

Phaechromocytoma during pregnancy: Ultrasound and MRI appearances

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Introduction

Phaechromocytoma is an uncommon cause of hypertension during pregnancy and is rarely diagnosed before delivery. Phaechromocytoma in pregnancy not diagnosed before onset of labour is associated with 48% maternal death rate and a 54% fetal death rate¹. The maternal mortality has declined considerably, 10%² when diagnosis is made earlier in pregnancy and recently to zero³ and fetal mortality to 30%². We present a case of phaechromocytoma diagnosed in the first trimester using ultrasound and MRI.

Case Report

A 24 year old pregnant Filipino woman gravida two, para-one was admitted at six weeks gestation when a blood pressure of 180/120 mm Hg was measured at the antenatal clinic. She was a known hypertensive for the past one year and was on anti-hypertensive treatment. Her blood pressure was liable with blood pressure ranging from 120/180 to 180/120. Her first pregnancy resulted in a miscarriage at six weeks gestation and was apparently associated with problems of blood pressure control. The second pregnancy again resulted in rises in blood pressure for which her local general practitioner investigated with a fetal ultrasound examination. Sonography revealed a normal intrauterine pregnancy and incidental finding of 4 x 3 cm left suprarenal mass.

In retrospect she gave an unimpressive history for paroxymal catecholamine release, admitting only to infrequent headaches and dizziness. There was no history of diabetes or renal disease. Her grandfather had hypertension and ischaemic heart disease.

On examination, her pulse rate was 84/min, regular in time and amplitude and the blood pressure was 170/100 mm Hg. There was no associated oedema nor proteinuria on urinalysis. A provisional diagnosis of phaechromocytoma was made. Biochemistry was normal apart from a protein of 85 gm/L (normal 63 – 78) and a 24 hour urine collection revealed elevated noradrenaline, normetanephrine and VMA levels. Adrenaline and metanephrine levels were normal. Repeated ultrasonography (Fig 1 A & B) demonstrated a hypochoic mass arising between two limbs of the left adrenal gland. The mass was separated from the left kidney, spleen and pancreas.

An MRI scan (Fig 2 A,B & C) performed at Royal North Shore Hospital revealed a grossly enlarged left adrenal gland. The left adrenal mass demonstrated low Phaechromocytoma during pregnancy : Ultrasound & MRI appearances signal intensity on the T1-weighted images and high signal intensity

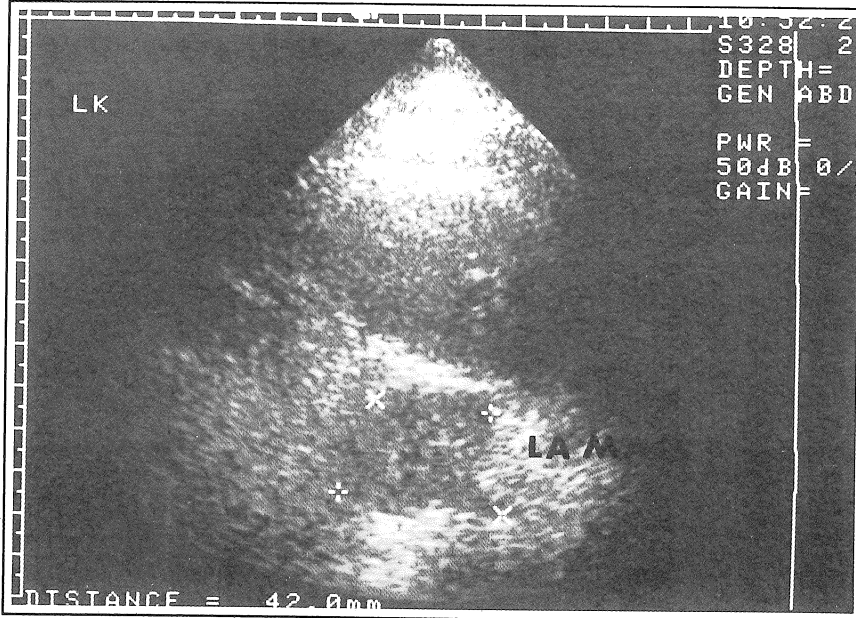


Fig. 1 (a)

