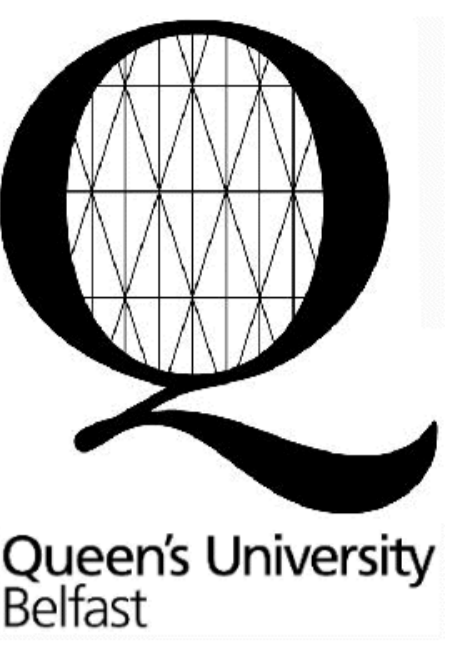


The Challenges of a Dopamine Secreting Paraganglioma

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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

	Volume (mls)	Dopamine (nmol/24h)	Noradrenaline (nmol/24h)	Adrenaline (nmol/24h)
Normal		300-3900	50-560	5-120
10/12/2009	2300	8273	265	35
24/12/2009	2600	6934	367	23
05/05/2010	2200	8653	240	26
06/05/2010	2600	5398	120	16

