Multiple Endocrine Neoplasia(MEN syndrome)

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outline

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- Classification
- MEN1 / MEN2
- epidemiology
- Clinical manifestations
- Diagnosis
- Treatment
- conclusion

Definition

- Multiple endocrine neoplasia syndrome is defined as a disorder with neoplasms in two or more different hormonal tissues in several members of a family.

- Usually at an earlier age.

- Currently three well-defined MEN syndromes – MEN 1, MEN 2a, MEN 2b.

- Several distinct genetic disorders predispose to endocrine gland neoplasia and cause hormone excess syndromes .

- DNA-based genetic testing is now available for these disorders.

- effective managment requires an understanding of endocrine neoplasia and the range of clinical features that may be manifested in an individual patient

. MEN1 (Wermer's syndrome)

- It is inherited as an autosomal dominant trait.
- Although the mechanism of tumor genesis at the cellular level is recessive.
- Clinical manifestations vary

Generally manifest by the 3rd or 4th decade

Although rare, MEN1 is the most common MEN syndrome with prevalence of 2–20 per 100,000 .

Multiple endocrine neoplasia (MEN) type I, also known as Wermer syndrome, is an autosomal dominant syndrome characterized by:

- parathyroid hyperplasia: with resulting hyperparathyroidism seen in 80–95% of patients
- islet cell tumors of the pancreas: 50% of patients, and a major cause of mortality, most often gastrinomas followed by glucogonomas
- pituitary adenomas: 30% of patients, most frequently prolactinomas

Other associations

1. lipomas
2. angiofibromas
3. adrenal cortical lesions
4. adrenal adenomas
5. adrenocortical hyperplasia
6. cortisol-secreting adenomas
7. adrenal carcinomas (rare)
8. carcinoid tumors
9. hepatic focal nodular hyperplasia
10. Zollinger-Ellison syndrome

- **Race**

No racial predilection is known.

- **Sex**

The incidence is equal for men and women.

Primary hyperparathyroidism

- The most common manifestation of MEN1 (95–100%).
- Hypercalcemia may develop during the teenage years,(hyperplasia of parathyroid glands)
- most individuals are affected by age 40 (adenoma of parathyroid gland).
- One of the cardinal features of endocrine tumors in MEN1—multicentricity.

- The neoplastic changes inevitably affect multiple parathyroid glands, making surgical cure difficult.
- **Diagnosis :**

1. demonstrating elevated levels of serum calcium
2. intact parathyroid hormone.

Clinical manifestations:

- Hyperparathyroidism of MEN1 do not differ substantially from those in sporadic hyperparathyroidism and include:

1. calcium-containing kidney stones,
2. kidney failure,
3. nephrocalcinosis,
4. bone abnormalities (i.e., osteoporosis, osteitis fibrosa cystica),
5. gastrointestinal and musculoskeletal complaints

- . Management of MEN1 is challenging because of;

1. early onset,
2. significant recurrence rates,
3. multiplicity of parathyroid gland involvement.
4. Ectopic/ occult glands.

- Hyperparathyroidism of MEN1 can be differentiated from other forms of familial primary hyperparathyroidism based on;

1. family history,
2. histology of resected parathyroid tissue,
3. presence of a *MEN1* mutation and,
4. long-term observation to determine whether other manifestations of MEN1 develop.

treatment

- **Criteria for surgery**

1. Individuals with serum calcium levels >3.0 mmol/L (12 mg/dL),
2. evidence of calcium nephrolithiasis or renal dysfunction,
3. neuropathic or muscular symptoms, or

bone involvement (including osteopenia) or individuals <50 years of age .

- When surgery is indicated , there are two approaches.

I. All parathyroid tissue are identified and removed and parathyroid tissue is implanted in the nondominant forearm.

- + Thymectomy because of development of malignant carcinoid tumors.

2. Remove 3–3.5 parathyroid glands from the neck (leaving ~50 mg of parathyroid tissue),