

A Rare Presentation of Multiple Endocrine Neoplasia Type 1 Syndrome as a Sphenoid Sinus Neuroendocrine Tumor

A Diagnostic Journey from Ectopic Pituitary Adenoma to Multiglandular Endocrine Disease

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An ectopic sphenoid prolactinoma may be the first clue to MEN1.



Introduction

MEN1 is an autosomal dominant syndrome involving the parathyroid glands, pituitary and gastrointestinal neuroendocrine tissues.

Ectopic pituitary adenomas account for <1% of pituitary adenomas and may mimic sinonasal neoplasms.



Case presentation

A 23-year-old woman first presented at age 19 with rhinorrhea and irregular menses. A sphenoid sinus mass was initially classified as a grade II neuroendocrine tumor. Endocrine evaluation identified an ectopic sphenoid prolactinoma, with biochemical improvement and radiological regression on cabergoline.

Multi-gland primary hyperparathyroidism and multifocal pancreatic NETs subsequently established the clinical diagnosis of MEN1.



As the story unfolds-Case Journey

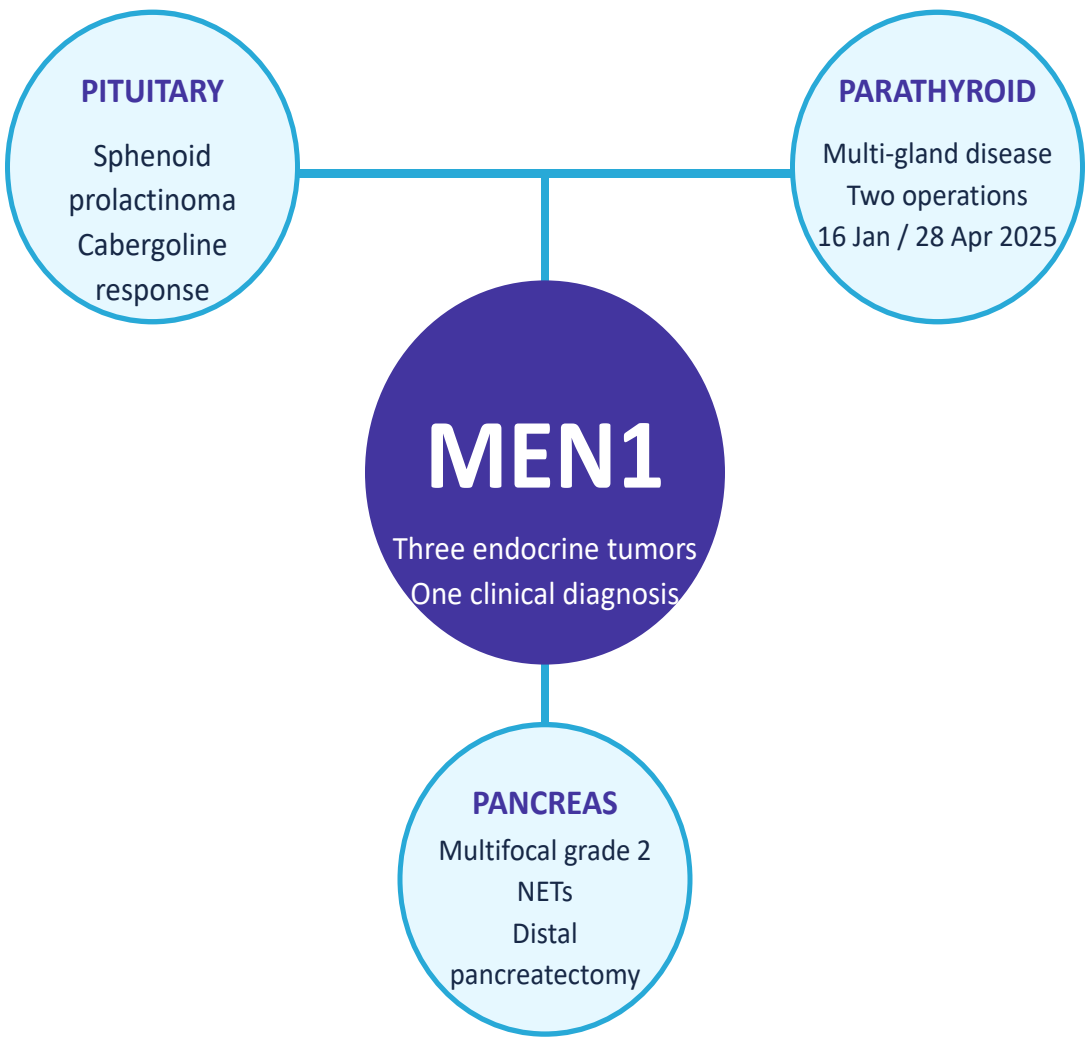
AGE 19 — Sphenoid prolactinoma; response to cabergoline.
AGE 21 — Multi-gland hyperparathyroidism; surgery and forearm reimplantation.
AGE 22 — Multifocal pancreatic NETs; distal pancreatectomy.
AGE 23 — Three tumor types establish clinical MEN1.



