Phaeochromocytoma in pregnancy

A report of 3 cases

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Summary

The coincidence of phaeochromocytoma and pregnancy is rare and potentially lethal. Three cases are reported; in 2 the fetus had died before the patient presented. With early diagnosis and appropriate treatment there should be no maternal mortality, and the fetal mortality rate should be reduced to less than 20%.

The incidence of phaeochromocytoma among hypertensive patients is 0.2 - 0.7% and if untreated and undiagnosed it is invariably fatal. In pregnancy there is a real danger to the mother and unborn child, the reported incidence of maternal and fetal death being as high as 50%. With its low incidence in the adult hypertensive population, phaeochromocytoma is a rare tumour, doubly so in pregnancy — in the 1970s only 128 cases were reported, with only 42 cases being diagnosed before delivery.

The diagnosis should be suspected in any pregnant patient who develops severe symptomatic hypertension and arrhythmia. Patients with phaeochromocytoma may have remittent, intermittent or sustained hypertension. Additional symptoms, such as palpitations, headache, sweating, pallor and anxiety, can occur spasmodically and should be carefully sought for in the pregnant patient with hypertension.

Case reports

Case 1

A 30-year-old woman presented to hospital in the 18th week of her first pregnancy with periodic attacks of anxiety, sweating and palpitations. The blood pressure was 220/130 mmHg and a mass was felt in the left hypochondrium — in 1970s only 128 cases were reported, with only 42 cases being diagnosed before delivery.

The diagnosis should be suspected in any pregnant patient who develops severe symptomatic hypertension and arrhythmia. Patients with phaeochromocytoma may have remittent, intermittent or sustained hypertension. Additional symptoms, such as palpitations, headache, sweating, pallor and anxiety, can occur spasmodically and should be carefully sought for in the pregnant patient with hypertension.

REFERENCES

Histological examination showed a typical benign phaeochromocytoma with a few mitotic figures.

The patient made an uneventful recovery. She left hospital on the 14th postoperative day by which time she was receiving no drugs. The blood pressure was 120/80 mmHg and the concentration of VMA in the urine was 23.6 \( \mu \text{mol}/24 \text{ h} \).

Case 2

A 23-year-old woman who had had 1 healthy child presented to hospital at 33 weeks' gestation complaining of headaches, anxiety, and sweating. She had a history of hypertension, which was controlled medically; her mother was also hypertensive. The concentration of VMA in the urine was 54 \( \mu \text{mol}/24 \text{ h} \) and the total metanephrines and normetanephrines 19.6 \( \mu \text{mol}/24 \text{ h} \) (normal < 5.5 \( \mu \text{mol}/24 \text{ h} \)). Fetal heart sounds were normal and ultrasonography showed a normal fetus corresponding to 33 weeks' gestation; it also showed a possible tumour in the left adrenal gland. Magnetic resonance imaging (MRI) restricted to the adrenal areas failed to show any abnormality.

The blood pressure was stabilised at 120/80 mmHg after 10 days' treatment with phenoxybenzamine 40 mg orally twice daily. Caesarean section was then performed through a long midline incision and a live infant weighing 3200 g delivered. During surgery, the patient's blood pressure was extremely labile and hypertensive episodes were controlled with nitroprusside. A small nodule was felt in the left adrenal gland, which was removed. The gland appeared to be normal. However, hypertension persisted after its removal so further exploration of the abdomen was undertaken. This led to the discovery and removal of a small tumour behind and to the left of the abdominal aorta above its bifurcation. Thereafter the blood pressure stabilised at 120/90 mmHg. The tumour weighed 8 g and histological examination showed it to be a benign phaeochromocytoma.

Postoperative recovery was uneventful and 6 weeks later the blood pressure was 120/70 mmHg. When last seen at the age of 14 months the child seemed normal.

Case 3

A 25-year-old woman presented to hospital in the 24th week of her 1st pregnancy with headaches and palpitations. She had a history of hypertension but the blood pressure was now 260/140 mmHg and the fetus was dead. The concentration of VMA in the urine was 93 \( \mu \text{mol}/24 \text{ h} \). Computed tomography (CT) showed a tumour in the right adrenal gland and gamma scanning after injection of metaiodobenzylguanidine labelled with iodine-131 showed this mass and also a second small mass medial and below the first one seen. This raised the possibility of multiple endocrine neoplasia but the serum calcium and calcitonin values were normal.

The blood pressure was stabilised at 130/90 mmHg after treatment with phenoxybenzamine 40 mg 3 times a day for 7 days. Before laparotomy (through an upper abdominal midline incision) the uterus was evacuated, and then the right adrenal gland was removed. Further exploration showed a second tumour behind the second part of the duodenum and this also was removed. Both tumours had the histological appearance of benign phaeochromocytoma.

The patient made a good recovery and 6 weeks later the blood pressure was 130/85 mmHg and the concentrations of VMA and metanephrines in the urine were normal. CT and isotope scans were also normal.

Discussion

Phaeochromocytoma is a rare but serious complication of pregnancy; the mortality is reported to be as high as 58% for the mother and 54% for the fetus. The diagnosis should be considered in any patient who presents with hypertension associated with palpitations, anxiety, headache, and sweating. Radiography is contraindicated during pregnancy, but raised concentrations of VMA and metanephrines in the urine, together with abnormalities on ultrasonography and where available — MRI should confirm the diagnosis. If the fetus is already dead, CT and isotope scanning are indicated.

Medical treatment should be started immediately to control the blood pressure and, if the pregnancy is in the 1st or 2nd trimester, the tumour should be removed and the pregnancy allowed to go to term. If the diagnosis is made during the 3rd trimester, the blood pressure should be controlled by drugs until the fetus is viable and then delivery should be by caesarean section, the tumour being removed at the same operation. The blood pressure should stabilise as soon as the tumour is removed; if it does not the presence of a second tumour should be suspected. Caesarean section is preferable to vaginal delivery because labour can cause a great increase in the concentration of catecholamines in the mother’s blood, prematurity of the placenta, and death of the fetus.

Swift diagnosis and appropriate treatment are essential if maternal and fetal mortality is to be kept to a minimum.

REFERENCES