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

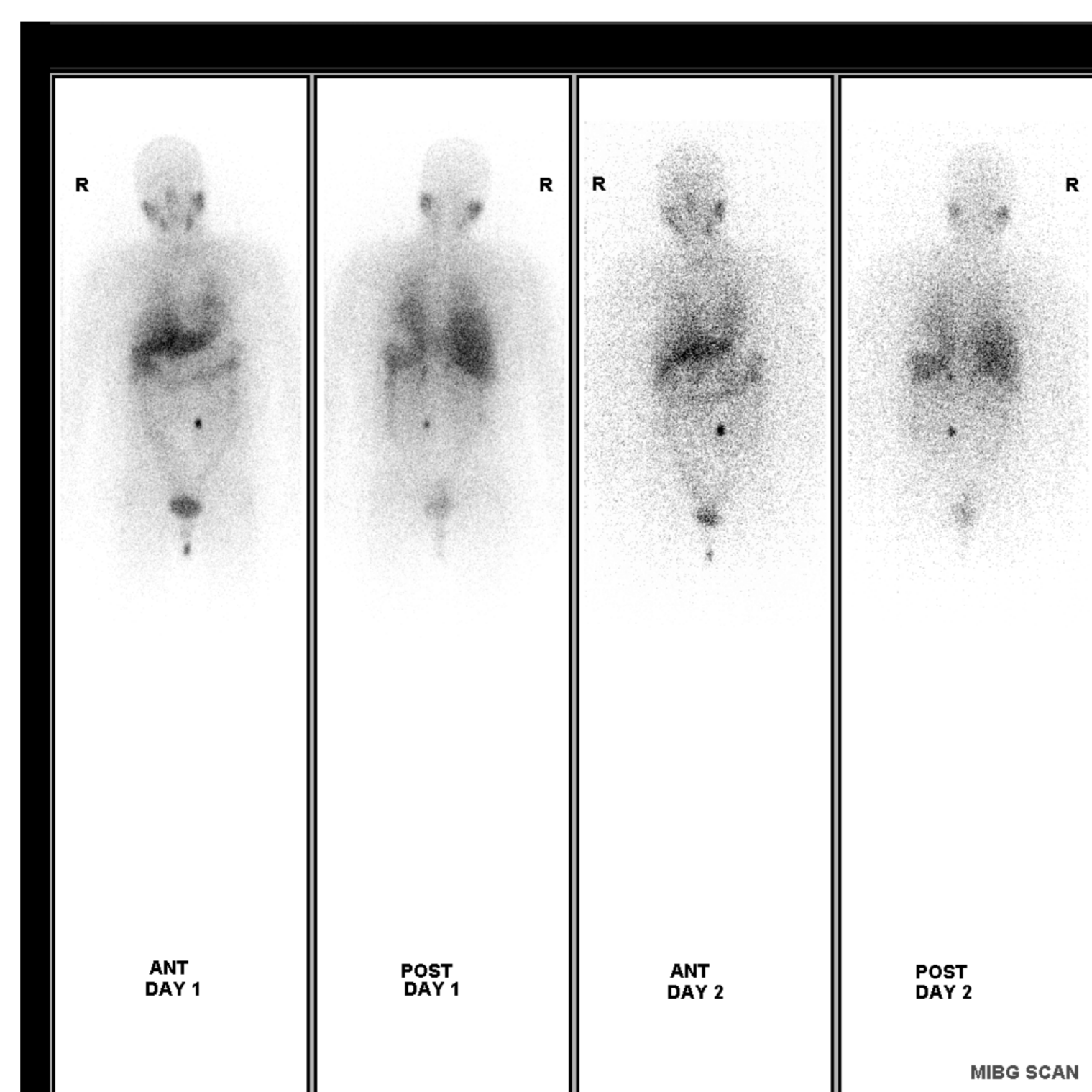


Fig 1. MIBG scan showing lower left para-aortic focus

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

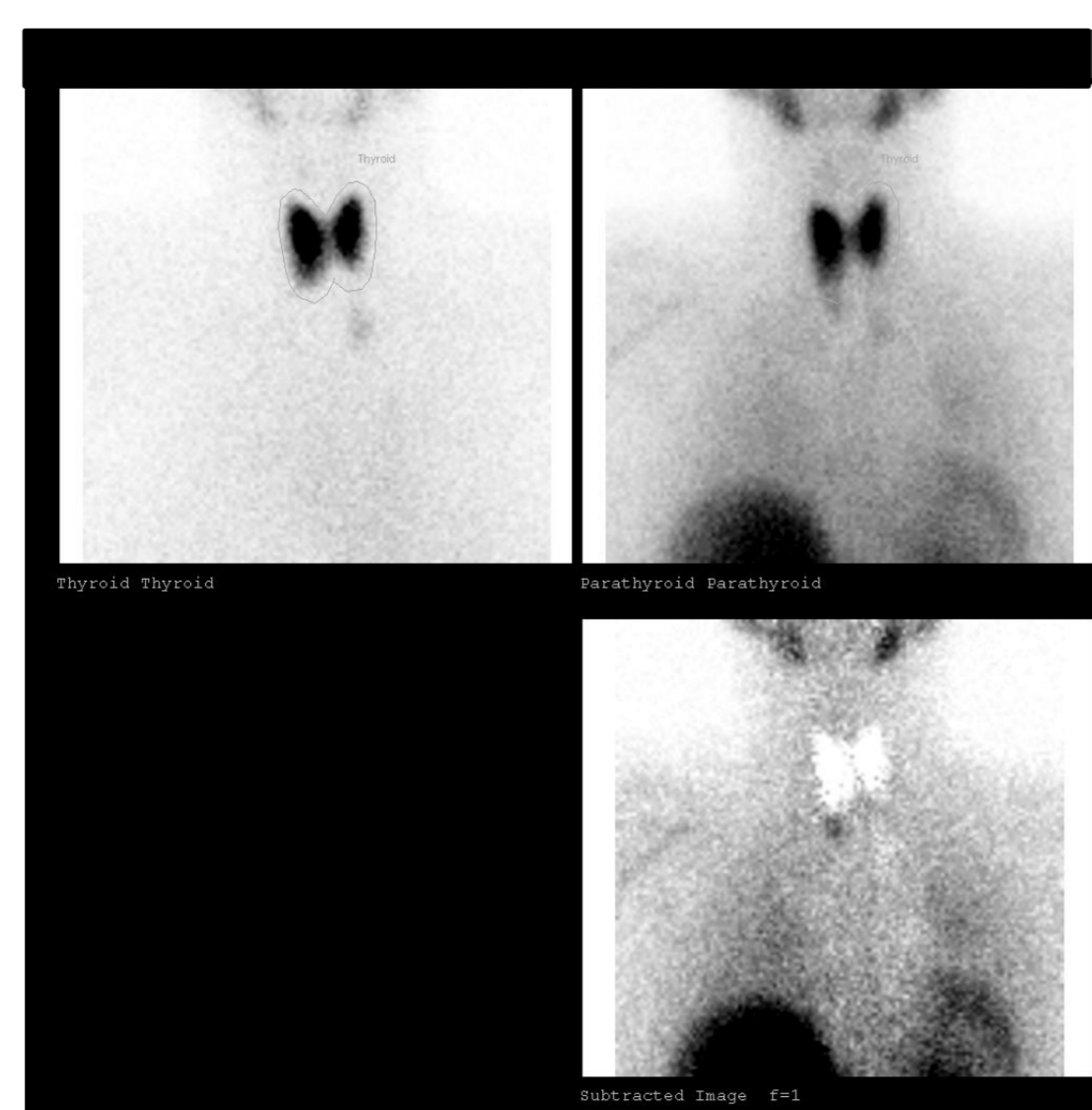


Fig 2. Parathyroid uptake scan showing a right lower parathyroid focus

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- Antihypertensives have now been fully withdrawn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

