CASE REPORTS:

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Phaecochromocytoma in Pregnancy: Report on two cases and a Review of the literature.

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SUMMARY

Two cases of phaecochromocytoma in pregnancy are described. One case was diagnosed at autopsy and in the other case the tumour was successfully removed. The world and particularly African literature on the subject is briefly reviewed. The clinical feature, diagnosis and management are discussed in the context of the reported cases.

INTRODUCTION

The phaeochromocytoma is an uncommon tumour of chromaffin tissue which discharges epinephrine and norepinephrine into the circulation, causes hypertension and only surgical removal of the tumour can cure the patient. The coincidence of phaeochromocytoma and pregnancy is a rare event and more sinister because of high maternal (48%) and foetal (54.4%) mortality (Schenker and Chowers, 1971). The presence of phaeochromocytoma in pregnancy has been reported ninety four times in the literature (Fox et al, 1969; Chukwuemeka et al, 1974; Templeton 1967; Sommers et al, 1967). Only 24 out of these 94 cases were diagnosed during pregnancy. Because of paucity of reports in the literature, it was thought to be a rare phenomenon for an African to have phaeochromocytoma and only four cases of phaeochromocytoma in pregnancy have been reported from Africa (Sommers et al, 1967; Templeton 1967; Chukwuemeka et al, 1974). Out of these four cases, only one patient was diagnosed correctly during pregnancy and the successful surgical removal of the tumour cured the patient (Chukwuemeka et al, 1974). All the other three patients died and were diagnosed on postmortem examination. Two cases of phaeochromocytoma in pregnancy are reported here, one was diagnosed at autopsy and the other one was successfully managed.

Case No. 1.

Mrs. C.M. Age 20, Para 1+1, Gravida 3, an African woman was referred from a peripheral clinic on 27.11.73 to the University Teaching Hospital, Lusaka because of hypertension (BP 160/95m.m. Hg) and palpitation at 28 weeks of gestation. The obstetric history revealed that the patient's first pregnancy ended in abortion at 4 months in 1969 and she had a

normal vaginal delivery in 1971 without any complications. On clinical examination, the size of the uterus was of 28 weeks gestation, the blood pressure was 120/70m.m. Hg and there was mild albuminuria. The patient was advised to attend the antenatal clinic four weeks later. The patient was admitted to the hospital after one month with the complaint of having palpitation and abdominal pain. On clinical examination, she was found to be anaemic, hypertensive (BP 150/90m.m. Hg) and she had tachycardia (pulse rate 100/min.). She was also thought to have hydramnios. After admission to hospital, for the next five days the blood pressure ranged between 120/70 and 168/100 m.m. H.g. Other investigations revelaed that she was suffering from hypochromic anaemic (HB. 6.9 gm%) and it was decided to transfuse her one pint of packed cell. On the day of transfusion, immediately after abdominal examination of the patient in the morning, the blood pressure was recorded to be 220/120 m.m. Hg. and she was heavily sedated with the intramuscular injection of 15mg of morphine and 25mg. of phenergan. A Multiple pregnancy was suspected and it was decided to take X-ray of the abdomen. The patient had one pint of packed cell transfusion in the evening without any complication. At about 1 a.m., the patient suddenly became dyspnoeic, started vomiting and died soon afterwards. The postmortem examination revealed mild myocarditis, congestion of lungs and a small retroperitoneal tumour in front of the second lumber vertebra which showed classical features of a phaeochromocytoma. There was also uniovular twin pregnancy in the uterus.

Case No. 2

Mrs. A.P., Age 32 years, para 2+1, gravida 4, an African woman was referred on 14th August, 1974, to the University Teaching Hospital, Lusaka, from a peripheral clinic for hypertension. On admission, the patient complained of headahce and abdominal pain for the last four days. On examination, the blood pressure was 160/110 m.m. Hg. and there was no oedema or albuminuria. The patient was sedated with sodium amytal 100mg. T.D.S. and the blood pressure remained stable between 120/80 m.m. Hg. and 100/60 m.m. Hg. On the fourth day of admission there was sudden rise of blood pressure to 210/120 m.m. Hg. and the patient was heavily sedated with intravenous injection of 20mg. valium and 80mg. valium was administered in 5% dextrose, as intravenous drip in 24 hours. On the following morning, the patient suddenly collapsed and she was diagnosed to have phaeochromocytoma.

A normal saline drip was put up and for controlling blood pressure, phenoxybenzamine (Dibenyline) and propranolol (Inderal) was used. Initially for two days the blood pressure settled but subsequently she started having episodic hyper and hypotension. The rogitine (phentolamine) test was positive but the the vaninyl-mandelic acid (V.M.A.) measured on one occasion was within normal range (5mg/24hrs.).

