International Journal of Preventive Medicine

Case Report Open Access

Pheochromocytoma after Cesarean Section

Elham Naghshineh, Azar Danesh Shahraki, Somaye Sheikhalian, Leila Hashemi

Department of Obstetrics and Gynecology, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran

Correspondence to:
Dr. Azar Danesh Shahraki, Department of Obstetrics and Gynecology, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran.
E-mail: danesh@med.mui.ac.ir

How to cite this article: Naghshineh E, Shahraki AD, Sheikhalian S, Hashemi L. Pheochromocytoma after cesarean section. Int J Prev Med 2016;7:60.

ABSTRACT

Pheochromocytoma is a catecholamine-producing tumor. There are a very few reported cases of clinical pheochromocytoma. Here, we report a 27-year-old woman para 1 live 1 with chief complaint of headache, confusion, nausea, and vomiting 2 days after cesarean section. She was anxious and had palpitation. On physical examination, fever, tachycardia, tachypnea, high blood pressure, and right thyroid nodule were found. She was managed as pregnancy-induced hypertension at first. In laboratory data, epinephrine, norepinephrine, metanephrine, normetanephrine, and vanillylmandelic acid were increased in 24 h urine collection. An adrenal mass was detected in abdominal computed tomography. Regarding clinical and paraclinical findings, pheochromocytoma was diagnosed. The patient received medical treatment, but it was not effective; hence, she underwent adrenalectomy.

Keywords: Adrenal gland neoplasm, cesarean section, pheochromocytoma, pregnancy

INTRODUCTION

Pheochromocytoma is a catecholamine-producing tumor. Incidence of pheochromocytoma is <0.2/10,000 pregnancies.[1] The classical triads of pheochromocytoma are episodic headache, sweating, and tachycardia. Pheochromocytoma in pregnancy can induce by several mechanisms. One of the most important causes of hypertension in pregnancy is pheochromocytoma but its occurrence is rare. It has high morbidity and mortality for mother and fetus.

Due to the similarity to other forms of hypertension, diagnosis of pheochromocytoma often missed in pregnancy. Diagnosis of pheochromocytoma is based on catecholamine's concentration (epinephrine, norepinephrine, dopamine, and vanillylmandelic acid [VMA]) in 24 h urine collection. Sensitivity of the test would be increased if 24 h urine collection begins at onset of a paroxysm.

Sensitivity of abdominal computed tomography (CT) for diagnoses of adrenal pheochromocytoma is 93–100% and extra-adrenal pheochromocytoma is 90%. Magnetic resonance imaging sensitivity is more than CT for extra-adrenal pheochromocytoma. Other diagnostic techniques included metaiodobenzylguanidine imaging, positron emission tomography imaging, and somatostatin receptor scintigraphy.[2]

Management of pheochromocytoma includes medical therapy (phenoxybenzamine, propranolol) and surgery.[3]

CASE REPORT

A 27-year-old woman para 1 live 1 admitted with chief complaint of sudden onset of headache, confusion,

Access this article online

Quick Response Code:

Website: www.ijpvmjournal.net/www.ijpm.ir

DOI: 10.4103/2008-7802.178534

nausea and vomiting, shortness of breath, tachypnea, and hypertension 2 days after cesarean section. Her cesarean section was performed at term pregnancy due to fetal distress under spinal anesthesia. Her pulse rate was 125/min; systolic blood pressure was labile (70–230 mmHg) and failed to respond to conventional treatment. She had tachypnea and was febrile (38°C). In laboratory data, antinuclear A antibodies and antineutrophil cytoplasmic antibody were normal (NL), but epinephrine = 65.2 (NL: 1.7–22.4 μg/24 h), norepinephrine = 364 (NL: 12.5–85.5 μg/24 h), metanephrine = 2071 (NL: 30–180 mcg/24 h), normetanephrine = 1337 (NL: 103–390 mcg/24 h), and VMA = 17.18 (NL: 1.4–6.5 mg/24 h) were increased in 24 h urine collection. Abdominal CT revealed left adrenal mass 58 mm × 50 mm × 30 mm.

Regarding clinical and paraclinical findings, pheochromocytoma was diagnosed. The patient was given medical treatment with phenoxybenzamine and propranolol, but treatment was not effective; hence, she underwent adrenalectomy.

