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Phaeochromocytoma in childhood: radiological-pathological correlation

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Appendix 4 - Data Collection Table

CASES FROM GREAT ORMOND STREET HOSPITAL

Pat. No.	Age	Date of surgery	Sex	Tumour histology	Clinical presenting complaint	Operative findings	MR features	CT features	U/S features	Nuclear Medicine	Out-come and FU
1	10	23/10/ 2000	M	4cm nodule, smooth surface, haemorrhagic centre. Marked trabecular pattern, associated paraspinal ganglioneuroma Capsular invasion	Headaches, dizziness and hypertension. High urinary VMA. ? MEN Thyroid lesion	Right adrenal tumour 4cm draining directly into IVC Paravertebral mass 6cm posterior to hilum of right lung		Adrenal tumour and right paraspinal mass CT thorax and abdo in Greece		MIBG pos right adrenal but not right paraspinal	
2	8	17/04/ 2003	F	Bladder nodule, 3cm diameter, classical 'nested' architecture, local invasion, lymph node metastases. Capsular invasion.	Haematuria. Cystoscopy showed a bladder neck tumour. Raised urinary noradrenaline and dopamine. BP normal. No flushing, sweating, palpitations	Adjuvant chemotherapy assisted excision – more mobile. Tumour adherent to pelvic wall	31/3/2003 Int low T2 Heterogeneous Low T1 Minimal to mild enhancement (STIR low)	CECT 7/5/2003 Peripheral enhancement. 3cm bladder tumour	6/5/2003 Good image correlates with CT findings Thicking of postrior bladder wall	MIBG pos	
3	14	03/06/ 2003	M	Adrenal with 6cm nodular mass, cystic and haemorrhagic, classical nested architecture, no capsular invasion	Acute onset severe right iliac fossa pain, thought to be appendicitis. Regular migraines, sweating at night, recent lethargy	Large tumour adherent to IVC, Aorta and inferior mesenteric artery		Peripheral enhancement. Central low density extracapsular Possible haemorrhage	20/5/2003 Internal septation with increased perinephric echos signifying a possible bleed. Peripheral vascularity noted Ultrasound found large right suprarenal mass 6x7cm	MIBG pos	
4	14	20/09/ 1990	F	Adrenal with a 4x3cm nodule, well circumscribed., classical nested architecture with no invasion	Nausea and vomiting, weight loss, tiredness chronic abdominal pain. Raised urinary catecholamines.	Right adrenal tumour					Lost

Pat. No.	Age	Date of surgery	Sex	Tumour histology	Clinical presenting complaint	Operative findings	MR features	CT features	U/S features	Nuclear Medicine	Outcome and FU
5	9	07/04/ 1994	M	Bilateral adrenal glands with tumours 5cmx2.5cm and two separate tumours in the left 0.5 and 0.2cm in diameter, encapsulated, classical nested architecture with mild nuclear pleomorphism but no invasion							
6	13	02/01/ 1997	M	Left paranephric, para-aortic tumour, 4cm diameter, nested architecture with thick fibrous bands, and local capsular invasion. Haemorrhage.	Nausea, weakness, vomiting after strenuous exercise. Sweating and cold extermeties. BP normal. Noradrenaline high in urine. 8 month delay in diagnosis	Tumour in and around left renal pedicle crossing the midline over the major vessels. Very vascular. Left renal vein in tumour. Left kidney sacrificed. Large Meckel's diverticulum excised					
7	7	18/04/ 2006	M	Left Renal hilum tumour, 3cm, encapsulated, nested architecture, no capsular invasion or pleomorphsim	Headaches, associated vomiting and eye pain, sweaty 2/12. Hypertensive at GP. Slightly elevated Urinary VMA.	Difficult resection, tumour surrounding renal vessels, very friable, not encapsulated.	T1 low heterogeneous STIR high ADC low Delayed peripheral, early intense enhancement	mass	Hypoechoic mass missed on original us	MIBG pos	
8	9	03/04/ 2001	M	Adrenal gland with 4cm nodule, classical nested architecture, no capsular invasion	Focal seizure Hypertension, 2/12 headaches, vomiting, Urine and plasma catecholamines positive.	Right adrenalectomy with tumour extending behind IVC			Adrenal mass on ultrasound		

