Pheochromocytoma in pregnancy

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Key words: pheochromocytoma, pregnancy

Introduction

Pheochromocytoma is not common. It is rare still during pregnancy.

Case report

A 22 years G2 P1 A0 L0 from rural background was referred on 4th April, 2004 with severe hypertension at 7 1/2 months of pregnancy (LMP-28th September, 2003). She had intrauterine fetal death (IUFD) at full term in her first pregnancy. Cause of IUFD was not known. Her blood pressure was 180/134 mmHg. There was mild pedal edema. She complained of mild headache and occasional restlessness. Urine showed +++ albumin. Hb-9 g/dL. White cell count, bleeding time, clotting time, prothrombin time, blood sugar, liver function tests, blood urea, serum creatinine, and platelet count were within normal limits. Blood uric acid was slightly raised to 5 g/dL. VDRL, HbsAg and HIV were negative. Blood group was B +ve. Fundoscopy showed bilateral disc edema of about 2 DP, scanty hemorrhage and soft exudates. ECG showed left axis deviation and sinus tachycardia. Sonography showed single fetus of 31 weeks gestation with corresponding placental maturity and Manning’s score. Diagnosis of severe preeclampsia was made. She was given hematinics, antihypertensives, and phenobarbitone orally besides other routine measures appropriate for the condition. Despite high doses of antihypertensives viz., aldomet 500 mg four times a day, nifedipine 20 mg four times a day, and labetalol 100 mg twice a day. Her blood pressure remained high varying from 150/100 to 190/140 mmHg. Blood pressure was measured every 4 hours. Although albuminuria and edema disappeared her fundoscopy findings showed deterioration. Unrelenting hypertension for more than 7 days made us review the case. Meanwhile she developed occasional headache and palpitation. Two hourly blood pressure recording showed wide variations and paroxysmal rise up to 250/150 mmHg. Cold extremities and excessive sweating made us suspect pheochromocytoma. At times systolic blood pressure dropped down to 80 mmHg especially on prolonged sitting / standing. Detailed sonography of the abdomen on 2nd April, 2004 showed adrenal tumor. Vanillyl mandelic acid (VMA) test done on 13th April, 2004 showed normal values. MRI on 14th April, 2004 confirmed presence of well-defined 39x31 mm left adrenal tumor in superior anterior location of kidney. She was put on prazosin 5 mg three times a day and nifedipine 10 mg four times a day. Phenoxymethazine was added on 20th April, 2004 with starting dose of 10 mg twice a day. It had to be increased up to 100mg/day. Aldomet was omitted. On addition of phenoxymethazine there was definite subjective improvement in the form of decreased sweating, normal skin temperature and decreased palpitation. Blood pressure was stabilized with its range varying between 130/90 to 250/150 mmHg. Cold extremities and excessive sweating made us suspect pheochromocytoma. At times systolic blood pressure dropped down to 80 mmHg especially on prolonged sitting / standing. Detailed sonography of the abdomen on 12th April, 2004 showed adrenal tumor. Vanillyl mandelic acid (VMA) test done on 13th April, 2004 showed normal values. MRI on 14th April, 2004 confirmed presence of well-defined 39x31 mm left adrenal tumor in superior anterior location of kidney. She was put on prazosin 5 mg three times a day and nifedipine 10 mg four times a day. Phenoxymethazine was added on 20th April, 2004 with starting dose of 10 mg twice a day. It had to be increased up to 100mg/day. Aldomet was omitted. On addition of phenoxymethazine there was definite subjective improvement in the form of decreased sweating, normal skin temperature and decreased palpitation. Blood pressure was stabilized with its range varying between 130/94 to 180/120 mmHg. She was placed on strict supervision with blood pressure, pulse and fetal heat sound recording every 4 hours. DFMC and nonstress test were done on alternate days. Liver and kidney function tests, platelet count, fundoscopy, ECG and sonography were done every week. Serial sonography showed progressive growth of the fetus. Color doppler studies done at 34 weeks of gestation showed normal placental and fetal blood flow studies. After few days of stable blood pressure it started fluctuating again with paroxysmal rise up to 240/140 mmHg. Fundoscopy on 17th May, 2004 showed bilateral papilledema along with increased hemorrhage and exudates. Cold sweats appeared again. Sonography on 19th May, 2004 showed fetal maturity of 35 weeks with definite presence of femur epiphysis. Anticipating need for preterm termination she was given weekly dexamethasone injections starting on 10th May, 2004. In view of her deteriorating condition it was decided on 20th May, 2004 to terminate her pregnancy by cesarean section. Anesthetist advised stoppage of phenoxymethazine 2 days prior to surgery to prevent post-adrenectomy hypotension. However she went into spontaneous labor in the morning of 21st May, 2004 before phenoxymethazine could be stopped. A live female baby weighing 2.75 kg with good apgar score was delivered by lower segment cesarean section. Surgery for pheochromocytoma was postponed because of inadequate preparation of the patient for surgery. Postoperative period was uneventful. Stitches were removed on 7th post-operative day. Antihypertensive drugs were continued. She was subsequently operated for removal of pheochromocytoma on 4th June, 2004 by anterior intraabdominal route. Histology of the tumor confirmed the diagnosis of pheochromocytoma. Her blood pressure levels came down to normal 3 days after the operation. She was discharged on 16th June, 2004. At the last followup on 21st October, 2004 she was asymptomatic with normal blood pressure and her baby was healthy and doing well.

Figure 1. Showing the tumor.

Figure 2. Ultrasonography.
Obstetric Case report

Discussion

Pheochromocytoma during pregnancy is rare, with an estimated prevalence of 1 in 50,000 to 54,000 in full term pregnancies. Before 1970 only 25% of the cases were diagnosed during pregnancy. Since 1990, 85% of the cases have been diagnosed antenatally suggesting an increased awareness of this rare but lethal condition. The maternal mortality rate is 2 to 4% if the tumor is diagnosed in the antenatal period, compared to 14 to 25% if it is diagnosed intrapartum or after delivery 1-3. Fatal hypertensive crisis can be precipitated by anesthesia, mechanical effects of the gravid uterus, vaginal delivery, hemorrhage into the tumor, and vigorous fetal movements. Fetal mortality is 11 to 15% when pheochromocytoma is diagnosed before delivery but approaches 55% if the diagnosis is delayed 4. The only way to diagnose a pheochromocytoma is first to think of it. The correct diagnosis was overlooked in our patient for 7 frightening days. The challenge is to differentiate preeclampsia from the hypertensive crisis of an unrecognized pheochromocytoma. Once the tumor is diagnosed, management with an appropriate α-adrenergic-receptor antagonist and delivery by cesarean section under the expert care of a perinatal team is ideal 4-6. Unique to this case was the use of MRI to diagnose pheochromocytoma and negative urinary VMA test 6. Had we awaited biochemical confirmation of the clinical suspicion the woman might have died. Multispeciality team work is a must for best maternal and fetal outcome of this potentially fatal condition 4-7. Surgery for pheochromocytoma can be undertaken early in pregnancy (before 24 weeks), during cesarean section, or after delivery at a later date depending on availability of facility and expertise 4,5,7.

References


Paper received on 24/04/2005 ; accepted on 14/11/2005

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