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Current Views on Oestriol

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140. Pheochromocytoma and multiple endocrine adenomatosis. Observation of unusual cases

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The diagnosis of pheochromocytoma (P) is based on elevated excretion of catecholamines (CA), adrenalin (A) and noradrenalin (NA), respectively their metabolites vanillylmandelic acid (VMA) and the metanephrines. Localization of the biochemically established tumor has to be performed in every patient before surgery. Though adrenal phlebography, ultrasonography (US) and computerized tomography (CT) have been shown to be useful, selective CA measurement during venous catheterization has been suggested to be a superior method for localization [1].

In recent years we have had the opportunity to observe several interesting and unusual cases (n = 5). In one female patient (42 y) the NA and A measurements in the venous blood (2) suggested a tumor in the area of the right atrium, although a $7 \times 4 \times 3$ cm tumor could be documented by CT and later proven by surgery in the right adrenal gland. The CA-concentration gradient in the right atrium was explained by the atypical venous drainage of the right adrenal. In a male patient (51 y) no CA-gradient was found by selective catheterization although the urinary excretion of NA was several times above 500 µg/24 h accompanied by the clinical picture of CA-excess. Although no other localization procedure could document the tumor, the patient was laparotomized. No tumor could be found during extensive surgical exploration. The tumor in this patient was probably equipped with multiple venous drainage and small as suggested by the high NA-excretion in the presence of normal VMA and metanephrines. In another male patient (52 y) with excessive NA excretion up to 6000 µg/24 h the highest NA concentration was found in both jugular veins on two separate occasions. US and CT did not reveal a tumor in the peritoneal or thoracic cavity, whereas tomography of the base of the skull showed enlargement of the right bulbus venae jugularis demonstrating a NA-producing paraganglioma. In a female patient (60 y) and her daughter (31 y) the described but rare combination of P with medullary thyroid carcinoma and hypercalcitoninemia was found. The mother with unilateral P had been thyroidectomized because of high calcitonin levels. However, the thyroid was free of carcinoma, which was only present in one lymph node of the neck. This finding can be explained by a previous thyroid resection 15 years ago for suspected endemic goiter without histological examination. The daughter had a bilateral P, which was operated 1968 on the left and 1979 on the right side. Total thyroidectomy led to normalization of the calcitonin levels.

The conclusions drawn from these cases are the following: Diagnostic procedures should be extended until tumor localization is established without doubt. Surgical exploration without tumor localization is not advisable. Calcitonin should be measured in every patient with pheochromocytoma. In cases of multiple endocrine adenomatosis type II family screening for P and elevated calcitonin is necessary.

References

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