CASES FROM THE JOHANNESBURG ACADEMIC HOSPITALS COMPLEX

Pat. No.	Age	Date of surgery	Sex	Tumour histology	Clinical presenting complaint	Operative findings	MR features	CT features	U/S features	Nuclear Medicine	Outcome and FU
9	14	20/09/ 2002	M	5 cm extra adrenal mass. Typical Zellballen type growth pattern and nested architecture. Capsular invasion noted. Multifocal tumour necrosis and haemorrhage. Liver deposit confirmed metastesis Left adrenal normal Later had lung mets	Complex congenital cardiac abnormality. Developed hypertension and palpitations discovered during routine follow up. Raised noradrenaline Adrenaline normal Hypertensive			3.7 x 4cm enhancing mass in mid abdomen. Suspicious L adrenal mass. Right lobe of liver mass in Segment 6 and 7 suspicious for metastasis.	Liver normal Lobulated mixed echogenic vascular mass mid abdomen anterior to aorta and IVC 3.2x3x3.7cm Adrenal mass 4x4 cm	MIBG pos	Hpt on Rx Followed up 83 months Lost to follow up, presumed RIP
10	9	28/08/ 2000	F	Two nodules in right adrenal gland, largest 3.5cm. Zellballen nested architecture. Capsular invasion. Focal haemorrhage and calcification within tumour.	Severe cardiogenic shock Recurrent hypertensive episodes Urine VMA positive			Mass in right adrenal gland on CT		MIBG pos	
	12	16/05/ 2005 Metachr onous	F	4cm tumour in left adrenal Zellballen nested architecture. Mild pleomorphism and mitoses. Cystic degeneration and focal haemorrhage. Capsular invasion.	Hypertensive on routine followup	Exploratory laparotomy 5x4cm left adrenal mass adherent to the diaphragm. Para-aortic mass not removed.		3x3.4x5.2cm L adrenal mass 6.4x3.8x5.6 cm paraaortic retropancreatic mass with central necrosis	24/3/2005 5x3.4x5 cm mass anterior to aorta no adrenal mass seen	MIBG pos	Metastatic on ant hypertensive's. Multiple therapeutic MIBG's HPT but well 134 months

Pat. No.	Age	Date of surgery	Sex	Tumour histology	Clinical presenting complaint	Operative findings	MR features	CT features	U/S features	Nuclear Medicine	Outcome and FU
11	11	12/08/ 2002	M	Right sided retroperitoneal and intracaval tumour. Mild to moderate nuclear pleomorphism and mitoses. Lobulated. Nested architecture with necrosis Only pieces removed. Extraadrenal paraganglioma. Capsular invasion	2 year history. Loss of weight, feeling hot, sweating, intermittent vomiting, headaches, stomach pain. Hypertensive. High urinary VMA	Unresectable tumour >5cm involving IVC and right renal vein. Retroperitoneal pale tumour with surrounding fibroblastic reaction. Tumour extending from bifurcation to just below hepatic veins and into renal vein. Tumour debulked.		Encasement of the aorta	4x3cm Paraaortic extension displacing pancreas anteriorly and medially. IVC attenuated	MIBG pos	Well HPT on Rx 110 months
12	13	29/10/ 2009	M	L adrenal tumour 35x30x30mm no necrosis no hgg Zellballen No capsular invasion Normal mitotic activity	HPT + papiloedema High urinary VMA LVH on echo Presented with acute gastro, ?seizure, renal artery bruit. Possible renal artery stenosis secondary to the mass	Left adrenal tumour. Uneventful surgery.	Left adrenal mass seen on MRA which was arranged to investigate HPT. 6/10/2009 at Bara, Normal proximal renal arteries	22/9/2009 missed the adrenal mass. Small L renal infarcts	3.5x2.5cm L suprarenal mass 9/10/2009	MIBG pos	Well 24 months
13	12	7/12/ 2009	F	55x35x25mm L adrenal mass no necrosis, no other features of malignancy declared benign. Zellballen Capsular invasion	Dizziness constipation, night sweats, LOW, headaches, Blurred vision, polyuria. Admitted for HPT crisis and acute abdominal pain LVH on echo HPt High urinary VMA	L adrenal tumour stuck down to surrounding structures, difficult surgery. Renal vessels preserved. Aggressive looking.		2x2 cm hyperdense L adrenal mass. No contrast given. 16/11/09 at Sebokeng		MIBG pos	Well 22 months

