MEN 2A (Multiple Endocrine Neoplasia)

MEN 2A syndrome is a group of neoplasms that often occur together due to a mutation in the RET oncogene. The neoplasms typically originate from medullary thyroid cells of the thyroid, the parathyroid glands, and the chromaffin cells of the adrenal glands resulting in a pheochromocytoma.



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Pathophysiology

Autosomal Dominant
Domino
The disease is passed in an autosomal dominant fashion. Therefore, there is often a significant family history in patients that present with this disease.

RET Gene
Roulette
RET is a tyrosine kinase receptor on chromosome 10. It is an oncogene that develops a gain of function mutation in the MEN2A and MEN2B syndromes.

Signs and Symptoms

Medullary Thyroid carcinoma
Medal Thyroid
Medullary carcinomas of the thyroid are neuroendocrine tumors derived from the C cells, or parafollicular cells of the thyroid and are often seen in MEN 2A and MEN2B syndromes. Cases associated with the MEN syndrome occur in younger persons while sporadic cases develop much later in life. These tumors secrete calcitonin, and altered calcitonin polypeptides can deposit as amyloid, which has a characteristic red stain on Congo dye.

Calcitonin
Cow-throne
Calcitonin plays a role in calcium homeostasis and is produced by medullary cells of the thyroid. Calcitonin polypeptides can deposit as amyloid, which stains red with Congo dye and appears apple green under birefringence.

Parathyroid
Parachute-parathyroid
Parathyroid tumors are characteristically involved with MEN2A due to the mutation in the RET gene. These tumors can present as hypercalcemia with complications like kidney stones.

Pheochromocytoma
Phiat-chrome
This is a neuroendocrine tumor of the chromaffin cells which secrete catecholamines. This tumor is of neural crest cell origin and is often found in the adrenal medulla. It can present with the classic triad of intermittent palpitations, headache, and diaphoresis.

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