44 A diagnostic challenge: Malignant and metastatic pheochromocytoma

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Introduction & Objectives: Pheochromocytoma (PCC) is a rare catecholamine producing tumor, which arises from chromaffin cells of the adrenal medulla, occurring in less than 0.2 percent of patients with hypertension. They are mostly benign, but they may also transform into malignancy in 10% (8% to 12.5%) of the cases.

Materials & Methods: We present the clinical history, imaging studies and comprehensive therapy of a 65-year-old Caucasian male, who was suspected of having myocardial ischemia and later diagnosed to have malignant pheochromocytoma.

Results: The patient was referred to our hospital with night sweats, labile hypertension and a positive cardiac stress test. Elective coronary catheterization detected 80% stenosis of the left circumflex artery and percutaneous intervention with DES implantation was performed. One week after discharge, he presented with chest pain to the emergency room and underwent re-angiography, without finding any significant stenosis or stent occlusion. An abdominal ultrasound revealed a 6x10 cm large enhancing renal mass. A contrast-enhanced CT confirmed the left renal mass and in addition it showed para-aortic lymph node enlargement too. The presumptive diagnosis of renal cell carcinoma (RCC) was made. Extended tumor nephrectomy with para-aortic lymphadenectomy was carried out, complicated by hypertensive crises episodes during and after surgery. Histopathological examination of the sample revealed malignant pheochromacytoma pT3 pL1 pV1 pN1 (two from four lymph nodes positive). An ^{18}F-DOPA PET study showed two suspected bone metastases in the 7th vertebral body and in the 11th rib. Due to the implanted non-MRI-compatible stent, the preferable MRI was not performable. Further staging with Iodine-123 metaiodobenzylguanidine (^{123}I-MIBG) scintigraphy was negative. High normetanephrine plasma level (290,7 pg/ml) was found postoperatively. According to the decision of the multisciplinary oncology team the two suspect areas were irradiated with 43,01 Gy. A control DOPA/PET in 3 months showed a tumor regression in the irradiated areas but new bone lesions along the vertebra and one new positive paraaortic lymph node were detected. The normetanephrine plasma level increased to 658 pg/ml. The patient was referred to a specialized center, where a DOTA-TOC-PET was performed with positive result. Targeted radio nuclide therapy (DOTA-TOC therapy with radioactive lutetium-177) was planned, the first treatment cycles have already been administrated.

Conclusions: Malignant and metastatic pheochromocytoma is a rare neuroendocrine tumor, the proper identification is often challenging. Because of the rarity of the disease and the consequently limited number of patients available for clinical study, there is no good evidence for a standard of care; individual approach should be applied for each patient.