DISCUSSION

Pheochromocytoma is a neuroendocrine tumor derived from adrenal chromaffin cells, and extra-adrenal paraganglioma could cause secondary hypertension.[1] High amount of catecholamine's release (norepinephrine, epinephrine, and dopamine) causes the typical manifestations of pheochromocytoma. Vasoconstriction is one of the α-receptors' stimulation symptoms, whereas vasodilation is due to β2-receptors' stimulation. Stimulations of β1-receptors are increased heart rate and contractile force of myocardium.[4] Neuronal norepinephrine is inhibited by presynaptic α2-receptors.[3] Pheochromocytoma is very rare but it occurs in every age. A few cases of pheochromocytoma have been diagnosed each year.[5-8] Increases in intra-abdominal pressure, fetal movement, uterine contraction, delivery process, abdominal surgical intervention, and even general anesthesia can make pheochromocytoma clinically overt in pregnancy.

Signs and symptoms in pheochromocytoma are similar to other forms of hypertension, including the new-onset hypertensive syndromes in pregnancy, gestational hypertension, and preeclampsia. Hypertension can be insidious, and the pregnant women may even symptomatic till delivery.[1]

Signs and symptoms of pheochromocytoma are nausea and vomiting, abdominal pain, severe constipation (megacolon), chest pain, congestive heart failure, cardiac dysrhythmia, and conduction defects.

Preeclampsia and pheochromocytoma have similar manifestations. Both are characterized by hypertension. Initially, many of pheochromocytoma patients managed as preeclampsia. Preeclampsia is usually manifested by proteinuria, sudden weight gain, edema, rise in liver enzymes, and coagulation disorders.[4]

Pregnant woman with pheochromocytoma rarely has proteinuria, liver or coagulation abnormalities, sudden weight gain, and edema.

Initial management of pheochromocytoma is medical management with α-blockers. It should be started as soon as the α-methyl-parathyrosine (a tyrosine hydroxylase inhibitor) diagnosis is established and should be given for ≥10–14 days.[9] The best treatment in pregnancy is phenoxybenzamine (pregnancy Class C).[10] Other selective α1-blocker drugs, such as doxazosin, also can be used.[11] The β-blockers should not prescribe before α-blocker drugs because β-blockers alone can rise blood pressure dramatically due to unopposed α-adrenergic effects in patients with epinephrine-secreting tumors especially.[12,13] In patients with intolerance to combine α- and β-blockers, we can use α-methyl-parathyrosine but its safety during pregnancy is questionable. Methyldopa may worsen the symptoms of pheochromocytoma and not recommended.[11]

Treatment of choice for pheochromocytoma is surgery. The surgery time is controversial depending on factors such as gestational age of pregnancy, clinical response to treatment, the accessibility of the tumor for surgery, and fetal condition. Some physicians recommended surgical tumor removal after adequate medical treatment in early gestational age by laparoscopic adrenalectomy if tumor size is <7 cm. After 24 weeks of gestation, an elective cesarean section done then surgical procedure is recommended.[14] Cesarean section is the preferred mode of delivery.[7] Vaginal delivery is accompanied with higher mortality rate (31%) as compared with cesarean section (19%).[15,16]

In pregnant women, early diagnosis and treatment of pheochromocytoma are accompanied with better fetal outcomes. If appropriate management was done for pheochromocytoma, pregnancy loss is seen only in 11% of patients.[17] Catecholamines do not cross the placenta, but placental abruption due to paroxysmal hypertension and then rebound hypotension can cause severe hypoxia and fetal wastage. Maintain of adequate uterine perfusion is necessary during hypertension treatment. A adrenergic stimulation effects uteroplacental circulation and elevated catecholamine levels result in vasoconstriction.[17,18]

Early diagnose and management has been attributed to improvement in maternal outcomes. With earlier diagnosis and presurgical preparation, maternal mortality has decreased to 2% from 48% in older literature.[17]

In patients with a suspicious family history of pheochromocytoma, paraganglioma, or evidence of genetic cause, genetic testing should be considered.[17,19]

CONCLUSIONS

One of the rare but important causes of hypertension in pregnancy is pheochromocytoma with high morbidity and mortality of mother and fetus. Phenoxybenzamine is safe in pregnancy, but β-adrenergic blockers must be initiated only if necessary because their association with intrauterine growth retardation.[20,21] During the second trimester, resection of tumor can often be safe or tumor resection simultaneous with cesarean section in mature fetus.[22] Individualized approach for each patient is recommended. Pheochromocytoma presentations are different in pregnancy, but in the second half of pregnancy, new-onset or superimposed preeclampsia may be mistaken with it.[5] High clinical suspicion, early diagnosis and treatment, and collaboration among specialists are required for better management of patients and improved outcome.