Histopathology

Pituitary adenoma and **parathyroid adenoma**.
Pancreas: multifocal WHO grade 2 NETs, Ki-67 10%, largest 1.5 cm, mpT1N0 with negative margins.

THE MEN1 CONNECTION



Discussion

MEN1 emerged through sequential endocrine manifestations. The unusual sphenoid presentation initially obscured the pituitary origin and the wider syndrome.

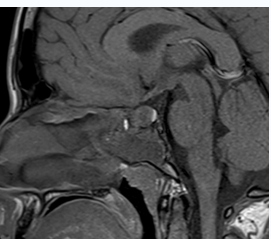
Hyperprolactinemia, seller continuity on MRI, and response to cabergoline supported a prolactin-secreting pituitary lesion. Joint endocrine, radiological and pathological review helped establish the diagnosis.

Persistent biochemical disease after the first parathyroid operation required repeat localization and surgery, highlighting the complexity of multi-gland disease. Surveillance identified multifocal, node-negative grade 2 pancreatic NETs.

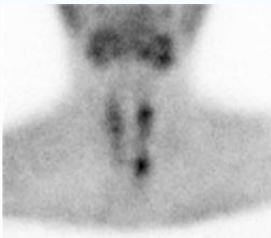
Lifelong multidisciplinary surveillance, genetic counselling and family evaluation remain essential.



Radiology



MRI: right pituitary lesion 10 × 12.3 × 10 mm with leftward stalk deviation, subtle peripheral enhancement and no optic chiasm compression.
Sphenoid solid–cystic mass 2.8 × 3.4 × 2.4 cm, inseparable from the pituitary fossa.



MIBI: *two left-sided hyperfunctioning glands*—0.5 × 0.7 cm behind the upper pole and 1.0 × 0.8 cm below the lower pole. Focal uptake persisted at 60 and 120 minutes, with washout from the thyroid lobes.



Biochemical profile

Parameter	Peak pre-op/Treatment	Post-op / treatment
Calcium (mg/dL)	11.31	8.804
Prolactin (ng/mL)	132	23.96
PTH (pg/mL)	167.1	7.5



Management and course

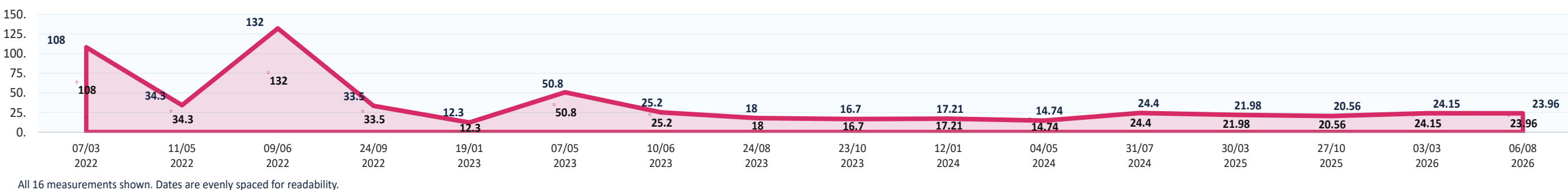
Cabergoline improved prolactin levels and imaging. Parathyroid operations on 16 January and 28 April 2025 addressed multi-gland disease. Distal pancreatectomy treated multifocal pancreatic NETs; ongoing endocrine surveillance remains essential.



Prolactin trend (ng/mL)

Bromocriptine held: symptoms recurred

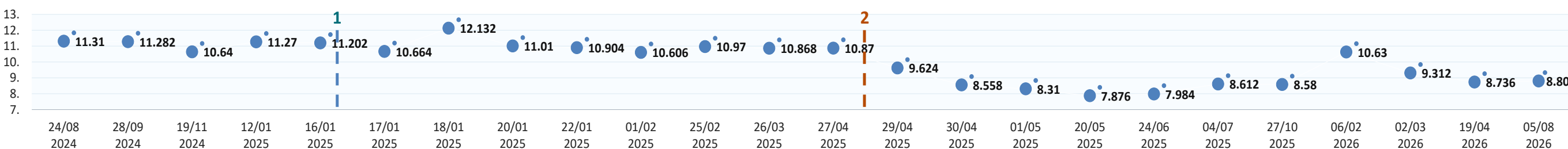
Cabergoline initiated after 9 June 2022



Corrected calcium trend (mg/dL)

16 Jan 2025 · FIRST PARATHYROID SURGERY

28 Apr 2025 · SECOND PARATHYROID SURGERY



Recognize the unusual. Connect the endocrine clues.

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