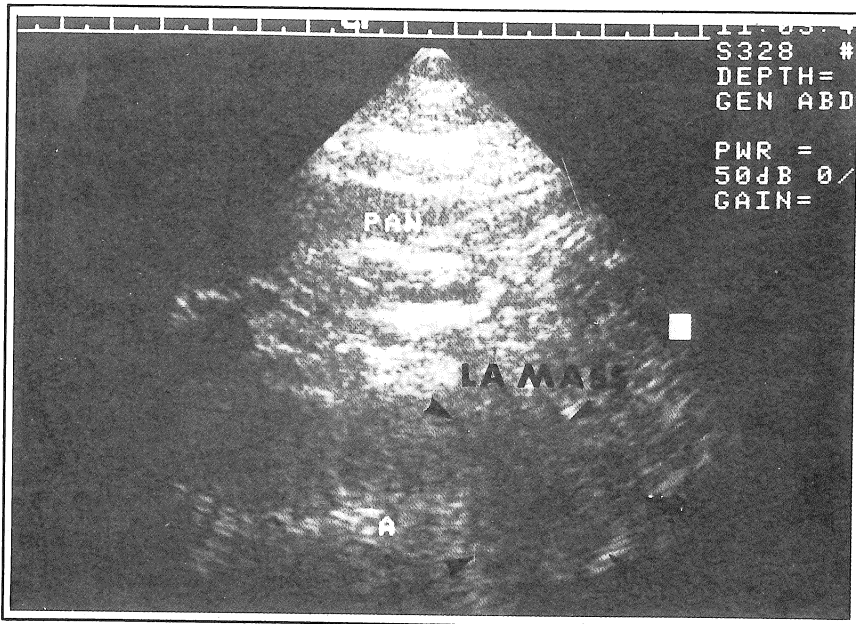


Fig. 1 (b)

Fig. 1 (a) Longitudinal and (b) transverse sonograms reveal a hypoechoic left adrenal mass.

in the T2-weighted images, consistent with a pheochromocytoma. The patient was commenced on alpha and beta blockers in preparation for surgery which was performed at approximately twelve weeks gestation.

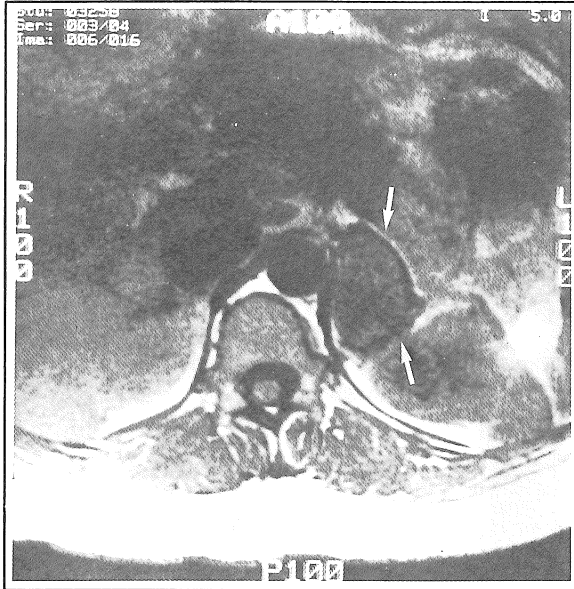


Fig. 2(a) :
 Axial T1-weighted MR image (800/20) shows a left adrenal mass (arrows) slightly hypointense compared with liver.

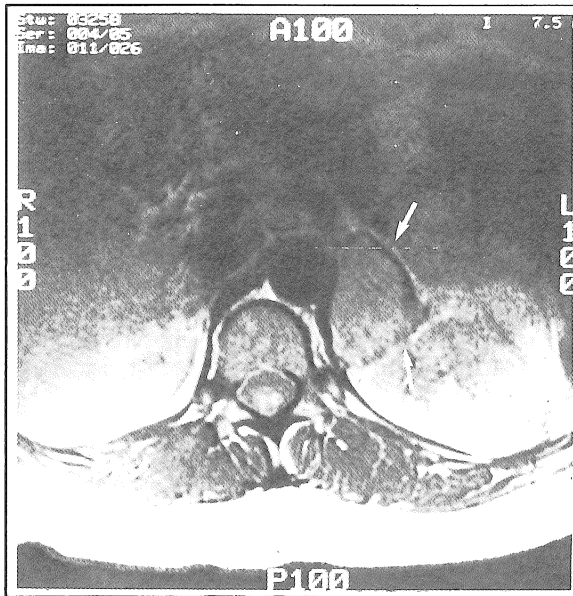


Fig. 2(b) :
 Axial T2-weighted MR image (2000/30) shows the left adrenal mass (arrows) is markedly hyperintense compared with the liver.

