ONE FAMILY SYNDROME MEN 2 A HISTORY

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Background/Purpose: MEN-2a (Sipple syndrome) - include medullar thyroid carcinoma (97 – 100 %), pheochromocytomas of adrenal gland (40 – 60 %), parathyroid tumor 20 – 60 %).
Methods: 5 cases of MEN 2A Syndrom in one family.
Results: Patient C. 25 years old male, underwent thyroidectomy concerning medullary thyroid carcinoma (MTC) in 1998. Presented to the clinic in January 2000 with complaints on headache attacks with tremor, fear, blood pressure rising up to 175mm Hg. Pheochromocytomas of left adrenal gland was revealed and adrenalectomy was performed. In 2004 metastases of MTC were revealed, left modified radical neck dissection was performed. His younger 23 years old brother suffered from congenital hydronephrosis, underwent right nephrectomy in 1 year old age. MTC was recognized at the age of 19, thyroidectomy was performed. Right adrenal Fore years later right adrenal Pheohromocytomas was recognized and removed. Their father was operated on concerning MTC when he was 23 years old man. Later pheocromocytomas was revealed but he refused from surgery. In 2000 patient suddenly dead. On autopsy ruptured 1kg Pheochromocytomas was revealed. In the sun of the oldest brother of ovarian twins was examined. The defective gene (mutation in T1900C (C634R) in 11 exon gene RET) was found. Prophylactic thyroidectomy was performed and MTC 2 mm focuses was fined. The same pathological gene was found. Patient will be operated on three month later.
Discussion & Conclusion: if even one symptom of MEN syndrome is revealed, comprehensive investigation patients and their family should be performed.