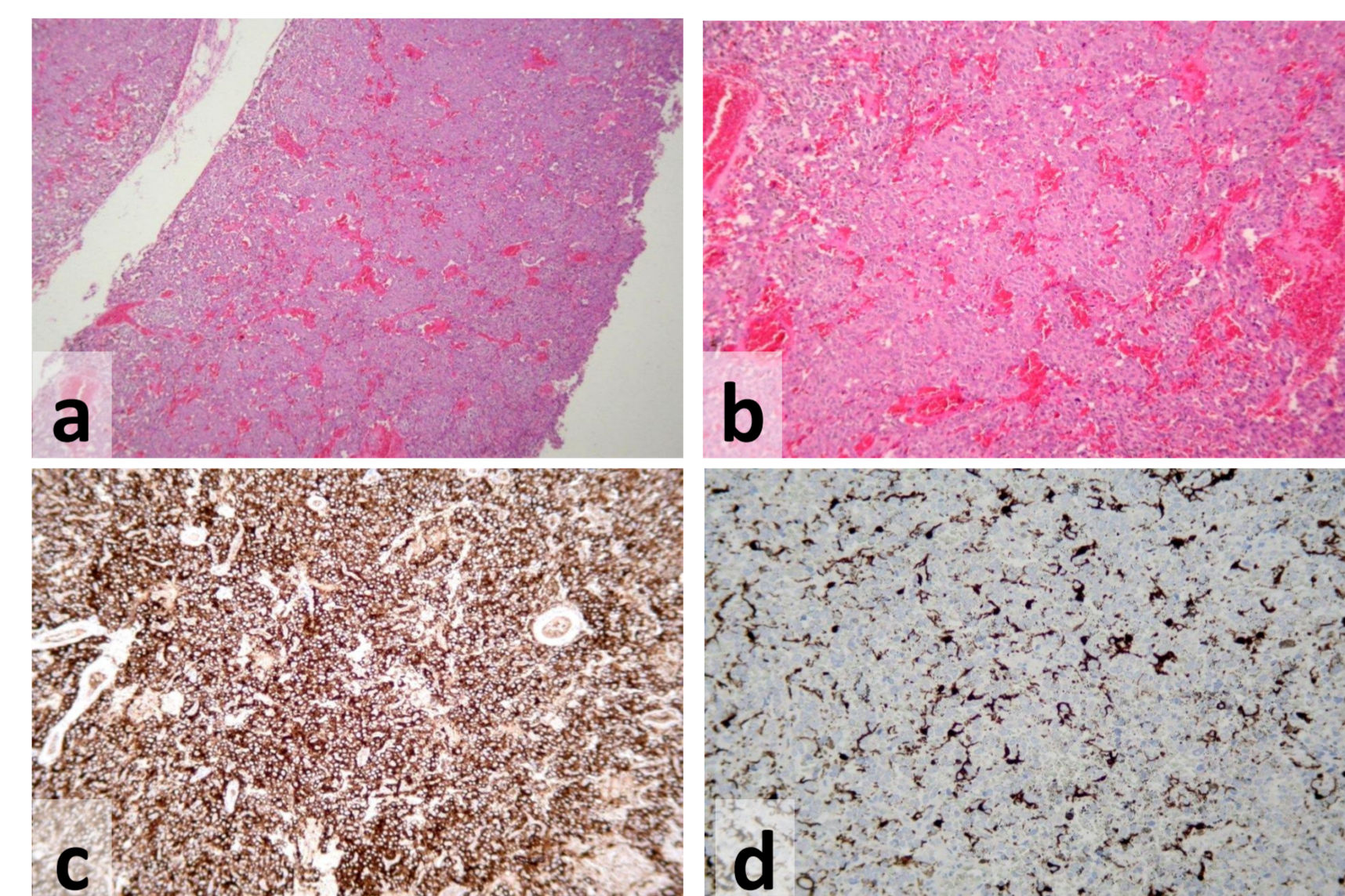


Fig 3. Histology from this patient's paraganglioma (a) and (b) H&E stain; (c) chromogranin A staining diffusely positive; (d) S100 stains the sustentacular cells

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

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