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A lady with MEN -2

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Introduction: Multiple endocrinal neoplasia type 2 A is a rare familial cancer syndrome caused due to mutation in the RET proto-oncogene characterized by medullary carcinoma of thyroid, pheochromocytoma, parathyroid hyperplasia.
Presentation: 47 year lady with known case of pheochromocytoma (left adrenalectomy since 21 years ago) presented again with paroxysmal spells, right lower chest pain and goiter. Family history revealed her mother had goiter and type 2 diabetes. On examination, there was multinodular goiter, BP ranging from 140/90- 240/130 mmHg (on irregular antihypertensive medications), Lab results revealed urinary VMA- 31.3 mg/24hr (Ref 1-11mg/24hr), calcitonin-1840 pg/ml (Ref 0-10 pg/ml) and CEA-70.14 ng/ml (Ref 0-4.7 ng/ml). Thyroid profile, Parathyroid hormone, calcium and phosphate were normal, MRI (abdomen & pelvis) – right adrenal mass, mixed cystic and solid lesion (6x 6.9 cm), left adrenal solid mass (1.7 x 1.8 cm). She was diagnosed as MEN 2 A involving pheochromocytoma (bilateral adrenal tumor) and medullary thyroid carcinoma. Laparoscopic right adrenalectomy and total thyroidectomy were done. Histopathological reports were consistent with phaeochromocytoma and medullary thyroid carcinoma. Two weeks after operation, BP was normal without antihypertensive medications. Her calcitonin level also fell to normal. She is taking steroid and thyroxine replacement together with calcium and vitamin D.
Discussion: Should keep in mind to observe the recurrent of phaeochromocytoma and also subsequent appearance of other components of MEN-2.