The Challenges of a Dopamine Secreting Paraganglioma

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HISTORY
• 39 year old female
• 18 month history of borderline hypertension, headaches, palpitations and some anxiety symptoms
• Family history of hypertension in both parents

EXAMINATION
• BP 160/102
• No medications
• Large single cafe au lait spot
• No neurofibromata
• Remaining examination unremarkable

URINARY CATECHOLAMINES

<table>
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<tr>
<th>Date</th>
<th>Volume (mls)</th>
<th>Dopamine (nmol/24h)</th>
<th>Noradrenaline (nmol/24h)</th>
<th>Adrenaline (nmol/24h)</th>
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IMAGING
Meta-iodobenzylguanidine (MIBG I-123) scanning indicated a single focus of activity further defined on SPECT-CT as a 12 mm mass in the lower para-aortic region

FURTHER INVESTIGATIONS
• Calcium 2.83mmol/L (N = 2.2-2.6)
• PTH 274pg/ml (N = 15-70)
• Calcitonin <5.0ng/L
• Nuclear uptake scan indicated a right lower parathyroid focus

MANAGEMENT
• BP controlled with amlodipine and lisinopril prior to surgery
• After a 30 second asystolic episode during first manipulation of the tumour a black lobulated paraganglioma was removed at the organ of Zuckerkandl
• Urinary dopamine normalised post-operatively (2147nmol/24h) and blood pressure settled
• Right lower parathyroid adenoma was removed uneventfully 5 months later and calcium normalised (2.52mmol/L)
• Antihypertensives have now been fully withdrawn
• Genetic testing to date has been negative including MEN2a, succinate dehydrogenase B, C and D mutations

DISCUSSION
• Dopamine secreting paragangliomas are extremely rare and are usually metastatic at diagnosis
• They are usually associated with nonspecific symptoms, normotension and present with mass effects
• Alpha blockade is contraindicated because of its association with cardiovascular collapse which is due to the unopposed hypotensive action of dopamine when the pressor catecholamines are blocked
• They are less likely to enhance with MIBG scanning
• Lifelong surveillance is recommended given high recurrence and malignancy rates
• To our knowledge this is the first reported case of a dopamine secreting paraganglioma presenting with a coexistent parathyroid adenoma

USUAL CAUSES OF RAISED URINARY Dopamine LEVELS
• Over- collection
• Tricyclic Antidepressants
• Levodopa
• Drugs containing adrenergic receptor agonists (eg decongestants)
• Amphetamines
• Buspirone and most psychoactive agents
• Prochlorperazine
• Reserpine
• Withdrawal from clonidine and other drugs
• Ethanol