

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 1, 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.



PLAY PICMONIC

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 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 on 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.