CASE REPORT OF MALIGNANT METASTATIC PHEOCHROMOCYTOMA

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Objective: To report a case of malignant metastatic pheochromocytoma.

Case presentation: A 22 year old gentleman was admitted with uncontrolled hypertension. He had a history of right adrenalectomy when he was four years old secondary to hypertension and pheochromocytoma. He remained stable after that until the age of 12 years when he developed hypertension again. Due to poor compliance with medications, his hypertension had remained uncontrolled. His mother and his maternal aunt also had history of surgery for pheochromocytoma. Plasma normetanephrine levels were elevated at 827 pg/mL. His calcitonin, intact PTH and calcium were normal at 4.9 pg/mL, 31 pg/mL and 9.4 mg/dL respectively. A CT scan of abdomen showed two nodules in left adrenal measuring 1.7 X 1.5 cm with two nodules in periaortic chain, right external iliac lymphadenopathy, right sided bladder mass, and multiple nodules in seminal vesicles. An I-123 MIBG scan was performed and it showed intense localization in left adrenal, right side of urinary bladder and right iliac lymphadenopathy, consistent with pheochromocytoma. Patient was deemed not to be a candidate for surgical resection and FDA approval was obtained to treat patient with I-131 MIBG ablation therapy.

Discussion: Pheochromocytomas are chromaffin tumors arising in adrenal medulla. They are unilateral in 90% of cases. Bilateral pheochromocytomas are common in familial pheochromocytoma syndromes. Treatment is surgical resection. Chemotherapy has been used with cyclophosphamide, vincristine and dacarbazine in 14 patients with a median survival of 3.3 years. Sunitinab was used in anecdotal case reports. Our patient has malignant recurrent metastatic likely familial pheochromocytoma. I-131 MIBG ablation therapy is not currently approved by FDA for treatment of malignant metastatic pheochromocytoma. However small case studies have shown improved survival with it (4.7 vs. 2.8 years in one study with 500 mCi). Dose ranges have been between 100 to 1690 mCi with more response seen at higher doses. Risk of hematological complications was 26% in one study with a dose of 600 mCi. High doses need stem cell harvest to be performed before ablation.

Conclusion: I-131 MIBG ablation therapy can be considered in a patient with metastatic malignant pheochromocytoma which is not amenable to surgery. However more data and clinical trials are needed.