C34

Surgical Resection of the Primary Tumor in Patients with Metastatic Pheochromocytoma and Sympathetic Paraganglioma: Impact on Survival and Quality of Life

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Background: Patients with metastatic pheochromocytoma and paraganglioma (PPG) exhibit a decreased overall survival. Current treatments for these tumors are not curative. The role of surgical resection of the primary tumor in patients with metastatic disease is still to be determined.

Methods: We performed a retrospective of MD Anderson Cancer Center database of patients with metastatic pheochromocytoma and paraganglioma. Demographic, clinical, biochemical, imaging, and pathological variables were analyzed.

Results: There were 113 patients with metastatic PPG; 89 had surgical removal of the primary tumor. 53 patients had synchronous metastases; of these 33 had resection of the primary tumor. The median diameter of the primary tumor in the whole cohort was 8.45 centimeters (range 1.90-24 cms).

Survival rates were longer in patients who underwent removal of the primary tumor when compared with patients who did not undergo resection (148 months, 95% CI 112.7-183.2 vs 36 months, 95% CI 27.2-44.8; $p<0.001$). In patients with synchronous metastases treated with surgery for the primary tumor, the survival rates were longer that in patients that were not treated with surgery (85 months, 95% CI 64.5 – 104.5 vs 36 months, 95% CI 29.7 – 42.3; $p<0.001$). No differences in age, race, primary tumor size or genetic background were noted in these cohorts. Survival rates in patients with metastatic disease were independent of the location of the primary tumor. In patients with hormonally active tumors treated with primary tumor resection, the symptoms of catecholamine excess including hypertension improved. Tumor burden rather than hormonal secretion is a more important determinant of overall survival in patients hormonally active metastatic PPG.

Conclusions: Resection of the primary tumor in patients with metastatic PPG is associated with an improved overall survival. In patients with hormonally active tumors, surgical resection is associated with better blood pressure control and symptom improvement.

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