G110 Pheochromocytoma Causing Unexpected Death – Two Unusual Presentations

Jacqueline L. Parai, MD, Ontario Forensic Pathology Services, Division of Anatomical Pathology, The Ottawa Hospital, 501 Smyth Road, Box 117 4th Floor, Ottawa, ON K1H 8L6, CANADA; and Iris Teo, MD, Itrat Ahmed, MD, and Christopher M. Milroy, MD, The Ottawa Hospital, 501 Smyth Road, Box 117 4th Floor, Ottawa, ON K1H 8L6, CANADA*

After attending this presentation, attendees will be able to appreciate the postmortem pathology of pheochromocytomas and sudden death.

This presentation will impact the forensic science community by presenting the medicolegal significance and varied clinical presentation of adrenal and extraadrenal pheochromocytomas including sudden death, their histological features and the possible genetic implications of their diagnosis.

Pheochromocytomas are rare tumors of paraganglionic tissue. Paraganglionic tissue is distributed throughout the body and tumors may occur in multiple sites. Patients may present with severe headaches, nausea, excessive sweating, palpitations due to tachycardia and anxiety, tremors, pain in the lower chest and upper abdomen, and weight loss. These symptoms are due to the fact these tumors produce, store and secrete catecholamines (epinephrine, norepinephrine). Patients typically have hypertension, which may be intermittent. Clinical diagnosis is made by urinary and plasma catecholamine measurement, along with imaging. The tumors may present in the adrenal medulla and extra- adrenal sites. They may rarely be associated with sudden death, and catecholamine induced damage to the myocardium may be present, as the so called catecholamine cardiomyopathy.

Two cases of sudden death due to pheochromcytomas are presented. Both patients were 34-year-old males. In the first case, the male presented with abdominal pain. An ECG showed left bundle branch block and changes of an inferior myocardial infarction. He was variably hypertensive and hypotensive. He went into cardiac arrest while undergoing radiological investigation. At autopsy there was a suprarenal mass measuring 8.5 x 7.5 x 4.5 cm, with 700 mL of blood in the peritoneal cavity along with retroperitoneal hemorrhage. On histology, the tumor had the characteristic appearance of a pheochromocytoma. There were typical Zellballen. The tumor cells stained positively with neuroendocrine markers including chromogranin and synaptophysin. The supporting sustentacular cells showed some S100 positivity.

In the second case, the male had a witnessed collapse and died unexpectedly. He had been diagnosed as a non-insulin dependent diabetic five days previously. On the day of his death, he was described as well and his glucose level had been measured within the normal range. At autopsy a mass was found adjacent to the kidney but below the adrenal gland 3.5 cm in diameter. Histology showed the characteristic appearance of a pheochromocytoma.

Pheochromocytomas secrete catecholamines which cause hypertension. They also modify glucose metabolism, which accounts for the hyperglycemia seen in the second case. Diagnosis depends on histology and the characteristic immunohistochemical appearance. Malignancy cannot be reliably diagnosed by morphological features. These tumors may also be associated with genetic syndromes – such as multiple endocrine neoplasia (MEN) syndromes. Although traditionally known as the 10% tumor because 10 % are extraadenal and 10% malignant, a higher proportion of the cases, of the order of 25%, are malignant when associated with familial syndromes.

In the two cases, their clinical presentations alone may have resulted in their deaths being erroneously attributed to more common causes of sudden death, such as hypertensive or atherosclerotic cardiovascular disease. An appreciation of the clinical and pathological features of pheochromocytomas however properly diagnoses these cases. As such, surviving relatives can be informed and screened.

Pheochromocytoma, Sudden Death, Adrenal

 ** Presenting Author*