A Benign Medullary Thyroid Cancer

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BACKGROUND

Metastatic dissemination occurs in up to 90% of patients with Medullary Thyroid Cancer (MTC) > 4 cm in diameter and Calcitonin > 500. Herein, we present a patient with a large tumor and extremely elevated calcitonin levels, who was subsequently found to show a “benign” behavior with no evidence of metastasis.

CLINCAL PRESENTATION

A 62-year-old woman presented to our endocrinology clinic for evaluation of a 2-year history of “lump” sensation in the neck. She reported no dysphagia, choking sensation, neck pain, hoarseness, or any symptoms of hypo/hyperthyroidism. She had no history of radiation to the neck. Physical examination was only remarkable for a 4-cm thyroid nodule on the right side which moved easily with swallowing. No cervical lymphadenopathy was appreciated. Pemberton’s sign was negative.

CLINCAL PRESENTATION..Cont’d

Ultrasound of the neck revealed a right sided hypoechoic nodule measuring 4.12 x 1.84 cm. Thyroid function test was normal. Calcitonin and CEA were 8754.0 pg/mL and 29.0 ng/mL, respectively. FNA cytology demonstrated monomorphic population of numerous spindle cells with very few neoplastic cells that stained focally for Chromogranin A, consistent with MTC. Pan CT revealed no evidence of metastasis. The patient underwent total thyroidectomy after exclusion of pheochromocytoma. Genetic studies were only remarkable for positive HRAS mutation. Result for RET mutation was negative, as were the results for PAX8/PPARG, NRAS and KRAS mutation.

Following surgery, the patient was initiated on Levothyroxine to maintain TSH levels within normal limits.

At 5 months of follow-up, calcitonin level declined to 3 ng/mL and CEA remained within normal range (1.5 ng/mL).

DISCUSSION

The relatively benign behavior of this patient's MTC despite extremely elevated calcitonin and CEA levels highlights the value of the results of genetic studies on disease prognostication. This case highlights the need for comprehensive research studies on genomic profiling in MTC to better understand the relationship of different genomic mutations with prognosis and outcome.

CONCLUSION

Presence of HRAS mutation in MTC is likely to be associated with better prognostic outcome.

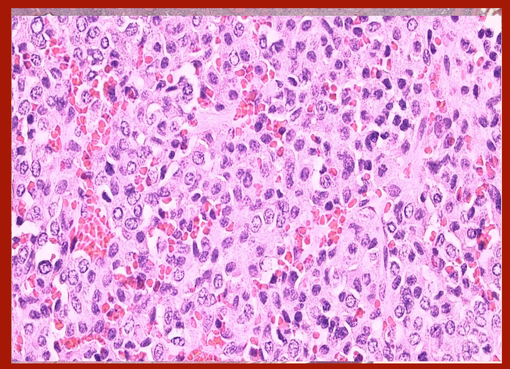

Figure 1

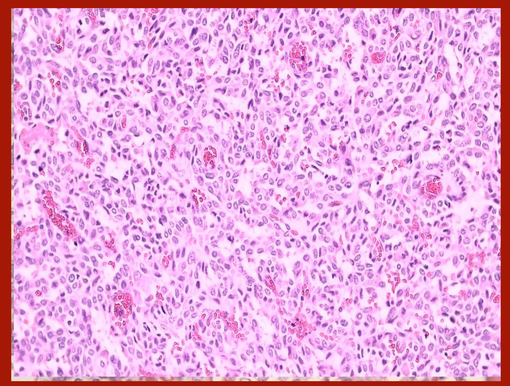

Figure 2

Figure 1. Shows solid sheets of epithelioid cells separated by delicate fibrovascular stroma. The cytoplasm is amphophilic. The nuclei are round or oval with a salt and pepper chromatin pattern suggestive of a neuroendocrine origin of the tumor.

Figure 2. Shows Round nuclei with finely stippled to coarsely clumped chromatin and indistinct nucleoli which stains positive for amyloid on Congo red staining