The tumour was removed under invasive monitoring and histopathology confirmed an adrenal phaeochromocytoma weighing 39 gm and measuring 4 x 3 x 0.5cm. The post operative course was complicated by a pulmonary embolus which was treated with heparin infusion. A follow-up fetal ultrasound revealed a normal fetus at 13 weeks gestation. A repeat urinary catecholamine was within normal limits two weeks post operatively. On discharge her pulse rate was 96/min and blood pressure 135/75 mm Hg.

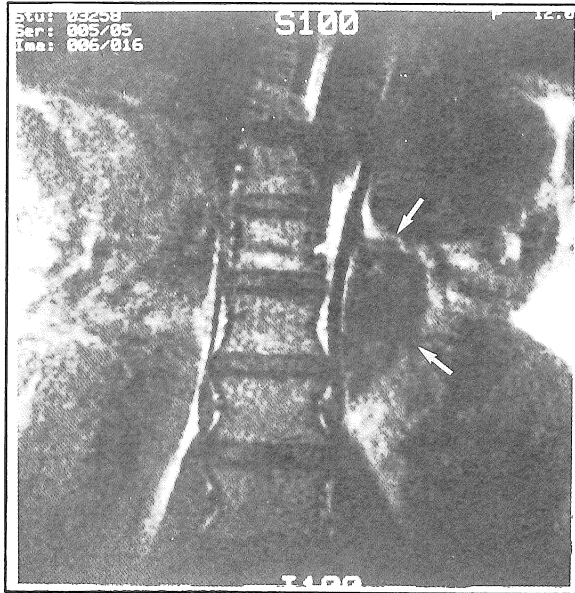


Fig. 2(c) :
Coronal T1-weighted MR image
(800/20) again demonstrates the
left adrenal phaeochromocytoma
(arrows)

Discussion

Phaeochromocytoma is a tumour which usually arises from the chromaffin cells in the adrenal medulla, but it may arise from other extra-adrenal chromaffin tissue. Phawchromocytoma is usually benign, single and adrenal in origin. They may be bilateral, multiple and extra adrenal in 10% of cases.

Schutz et al⁴, in a review of the medical literature, found a male to female ration of 4:3. The patient age at the time of diagnosis varied from 11 years^{1,2} to 78 years of age⁵, the peak age lying between the second and third decade. Total of 47 cases reported in the medical literature from 1980 – 1989³. Majority of patients present with flushing, palpitations, headache, sweating, marked anxiety, tachycardia and paroxymal or sustained hypertension. In pregnancy it is often misdiagnosed as pre eclampsia or uterine rupture when patient goes into shock during or after labour⁶. Phaeochromocytoma is associated with high maternal mortality (48%) and fetal (54%) mortality rate if diagnosis is not made during pregnancy. However, when diagnosed before term, maternal mortality has been considerably decreased² and fetal mortality less significantly.

When diagnosis is suspected clinically, the diagnosis is confirmed and after tumour localisation it should be removed immediately before 20 – 24 weeks gestation. After 24 weeks however the pregnancy should be allowed to continue under adequate adrenergic blockade until fetal maturity and delivery by caesarian section. Tumour resection can then be done either at the time of caesarian section or later³.

Diagnosis is best established by measurement of serum or urinary catecholamine or their metabolites. Preparative localisation has been done previously by intravenous pyelography, presacral gas insuffulation, arteriography, vena-cavagraphy and sampling of blood for catecholamine at multiple sites.

Scintigraphic localisation with ¹³¹I-mIBG has been utilised^{7,8}. During pregnancy tumour localisations are restricted. Computed tomography has been used in pregnancy⁹. Recently MRI has been utilised for tumour localisation during pregnancy^{3,10,11}.

We localised the tumour by ultrasound and MRI. Both are non-invasive, non-ionising radiation and provide images in multiple planes. Abdominal ultrasound although safe may be inadequate in diagnosis of multiple lesions and extra-adrenal sites. Computed tomography (CT) was not performed

in our patient to avoid radiation exposure to the fetus. Although CT has been used in pregnancy³ and no adverse side effects have been reported so far. Sonography confirmed the gestational age and demonstrated a 4cm solid tumour arising from the left adrenal gland, the right adrenal gland appeared normal. Ultrasound and MRI are not diagnostic. The only indicator of phaeochromocytoma is the presence of elevated urinary catecholamine levels.

