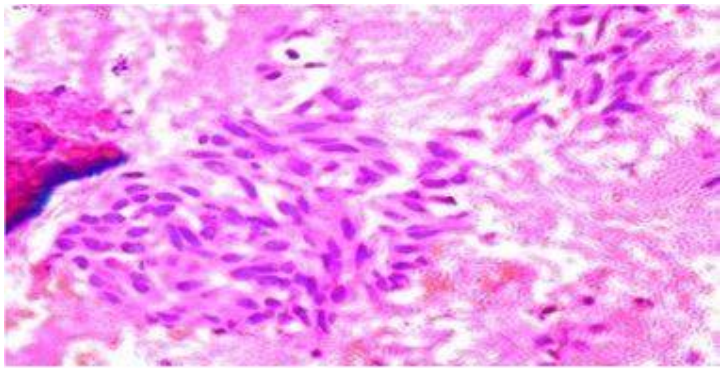

Figure 2.H & E Stained Section Show Spindle Cells in Clusters Showing Mild Pleomorphism

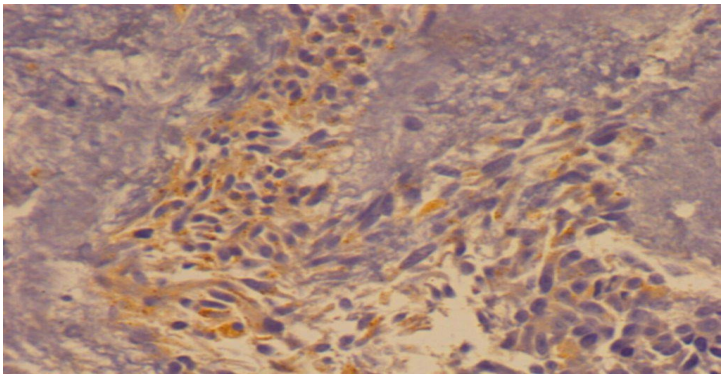

Figure 3.Spindle Cells Showing Positivity for Cytokeratin

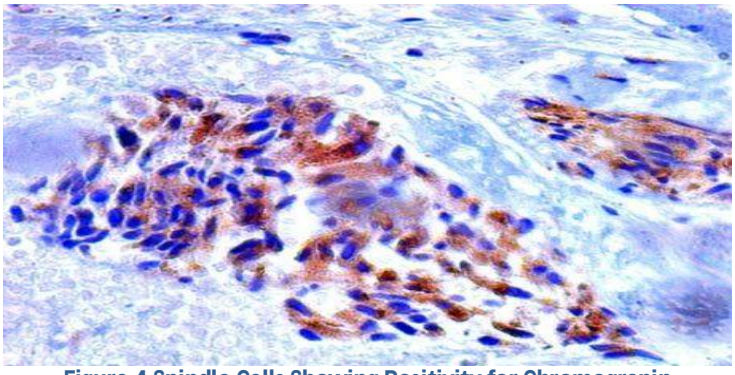

Figure 4.Spindle Cells Showing Positivity for Chromogranin

This tumor has a range of different cytologic patterns plasmacytoid, spindle cell, small cell, follicular, tubular and giant cell variants.[10]

Apart from the classic plasmacytoid cell pattern, the neoplastic cells in FNA smears may resemble spindle cells or small cells with scanty cytoplasm and moulding of nuclei.[10] In such cases, the detection of amyloid is a valuable pointer to the diagnosis. Congo Red staining helps to differentiate amyloid from colloid or hyaline fragments,[10] and is diagnostic for medullary thyroid carcinoma, except in rare cases of primary amyloidosis involving the thyroid.[11] The spindle cell tumor may mimic a fibroblastic tumor or even a melanoma.[10]

The cytological diagnosis of MCT is based on the neuroendocrine differentiation of the tumor cells and the presence of amyloid.[12]

If MCT is cytologically presumed but amyloid and azurophilic cytoplasmic granules are not demonstrated, the use of immunostaining is necessary for correct tumor typing. Generally, the use of calcitonin is sufficient for the diagnosis of MCT, since it is expressed in 80% of the tumors.[13,14] If calcitonin is negative and the cytologic criteria of neuroendocrine differentiation are not so apparent, then immunostaining using antibodies against chromogranin A or synaptophysin should be performed to verify neuroendocrine differentiation of the cells.[15]

Source(s) of Support: None

Conflict of Interest: None

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 ISSN: 2454-8642