Pat. No.	Age	Date of surgery	Sex	Tumour histology	Clinical presenting complaint	Operative findings	MR features	CT features	U/S features	Nuclear Medicine	Outcome and FU
14	8	12/11/ 2010	M	50x30x25mm Left adrenal mass necrosis hgg Zellballen Encapsulated no local invasion Moderate pleomorthism and abundant cytoplasm	Seizures, status epilepticus. Thought to be ADEM on MRI as BP not originally checked. Hpt in ward later and abdo sonar revealed paraaortic mass. Urinary VMA and serum adrenaline/nor adrenaline high. CT showed basal meningeal enhancement. LP showed high protein to thought to be TBM	Well encapsulated L adrenal tumour. Uneventful laparoscopic removal	22/10/2010 Of Brain showed high FLAIR signal in deep white matter	21/10/2011 22/10/2011 26x33mm left adrenal mass. Irregular thick wall with necrotic centre. Separate from surrounding structures	Left paraaortic mass	MIBG neg	Well not hypertensi ve 11 months
15	11	29/08/ 2011	M	50x35x15mm central hgg and necrosis Zellballen Pseudocapsule Extracapsular invasion	Hypertension, Nausea vomiting, headaches, seizures	Open laparotomy 8cm mass arising from R Adrenal Encasing IVC Encasing Aorta Local invasion Incompletely removed	Iso T1WI Iso with high intensity foci T2WI Retrocaval attenuating IVC. Renal vein and artery abutted but not invaded	Retrocaval Right adrenal masss attenuating ivc 60x29x30mm	R paraaortic mass Mixed echogenicity 47x37x25mm	Ga 68 DOTATO C PET/CT avid uptake	Well after 2 months

APPENDIX 5 – Data Summary Table

Patient number	Tumour number	Year of surgery	Age at diagnosis	sex	Hypertensive at time of presentation	Clinical Symptoms	Syndromic	Urinary metabolites
1	1	2000	10	M	-	Headaches, Dizziness	Thyroid lesion ? MEN	Yes
2	2	2003	8	F	No	Haematuria. No symptoms of catecholamine excess	No	Yes
3	3	2003	14	M	-	headaches, Right iliac fossa pain, night sweats, lethargy	No	-
4	4	1990	14	F	-	Nausea, vomiting, weight loss, lethargy, abdominal pain	No	Yes
5	5	1994	9	M	-	-	-	-
6	6	1997	13	M	No	Nausea, vomiting, weakness, sweating, cold peripheries	No	Yes
7	7	2006	7	M	Yes	Headaches, vomiting, sweating, eye pain	No	Yes
8	8	2001	9	M	Yes	headaches, Focal seizure, vomiting	No	Yes
9	9	2002	14	M	Yes	Complex congenital cardiac abnormality, palpitations discovered on routine follow up	No	Yes
10	10	2000	9	F	Yes	Severe cardiogenic shock	No	Yes
	11	2005	12	F	Yes	Hypertensive on routine follow up	No	-
11	12	2002	11	M	Yes	headaches, abdominal pain sweating, Loss of weight, vomiting,	No	Yes
12	13	2009	13	M	Yes	Seizure, Diarrhea and abdominal pain renal artery bruit, renal artery stenosis secondary to the mass	No	Yes
13	14	2009	12	F	Yes	Dizziness, constipation, night sweats, loss of weight, headaches, abdominal pain, blurred vision	No	Yes
14	15	2010	8	M	Yes	Seizures	No	Yes
15	16	2011	11	M	Yes	Nausea, Vomiting, Headaches and Seizures	No	Yes

Patient number	Tumour number	Anatomical location	Left or right adrenal	Local invasion noted by surgeon	Distant metastases time of diagnosis	Largest dimension on histology	Largest dimension on cross sectional imaging	Classical Histology	Capsular invasion on histology
1	1	Adrenal	right	No	No	4cm	-	Yes	Yes
2	2	Bladder	-	Yes	Lymph nodes positive	3cm	3cm	Yes	Yes
3	3	Adrenal	Right	Yes	No	6cm	7cm	Yes	No
4	4	adrenal	Right	No	-	4.5cm	-	Yes	No
5	5	adrenal	Bilateral (two tumours in left adrenal and one in right adrenal)	-	-	Right 5cm Left 0.5cm Left 0.2cm	-	Yes	No
6	6	Renal hilum	-	Yes	-	4cm	-	Yes	Yes
7	7	Renal hilum	-	Yes	No	3cm		Yes	No
8	8	Adrenal	Right	No	No	4cm	-	Yes	No
9	9	Mid abdominal para-aortic	-	-	Yes	5cm	4cm	Yes	Yes
10	10	Adrenal	Right	-	No	3.5cm	-	Yes	Yes
	11	Adrenal	Left	Yes	Yes	4cm adrenal mass	5cm adrenal mass with 6cm para-aortic extension	Yes	Yes Para aortic extension not removed
11	12	Para aortic	-	Yes	No	Only pieces removed. Total tumour larger than 5cm	5.4cm	Yes	Yes
12	13	Adrenal	Left	No	No	3.5cm	3.5cm	Yes	No
13	14	Adrenal	Left	Yes	No	5.5cm	2cm	Yes	Yes
14	15	Adrenal	Left	No	No	5cm	3cm	Yes	No
15	16	Adrenal	Right	Yes	No	5cm	6cm	Yes	Yes