MRI is useful in evaluating patients with phaeochromocytoma by virtue of the prolongation of the tumour T2 and T1, relative to adjacent tissue and the low signal emanating from patent vasculature. Adrenal phaeochromocytoma are frequently large at the time of diagnosis that T2-weighted SE images may be sufficient. However a T1-weighted sequence may be required for a complete assessment and for detection of small or ectopic tumours in which visualisation of tumour – fat interface is crucial.

Coronal and sagittal scans are useful in evaluating the cephalic and caudal extent of the tumours. Coronal scanning, demonstrates the sympathetic chain, a region which is at risk for ectopic phaeochromocytoma¹¹. In the absence of necrosis or haemorrhage within the tumour, phaeochromocytomas appear as homogenous low intensity on T1-weighted images and high signal intensity on T2-weighted images. Large necrotic tumours are heterogenous in appearance Phaeochromocytoma during Pregnancy : Ultrasound & MRI appearances especially in heavily T2-weighted images. Cystic changes within the tumour display very high signal intensity on T2-weighted images^{12,13,14}. In the absence of metastatic deposit it is not possible to differentiate benign and malignant tumours on the basis of MRI appearance. MRI, however is useful in follow-up of patients with previous tumour resection, previous surgery with surgical clips, or disruption of the retroperitoneal fat planes since surgical clips do not affect MR images, whereas they degrade CT images. There are no known deleterious effects from static magnetic field exposure^{15,16}. Fourteen patients with tumour recurrences or metastases in association with phaeochromocytoma in pregnancy have been reported². Post-operative follow-up with urinary catecholamine measurement is essential because of tumour recurrence or metastases may develop many years later².

References

1. Schenker J.C., Chowers I. Phaeochromocytoma and pregnancy. Review of 89 cases. *Obstet Gynaecol Surv* 1971; 26 : 736 – 47.
2. Schenker J.C., Chowers I. Phaeochromocytoma and pregnancy – an update appraisal. *Aust NZJ Obstet Gynaecol* 1982; 22 : 1 – 10.
3. Harper M.A., Murmaghan G.A., Kennedy L., et al. Phaeochromocytoma in pregnancy. Five cases and a review of the literature. *Brit J Obstet and Gynaecol* 1989; 96 : 594 – 606.
4. Schutz W., Vogel EL: Phaeochromocytoma of the urinary bladder : A case report and review of the literature. *Urol Int* 1984 ; 39 : 250 – 55.
5. Leetsma J.E., Price C.B. Jr. Paraganglioma of the urinary bladder. *Cancer* 1971; 28 : 1063 – 73.
6. Chatterjee T.K., Parek U. Phaeochromocytoma in pregnancy. *Aust NZJ Obstet Gynaecol* 1985; 25 : 290 – 1.
7. Sisson J.C., Frager M.S., Valk T.W. Scintigraphic localisation of phaeochromocytoma. *N Engl J Med* 1981 ; 305 : 12 – 7.
8. McEwan A.J., Davis C.L., Ackery D.M. Bladder phaeochromocytoma diagnosed by radio-ionated meta-iodobenziguamide imaging. *Br J Urol* 1987; 59(6) : 594 – 5.
9. Ober K.P. Hyperthyroidism or phaeochromocytoma? *N.C. Med J.* 1983; 44: 285 – 6.
10. Greenburg M., Mowad A.H., Wieties B.M., et al. Extra adrenal phaeochromocytoma: Detection during pregnancy using MR imaging. *Radiology* 1986: 161 : 475 – 6:
11. Combs C.A., Easterling T.R., Schmucker B.C. et al. Haemodynamic observations during paroxymal hypertension in a pregnancy with phaeochromocytoma. *Obstet Gynaecol* 1989; 74(3): 439 – 41.
12. Glazer G.M., Woolsley E.J., Borrello J. et al. Adrenal tissue characterization using MR imaging. *Radiology* 1986; 158 : 73 – 9.
13. Reinig J.W., Doppman J.L., Dwyer A.J. et al. Adrenal masses differentiated by MR. *Radiology* 1986; 158o: 81 – 4.
14. Fink I.J., Reinig J.W., Dwyer A.J. et al. MR imaging of phaeochromocytomas. *J Comput Assit Tomogr.* 1985; 9: 454 – 8.
15. McCarthy S.M., Stark D., Filly R.A. Obstetrical Magnetic Resonance Imaging: Maternal Anatomy. *Radiology* 1985; 154: 421 – 5.
16. Weinreb J.C., Lowe T.W., Santos-Ramos R. et al. Magnetic resonance imaging in obstetric diagnosis. *Radiology* 1985; 154: 157 – 61.