G87 A Rare Case of Cardiac Failure Due to Hypertensive Crisis in Pheocromocytoma: A Methodological Approach for Diagnosis

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The goal of this presentation is to present a rare case of cardiac failure due to hypertensive crisis in pheocromocitoma in an asymptomatic 25-year- old young man is presented. The rarity of pheocromocytoma makes the case peculiar and the complete pathologic investigation adopted (autopsy performing, immunoistochemical staining, and biochemical screening) is strongly recommended to pathologists to confirm diagnosis.

This presentation will impact the forensic science community by demonstrating how the rarity of pheocromocytoma makes the case peculiar. It is strongly suggest, in these cases, the relevance for pathologists of a complete methodological approach, integrating clinical data by means of autopsy findings, immunoistochemical staining and biochemical screening to confirm diagnosis.

Pheocromocytomas are rare but clinically important tumours of chromaffin cells that produce, store, release and metabolize catecholamines. Pheocromocytomas usually manifest clinically as hypertension which can be sustained or paroxysmal. The diagnosis of pheocromocytoma is based on measuring excessive amounts of catecholamines and their metabolites on blood and urine; more than 90% of patients with pheocromocytoma have elevated levels of catecholamines, metanephrine, and vanillyl-mandelic acid. Sensitivity and specificity of these measurements are 91%. Failure to diagnose the tumours can result in sudden, unexpected and potentially lethal complications; cause of death in these cases is generally a consequence of paroxysmal hypertension as well as cerebral vascular accidents, abrupt haemorrhage into the tumour or acute left ventricular failure.

A 25-year-old man, with a past medical history significant for recurrent episodes of cephalalgia, was transported to the local Hospital at 11:44 p.m. complaining of vomiting and headache. A prescription for symptomatic treatment was issued unsuccessfully. He had high blood pressure (180/80) and tachycardia (110 bpm). Neurological examination was unremarkable, abdomen showed no rigidity, peristaltic sounds were normal. Initial laboratory findings showed hyperglycaemia (177 mg/dl) and high level of amylase (125 U/L); further findings showed high levels of myoglobin (153 ng/ml). A 12 lead electrocardiogram on admission was performed showing sinus tachycardia, with right bundle branch block and ventricular bigeminy extra systoles; non specific repolarization change were also described. Few hours later, 8:15 a.m. the patient appeared pale and sweating; blood pressure was unappreciable and hypocontractility of left ventricle with low ejection fraction was observed on echocardiography (EF 25-30%). A state of haemodynamic shock was declared (FC 170, blood pressure was unappreciable). Pulmonary edema was observed on chest Rx examination and oro-tracheal intubation was performed substaining ventilation in intensive care unit. At 9:15 a.m. ECG monitor showed cardiac arrest; resuscitation manoeuvres were attempted unsuccessfully.

A complete postmortem examination was performed two days after death. External examination was unremarkable except for food residuals in were recorded at oesophagus exploration; heavy lungs presenting white foam on the main bronchi was also detected. Heart was fixed in formalin, cardiac size was normal, with conical shape. Macroscopic study (cut in cross-section 3 mm intervals) of coronary arteries was unremarkable. A well circumscribed encapsulated lobulated reddish and brownish suspected lump measuring 3.5x3x3 was attached to the medial aspect of the left kidney; it was soft on section and presented aspect of necrosis and haemorrhage. Adrenal tissue was attenuated over the upper part of the mass; aspect of minimal haemorrhage was observed on pancreas examination.

Histological examination revealed polyvisceral stasis, mild cerebral edema: massive pulmonary edema was recorded. Cardiac myofibers varied considerably in size with many large fibers and aspect of fibrosis suggesting for hypertension; the pathological myocardial picture included fragmentation of the whole myocyte (pancellular lesion) which ranged from early break- down in pathological band (intense hyperosinophilia of the hypercontracted myocardial cells with rexis of the myofibrillar apparatus into cross-fiber, anomalous and irregular) to a total granular disruption (myofibrillar degeneration). Histological examination of the suspected lump addressed diagnosis for a benign pheocromocitoma with the presence of well-defined nests (Zellballen) bound by a delicate fibrovascular stroma, which contain amyloid. The cells varied considerably in size and shape and had purely granular basophilic cytoplasm; the nuclei were round with prominent nucleoli.

An immunoistochemical study was performed and it showed a positive reaction for chromogranin and synaptophysin.

Dosage of catecholamines and their metabolites on a blood and urine samples was performed showing high levels of catecholamines, metanephrine and vanillyl-mandelic acid.

Clinical data, autopsy findings, data collected from immunoisto- chemical staining and biochemical analysis led us to conclude that cardiac failure due to hypertensive crisis in adrenal pheocromocytoma was the cause of death.

In conclusion, the rarity of pheocromocitoma makes the case peculiar. The authors strongly suggest, in these cases, the relevance for pathologists of a complete methodological approach, integrating clinical data by means of autopsy findings, immunoistochemical staining and biochemical screening to confirm diagnosis.

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Pheocromocitoma, Cardiac Failure, Hypertensive Crisis

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