Patient number	Tumour number	Anatomical location	Ultrasound	MDCT	MRI	MIBG
1	1	Adrenal	-	Adrenal tumour	-	Positive
2	2	Bladder	Thickening of posterior bladder wall	Peripheral enhancement	Low signal T1WI, T2WI and STIR Mild enhancement	Positive
3	3	Adrenal	Adrenal mass with septations with perinephric echogenicity thought to be haemorrhage	Adrenal mass Peripheral enhancement Central hypodensity	-	Positive
4	4	Adrenal	-	-	-	-
5	5	Adrenal	-	-	-	-
6	6	Renal hilum	-	-	-	-
7	7	Renal hilum	Hypoechoic renal hilum mass[a]	Extra adrenal mass	Low signal T1WI High Signal T2WI High signal STIR Intense enhancement	Positive
8	8	Adrenal	Adrenal mass	-	-	-
9	9	Mid abdominal para-aortic	Mixed echogenic adrenal mass[b] and para-aortic mass Liver normal on Sonar[c]	Enhancing para-aortic mass Adrenal mass [b] Hepatic mass	-	Positive
10	10	Adrenal	-	Adrenal mass	-	Positive
	11	Adrenal	Para-aortic mass No adrenal mass[d]	Adrenal mass and para-aortic extension with central necrosis	-	Positive
11	12	Para aortic	Para aortic mass 4x3cm	5.4cm Para-aortic mass heterogeneous density abutting the aorta. Involves and IVC and right renal vein	-	Positive
12	13	Adrenal	Adrenal mass	Adrenal mass not seen[e]	Adrenal mass iso on T2WI Limited MRI performed to exclude renal artery stenosis	Positive
13	14	Adrenal	-	Adrenal mass hyperdense (non contrast)	-	Positive
14	15	Adrenal	Para-aortic mass Normal adrenal gland[f]	Adrenal mass hypodense center	-	Negative
15	16	Adrenal	Para-aortic mass, mixed echogenicity	Adrenal mass, attenuating IVC	Iso T1WI Iso T2WI and foci of high signal	Ga68 DOTATOC PET/CT positive

a. Renal hilum mass was missed on first ultrasound investigation but was picked up in a subsequent ultrasound test.
b. The left adrenal gland was assessed as normal at surgery however review of the MDCT confirms that the adrenal gland appears to be enlarged *Fig 4*.1
c. Liver metastases were missed on Ultrasound
d. falsely normal sonar of the adrenals
e. falsely normal on CT
f. falsely normal adrenal gland on Ultrasound

Patient number	Tumour number	Anatomical location	Follow up period in months until October 2011	Outcome	Hospital *
1	1	Adrenal	-	-	GOSH
2	2	Bladder	-	-	GOSH
3	3	Adrenal	-	-	GOSH
4	4	adrenal	-	-	GOSH
5	5	adrenal	-	-	GOSH
6	6	Renal hilum	-	-	GOSH
7	7	Renal hilum	-	-	GOSH
8	8	Adrenal	-	-	GOSH
9	9	Mid abdominal pre-aortic	83	Hypertensive for until last clinic visit. Presumed diseased	CMJAH/CHB
10	10	Adrenal	-	Hypertension at f/u prompted re-evaluation for possible metachronous tumour	CMJAH/CHB
	11	Adrenal	134	Hypertensive with known secondary deposits	CMJAH/CHB
11	12	Para-aortic	110	Hypertensive on treatment	CMJAH/CHB
12	13	Adrenal	24	Well and not hypertensive	CMJAH/CHB
13	14	Adrenal	22	Well and not hypertensive	CMJAH/CHB
14	15	Adrenal	11	Well and not hypertensive	CMJAH/CHB
15	16	Adrenal	2	Well and not hypertensive	CMJAH/CHB

* GOSH Great Ormond Street Hospital
CMJAH Charlotte Maxeke Johannesburg Academic Hospital
CHB Chris Hani Baragwanath Hospital