MEN 1 (Multiple Endocrine Neoplasia)

Multiple endocrine neoplasm type one is a group of neoplasms occurring together due to a specific gene mutation. This gene mutation is inherited in an autosomal dominant fashion which results in characteristic patient presentations. In MEN I, the neoplasms typically originate from the pancreas, the parathyroid gland, and the pituitary gland. Tumors in the pancreas include several tumor types including gastrinomas, insulinomas, VIPomas, and glucagonomas. Excess parathyroid hormone can present as hypercalcemia and pituitary tumors often produce prolactin or growth hormone.



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Pathophysiology

Autosomal Dominant
Domino
This disease is inherited in an autosomal dominant fashion.

Menin Mutation (Tumor Suppression Gene)
Minion Mutant
Menin is coded by the MEN 1 gene and is a tumor suppressor gene. This mutation on chromosome 11 leads to tumor growth.

Chromosome 11
Chromosome (11) Double-wand
This mutation on chromosome 11 leads to MEN 1 syndrome.

Signs and Symptoms

Pancreatic Endocrine Tumors
Pancreas and Tumor-guy
Tumors of the pancreas, as well as the duodenum, are one of the culprits of MEN1. There are several types of tumors in the pancreas, including gastrinomas, insulinomas, VIPomas, and glucagonomas. Depending on the type of pancreatic tumor, patients can present in a variety of ways, including ulcers, hypoglycemia, or hyperglycemia.

Pituitary Tumors
Pitbull-terrier Tumor-guy
Pituitary tumors are characteristic of MEN1 and are most commonly prolactinomas. Growth hormone tumors can also occur. Enlargement of a pituitary tumor can compress the optic chiasm, causing bitemporal hemianopia.

Parathyroid Adenomas
Para-thigh-droid Add-gnome
Parathyroid tumors are characteristically involved with MEN1 and present as hypercalcemia. Many times, patients will present with kidney stones as a consequence of prolonged hypercalcemia.

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