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A young man with acute onset shortness of breath: a case of phaeochromocytoma with normal urinary metanephrines

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Introduction: Phaeochromocytomas are neuroendocrine tumors of the adrenal medulla or extra-adrenal chromaffin tissue. Diagnosis of phaeochromocytoma is based on measurement of catecholamines and metanephrines in plasma or through a 24 hour urine collection. Plasma and urinary catecholamines can be normal in episodically secreting tumours, when measured between paroxysmal attacks.

Case report: A 38 year old patient was admitted with acute onset of shortness of breath of 3 days duration, associated with orthopnoea. On examination there were bilateral extensive fine inspiratory crepitations in lung bases. His BP was 170/100 mmHg. Cardiac apex was heaving in nature and shifted to left.

His ECG and 2D echocardiogram revealed left ventricular hypertrophy. His ejection fraction was 42%. Ultra sound scan and contrast CT of the abdomen showed a solid mass compatible with a left suprarenal tumour. His 24 hour urinary metanephrines were 8.9 micrograms (25-312). Phaeochromocytoma was diagnosd, despite having normal urinary metanephrines. This could be explained by the fact that certain phaeochromocytomas secrete catecholamines only intermittently. In such patients phaeochromocytoma can exist with normal urinary metanephrines.

Discussion: He was started on antihypertensives. Resection of the tumour was done under the cover of alpha blockers at National Hospital of Sri Lanka and histology of the tumour confirmed the diagnosis of phaeochromocytoma. This was one of the rare cases of a symptomatic phaeochromocytoma without elevated urine catecholamines and metanephrines. Plasma free metanephrines would be the ideal test for biochemical detection of the tumour. But tumours can exist even with normal metanephrines.