Medullary Thyroid Carcinoma

There are four main types of thyroid cancer, listed in descending order of prevalence are papillary, follicular, medullary, and anaplastic. Medullary thyroid carcinoma, the third most common type of thyroid cancer is divided into sporadic and familial types. This cancer arises from parafollicular C cells, may produce calcitonin and manifest with hypocalcemia or produce ACTH and manifest with Cushing syndrome. It is characterized histologically by amyloid-staining sheets of cells and will exhibit calcitonin release with pentagastrin administration. Two major risk factors for medullary carcinoma are a history of head and neck radiation and family history of MEN 2A and 2B.



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Sporadic and Familial Types
Sporadic-spear and Familial-family
The majority of medullary carcinomas are sporadic and have no association with other genetic factors or syndromes. The other remaining cases are due to an autosomal dominantly inherited RET gene mutation that can lead to familial medullary thyroid cancer or to either MEN 2A or MEN 2B syndromes, in which medullary thyroid cancer is commonly present. Familial cases generally carry a better prognosis than sporadic.

Arises from Parafollicular C cells
Pair-of-folliciles C-cat
These neuroendocrine cells within the thyroid gland produce and secrete calcitonin.

Produces Calcitonin
Cow-throne
Calcitonin is a peptide hormone that has antagonistic effects to parathyroid hormone (PTH); it is secreted in response to high blood calcium levels and its primary function is to lower calcium levels by decreasing absorption in the GI tract, modifying bone resorption, and increasing excretion by the kidneys. By measuring serum levels of calcitonin, it can be used as a tumor marker for medullary carcinoma.

Hypocalcemia
Hippo-calcified-cow
Since calcitonin downregulates calcium in the blood, an excessive amount of calcitonin may produce hypocalcemia, which can manifest as tetany, paresthesias, seizures, and EKG abnormalities.

May Produce ACTH
Air-conditioning
Adrenocorticotropic hormone is a peptide hormone normally produced by the anterior pituitary gland. Like many other cancers, medullary thyroid carcinomas can produce ectopic ACTH and therefore patients can present with signs and symptoms of Cushing syndrome such as weight gain, hypertension, abdominal striae, etc.

Diagnosis

Sheets of Cells with Amyloid Deposition
Bed-sheets of Cells and Armadillo
Medullary carcinoma cells arrange themselves into sheets; the cells that form these sheets contain excess calcitonin that is eventually converted into amyloid protein. These amyloid deposits can be stained with Congo Red dye and will exhibit the pathognomonic apple-green birefringence when viewed under polarized light.

Increased Calcitonin with Pentagastrin Infusion
Up-arrow Cow-throne with Pentagram-gas
As a diagnostic test, a synthetic peptide hormone called Pentagastrin can be administered to a patient in order to stimulate the release of calcitonin from C cells, which can then be measured and compared to baseline levels. Patients with medullary thyroid cancer will have levels markedly above normal or baseline after pentagastrin infusion.

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Risk Factors

MEN 2A and 2B

2 MEN with (A) Apple and (B) Bee

These two subsets of the multiple endocrine neoplasia (MEN) syndromes both involve medullary thyroid cancer and pheochromocytomas. MEN 2A also classically includes parathyroid hyperplasia, while MEN 2B also manifests with mucosal neuromas, or more specifically oral ganglioneuromatosis. Both of these syndromes are inherited in an autosomal dominant fashion via the RET oncogene on chromosome 10.

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