

A case of adrenal composite pheochromocytoma-ganglioneuroma with intrathoracic goiter

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Adrenal composite pheochromocytoma-ganglioneuroma is extremely rare. We report a case of a 70-year-old female with compound adrenal medullary tumor and intrathoracic goiter. She had hypertension and diabetes for 13 years and 3 years, respectively. She complained of epigastric discomfort 2 months ago. On esophagogastroduodenoscopy, an extrinsic compression finding on the mid-esophagus was noted. A chest computed tomographic (CT) scan revealed an intrathoracic goiter and an about 8-cm-sized thick walled cystic lesion around the pancreatic tail. She had no family history of thyroid disease, and her serum TSH, free T4, and T3 level was 0.16 μ IU/mL, 1.03 ng/dL, and 137.02 ng/dL, respectively. A thyroid peroxidase (TPO) antibody test was negative. A pancreatic magnetic resonance imaging (MRI) also showed an septated cystic mass mimicking an pancreatic cystic neoplasm in the same region. She underwent explorative laparotomy, however no pancreatic mass was detected on palpation. In stead, left adrenalectomy was done, because an adrenal mass was noted. Pathologic diagnosis was a compound adrenal medullary tumor, composed of pheochromocytoma and ganglioneuroma. Ultrasound (US) guided thyroid fine needle aspiration cytology was done, because there were several heterogenous lesions on thyroid US. There was no evidence of medullary thyroid carcinoma (MTC) on pathologic exam, and calcitonin level was 2.7 pg/mL. Therefore, she could not be diagnosed with multiple endocrine neoplasia type 2 (MEN2) syndrome. Removal of the adrenal mass improved her blood pressure and blood glucose level.

폐 유암종을 동반한 제1형 다발성 내분비선종증 1예

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제1형 다발성 내분비선종증은 상염색체 우성으로 유전 되는 질환으로 부갑상선, 췌장, 뇌하수체 등에 내분비 종양이 동시에 발생하는 질환이다. 그 이외에 위장이나 흉선 및 폐 등에 유암종이 동반되는 경우가 보고되고 있으며, 폐 유암종은 빈도가 매우 드물어 국내에는 아직 보고된 예가 없었다. 저자들은 상복부 통증을 주소로 내원한 62세 여자 환자에서 시행한 상부 내시경 검사상 다발성 궤양을 관찰 하였으며, 호르몬론 검사, 방사선 검사 및 옥트레오티드(octreotide)를 사용한 somatostatin receptor scintigraphy (SRS)를 통하여 췌장 두부의 가스트린종, 프로락틴 분비 뇌하수체 선종, 원발성 부갑상선 기능항진증 및 폐 유암종을 진단하고 유전자 검사를 통하여 제1형 다발성 내분비선종증을 확진 하였다. 국내에서는 폐유암종을 동반한 제1형 다발성 내분비 선종의 보고가 아직까지 없었기에 문헌 고찰과 함께 보고하는 바이다.

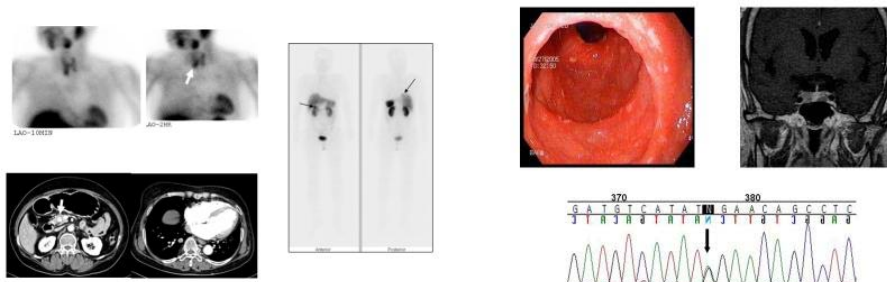


Figure 1

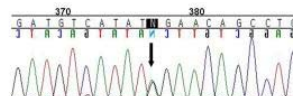


Figure 2