Case Report

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Medullary thyroid carcinoma spindle cell variant: a rare case report

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ABSTRACT

Medullary thyroid carcinoma (MTC) accounts for only 4% of thyroid tumors and originates from parafollicular cells secreting calcitonin. However, it accounts for 13% of all thyroid cancer related death. We herein report a case of 50 years male who presented with a right neck swelling measuring $3 \times 2 \times 2$ cm for 2 years, which was gradually increasing. Ultrasonography of the neck revealed a hypoechoic nodule in the right lobe of thyroid. There was no relevant family history. Patient was clinically euthyroid. Fine needle aspiration smears showed many dispersed spindle shaped cells and loosely cohesive clusters of cells. Nuclei of these cells are spindle shaped and showing moderate anisokaryosis. Clumps of hyaline material also seen. A provisional diagnosis of MTC spindle cell variant had been made, which was confirmed by histopathology. Early diagnosis of MTC is very important. Because if patient do not have metastatic disease usually cured by total thyroidectomy.

Keywords: MTC, RET proto-oncogene, MEN syndrome

INTRODUCTION

Thyroid cancer is one of the most common cancers worldwide, with an incidence of 600,000 cases per year. It is common in both developing as well as developed countries.^{1,2} Thyroid cancers are of various types in which medullary thyroid cancer (MTC) is relatively rare and accounts approximately 1% to 2% of all thyroid cancers.¹⁻³ It is a rare neuroendocrine thyroid malignancy and arises from parafollicular C-cells of thyroid. Majority of cases approximately 70% are sporadic with 10-20% familial cases. Hereditary MTC is an autosomal trait associated with RET proto-oncogene located on chromosome 10q11.2. It may present alone as familial medullary carcinoma thyroid or associated with multiple endocrine neoplasia (MEN) type 2A or 2B.4 MTC is usually seen in middle age group with a female preponderance and usually presented as a painless, firm, solitary thyroid nodule. Therefore, fine needle aspiration cytology (FNAC) is used as initial investigation.⁵ In spite of low incidence MTC represent high mortality rate of 8.6% of thyroid cancer-related deaths.¹ MTC may take an aggressive course in some patients, causing early metastasis and leading to marked morbidity and mortality. Total thyroidectomy is the mainstay of treatment, which may be combined with central only or central and lateral neck dissection as it is an aggressive tumor and show frequent cervical lymph node metastases.⁶ Due to its aggressive nature and association with MEN syndrome early diagnosis is important for treatment as well as screening of family members associated with multiple endocrine neoplasia (MEN) syndrome. For this purpose, we can use FNAC as it is a sensitive and specific technique for pre-operative evaluation of thyroid tumours.

CASE REPORT

A 50-year-old male having a nodular, painless swelling on right side of neck since the last 2 years, which was gradually increasing in size got referred to FNAC from ENT outpatient department. Ultrasonography of the neck revealed hypoechoic nodule in the right lobe of thyroid. The patient complained of hoarseness of voice, dysphagia and dyspnoea for last 6 months. On physical examination swelling was 3×2×2cm in size, firm and moved on deglutition. The patient was euthyroid and there was no family history of thyroid swelling or other endocrine tumours. Serum calcitonin was raised, FNAC smears shows high cellularity which comprises mainly dispersed cells and loosely cohesive clusters of cells. These cells are showing cellular and nuclear pleomorphism. Predominantly cells are spindle shaped with few plasmacytoid cells. Nuclei of the spindle cells are spindle shaped and showing moderate anisokaryosis with scanty eosinophilic cytoplasm, amorphous pin material is also seen, few cells show prominent coarse cytoplasmic granularity. Clumps of amyloid material is also seen. Based on these characteristics a provisional diagnosis of spindle cell variant of MTC (Bathesda category 5) was given (Figure 1-4) which was further confirmed by histopathology outside our department. Thus, a diagnosis of MTC spindle cell variant was made.



Figure 1: FNAC smear showing good cellularity with many dispersed cells and clusters (100X MGG).



Figure 2: FNAC smear showing many spindle cells and few plasmacytoid cells (400X).



Figure 3: Cytology smear shows few cells with prominent coarse cytoplasmic granularity andstains bright red (400XMGG).



Figure 4: Cytology smear show spindle cells with moderate anisocytosis, amorphous pinmaterial is also seen (Oil immersion MGG).

DISCUSSION

Horn et al first described MCT but he failed to separate it from undifferentiated thyroid carcinoma.⁷ Hazard et al in gave the term "medullary thyroid carcinoma" (MTC) and described its histopathological characteristics as amyloid stroma.⁸ Giard et al in their study correlated cytological findings with histology to know the accuracy of fine needle aspiration results in thyroid carcinoma and identified amyloid in 43-81% of cases of MTC cases.⁵ Orell et al described that MTC show different cytological patterns like plasmacytoid, spindle cell, follicular, tubular and giant cell variants.⁹ Giard et al, Orell et al and Sangalli et al in their study found that pure spindle cell variant of MTC is rare and usually spindle cells are mixed with other types of cells. Hyalinizing trabecular adenoma and nodular fasciitis variant of papillary differentials.5,9,10 carcinoma are two close Immunohistochemistry (IHC) for calcitonin is considered as gold standard for definitive cytodiagnosis although some cases are negative. Kaushal et al studied 78 cases and made a definitive diagnosis in 54 cases i.e., 87.1% based on cytomorphology alone and in 12.9% based on IHC for calcitonin.11 This study favours our study and demonstrate the importance of FNAC alone in diagnosis of MTC. Mondal in his study found the correct diagnosis of majority of MTC cases can be made by cytological study but should be correlate with histopathological examination.

CONCLUSION

MTC is a rare aggressive thyroid carcinoma and have different variants. Mostly diagnosis can be made on cytological examination and with serum calcitonin level. FNAC plays very important role in MTC diagnosis as it is simple, cost-effective, less invasive procedure and helps in early diagnosis as well as guidance for the treatment.

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