It was decided to terminate the pregnancy by caesarean section and to remove the tumour at the same time. On the eighth day of admission, the patient refused to undergo operation and took her own discharge from the hospital. Two months later at 36 weeks of gestation the patient was admitted in labour. On examination, the blood pressure was 180/120m.m. Hg., protinuria ++ and there was no oedema. The obstetric examination revealed the baby to be lying transversely with the elbow presenting, the cervix 3 c.m. dilated and the membrane already ruptured. A lower segment caesarean section was done under general anaesthesia and a healthy baby weighing 7lb 9oz with an apgar score of 7/10 was delivered. On exploring the upper abdomen a firm tumour of 3" diameter was detected on the upper pole of left kidney.

During the postoperative period after caesarean section, the patient was put on Phenoxybenzamine (Dibenyline) 20mg. 6 hourly and propranolol (Inderal) 20mg. 4 times daily. Two days later, as it was becoming difficult to control the blood pressure it was decided to remove the tumour. The abdomen was opened by transverse upper abdominal incision and the veins draining from the left suprarenal gland was ligated first before the tumour was dissected out. The postoperative period was uneventful and her blood pressure remained normal. She was discharged from the hospital 3 weeks later. The histology of the tumour showed the features of typical phaeochromocytoma.

DISCUSSION

The outstanding clinical picture in phaeochromocytoma is the episodes of paroxysmal hypertension occuring spontaneously or initiated by mechanical stimulation of the tumour. The first case that is reported here, although had episodic hypertension on palpation of the abdomen but unfortunately the phaeochromocytoma was not suspected at that time. Another peculiarity of the first case was that, although the patient was referred from the peripheral clinic for hypertension, at our antenatal clinic the blood pressure was found to be within normal range (120/70 m.m. Hg.). This was due to the fact that phaeochromocytoma does not always discharge catecholamines into the circulation and sometimes it can even cause sustained rise of blood pressure or simulate pre-eclamptic toxaemia.

Other associated clinical features e.g. palpitation and tachycardia were also missed in the first patient. The phaeochromocytoma is sometimes associated with headache, vomiting, pallor of extremites, sweating and glycosurea. Because of these bizarre clinical features often phaeochromocytoma has been mis-

diagnosed in obstetric practice as pre-eclampic toxaemia and eclampsia.

The second case had typical features of episodic hypertension followed by shock like state and it was correctly diagnosed. It was a miracle that she was alive for two months, between the time that she went home against medical advice and when she came back in labour.

The pharmacological tests of using histamine vasopressor or rogitine (phentolamine) to diagnose phaeochromocytoma is not suitable during pregnancy because of serious side effects. The rogitine test was positive in the second patient. The estimation of excess urinary catecholamines or their metabolite (V.M.A.) or the blood catecholamine level should be confirmatory provided the tumour was secreting at the time the specimen is collected. In the second patient, the urinary vaninyl-mandelic acid (V.M.A.) level was within normal limit (5mg/24 hrs.), although it was clinically suggestive of phaeochromocytoma.

Once the clinical and chemical tests suggest phaeochromocytoma, it is necessary to localize the tumour. The intravenous pyelogram, presacral insufflation of Carbon Dioxide, aortography and venocavography have all been utilized, but they all have their limitations and danger. The tumour was localized during pregnancy by laparotomy in 23 out of 24 causes of phaeochromocytoma that are reported in the literature (Schenker and Chowers, 1971; Chukwuemeka et al, 1974). In the second patient, the opportunity of localizing the tumour was taken when the abdomen was opened for caesarean section.

Once a correct diagnosis of phaeochromocytoma is made, the tumour must be removed irrespective of the stage of gestation because of high maternal and foetal mortality.

As labour and vaginal delivery is associated with high maternal mortality (33%) so it is advisable to deliver the baby by caesarean section (Schenker and Chowers, 1971). If the operation is a planned procedure then it is better to remove the tumour at the same time. In the second patient, the tumour was not removed at the time of caesarean section as the operative mortality is reported to become double in unplanned surgery (Rideli et al, 1963).

A special care was also taken in inducing anaesthesia and in avoiding cyclopropane or halothane gas because they produce cardiac arrhythimia in the presence of excess catecholamine.

In the past twenty years, the use of alpha and beta-receptors blocking drugs have reduced the hazards of operation and are beneficial in treating phaechromocytoma while the patient is prepared for the surgery. In the second case, although the alpha and beta-receptors blocking drugs helped to control the blood pressure for a short time yet it became exceedingly difficult to maintain a sustained level until the tumour was removed.

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REFERENCES

1. *Chukwuemeka A.C., Paton A.C., Gebbie A.M., Ayim E.N. and Dhall D.P. (1974): East African Medical Journal. Vo. 51:6:496—499.*

2. *Fox L.P., Grandi J. and Johnson A.H. (1969): American Journal of Obstetrics and Gynaecology. Vol. 104: 289.*

3. *Ridell D.H., Schull I.G., Frist T.F. and Baker T.D. (1963): Annals of Surgery. 157:980.*

4. *Schenker J.G. and Chowers I. (1971): Obstetrical and Gynaecological Survey. Vol. 26:11:739—747.*

5. *Sommers K., Shepherd J.J. and Rwabugabya H.G.K (1967): Tropical and Geographical medicine. 19:294—299.*

6. *Templeton A.C. (1967): East African Medical Journal. Vol. 44:7:271—277.*