Acknowledgements
We want to thank from Alzahra Hospital ICU nurses for their cooperativeness.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

Received: 06 Aug 13 **Accepted:** 16 Jan 16
Published: 10 Mar 16

REFERENCES

1. Kondziella D, Lycke J, Szentgyörgyi E. A diagnosis not to miss: Pheochromocytoma during pregnancy. J Neurol 2007;254:1612-3.
2. Ilias I, Pacak K. Current approaches and recommended algorithm for the diagnostic localization of pheochromocytoma. J Clin Endocrinol Metab 2004;89:479-91.
3. Eisenhofer G, Siegert G, Kotzerke J, Bornstein SR, Pacak K. Current progress and future challenges in the biochemical diagnosis and treatment of pheochromocytomas and paragangliomas. Horm Metab Res 2008;40:329-37.
4. Manger WM. An overview of pheochromocytoma: History, current concepts, vagaries, and diagnostic challenges. Ann N Y Acad Sci 2006;1073:1-20.
5. Oliva R, Angelos P, Kaplan E, Bakris G. Pheochromocytoma in pregnancy: A case series and review. Hypertension 2010;55:600-6.
6. Oger P, Raiffort C, Plouin PF, Mandelbrot L. Pheochromocytoma and pregnancy. Case report. Gynecol Obstet Fertil 2006;34:323-5.
7. Dugas G, Fuller J, Singh S, Watson J. Pheochromocytoma and pregnancy: A case report and review of anesthetic management. Can J Anaesth 2004;51:134-8.
8. Harper MA, Murnaghan GA, Kennedy L, Hadden DR, Atkinson AB. Phaeochromocytoma in pregnancy. Five cases and a review of the literature. Br J Obstet Gynaecol 1989;96:594-606.
9. Witteles RM, Kaplan EL, Roizen MF. Safe and cost-effective preoperative preparation of patients with pheochromocytoma. Anesth Analg 2000;91:302-4.
10. Kinney MA, Narr BJ, Warner MA. Perioperative management of pheochromocytoma. J Cardiothorac Vasc Anesth 2002;16:359-69.
11. Kalra JK, Jain V, Bagga R, Gopalan S, Bhansali AK, Behera A, *et al.* Pheochromocytoma associated with pregnancy. J Obstet Gynaecol Res 2003;29:305-8.
12. Bravo EL, Tagle R. Pheochromocytoma: State-of-the-art and future prospects. Endocr Rev 2003;24:539-53.
13. Asensio Martín MJ, Pavón Benito A, Barrena Sotes J, Zabaleta Zúñiga A, Salvador Bravo M. Anesthesia for surgical removal of a pheochromocytoma during the first trimester of pregnancy. Rev Esp Anestesiol Reanim 2009;56:129-31.
14. Reisch N, Peczkowska M, Januszewicz A, Neumann HP. Pheochromocytoma: Presentation, diagnosis and treatment. J Hypertens 2006;24:2331-9.
15. Junglee N, Harries SE, Davies N, Scott-Coombes D, Scanlon MF, Rees DA. Pheochromocytoma in pregnancy: When is operative intervention indicated? J Womens Health (Larchmt) 2007;16:1362-5.
16. Kariya N, Nishi S, Hosono Y, Hamaoka N, Nishikawa K, Asada A. Cesarean section at 28 weeks' gestation with resection of pheochromocytoma: Perioperative antihypertensive management. J Clin Anesth 2005;17:296-9.
17. Ahlawat SK, Jain S, Kumari S, Varma S, Sharma BK. Pheochromocytoma associated with pregnancy: Case report and review of the literature. Obstet Gynecol Surv 1999;54:728-37.
18. Dahia PL, Hayashida CY, Strunz C, Abelin N, Toledo SP. Low cord blood levels of catecholamine from a newborn of a pheochromocytoma patient. Eur J Endocrinol 1994;130:217-9.
19. Young WF Jr. Adrenal causes of hypertension: Pheochromocytoma and primary aldosteronism. Rev Endocr Metab Disord 2007;8:309-20.
20. Butters L, Kennedy S, Rubin PC. Atenolol in essential hypertension during pregnancy. BMJ 1990;301:587-9.
21. Montan S, Ingemarsson I, Marsál K, Sjöberg NO. Randomised controlled trial of atenolol and pindolol in human pregnancy: Effects on fetal haemodynamics. BMJ 1992;304:946-9.
22. Pawlu C, Bausch B, Reisch N, Neumann HP. Genetic testing for pheochromocytoma-associated syndromes. Ann Endocrinol (Paris) 2005;66:178-85.