Concurrent Parathyroidectomy and Caesarean Section in the Third Trimester

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Abstract

Background: Primary hyperparathyroidism is the most common cause of hypercalcemia in the general population. It is a rare complication of pregnancy that is difficult for clinicians to recognize, yet it can have important and devastating effects for both mother and baby.

Case: A 27-year-old primigravida at 32+3 weeks' gestation had a serum calcium level in excess of 2.75 mmol/L and evidence of HELLP syndrome. She underwent concurrent parathyroidectomy and Caesarean section. Neither the mother nor the neonate developed hypocalcemia postoperatively. The mother's parathyroid tissue was pathologically atypical, and a left hemithyroidectomy was performed at three months postpartum.

Conclusion: When hyperparathyroidism is diagnosed in the third trimester, concurrent parathyroidectomy and Caesarean section is a safe and reasonable option for management. This should be performed by a multidisciplinary team with careful monitoring of the calcium levels of both mother and neonate after surgery.

Résumé

Contexte : L'hyperparathyroïdie primaire constitue la cause la plus courante d'hypercalcémie au sein de la population générale. Bien qu'il s'agisse d'une complication rare de la grossesse qui est difficile à reconnaître pour les cliniciens, elle peut exercer d'importants effets dévastateurs tant chez la mère que chez l'enfant.

Cas : Une primigravide de 27 ans à 32+3 semaines de gestation présentait un taux sérique de calcium dépassant 2,75 mmol/l et des symptômes indiquant la présence du syndrome HELLP. Elle a subi, de façon concomitante, une parathyroïdectomie et une césarienne. Ni la mère ni le nouveau-né n'en sont venus à présenter une hypocalcémie postopératoire. L'examen pathologique a révélé que le tissu parathyroïde de la mère était atypique et une hémithyroïdectomie gauche a été menée à trois mois postpartum.

Conclusion : Lorsqu'une hyperparathyroïdie est diagnostiquée au cours du troisième trimestre, la tenue concomitante d'une parathyroïdectomie et d'une césarienne constitue une option sûre et raisonnable pour ce qui est de la prise en charge. Ces interventions devraient être menées par une équipe multidisciplinaire et s'accompagner d'une surveillance rigoureuse des taux de calcium de la mère et du nouveau-né après la chirurgie.

Key Words: Pregnancy, pregnancy complications, third trimester, hyperparathyroidism, Caesarean delivery, parathyroid neoplasms

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INTRODUCTION

Fewer than 200 cases of primary hyperparathyroidism in pregnancy have been reported since the condition was first recognized in 1931. To date, no case reports describing concurrent parathyroidectomy and Caesarean section have been published.

THE CASE

A 27-year-old primigravid woman presented to a rural emergency department in Northern Ontario at 31+5 weeks' gestation with a complaint of right upper quadrant abdominal pain, vomiting, and headache. She had a history of migraine headaches, but no other significant history. Her pregnancy had been complicated by prolonged hyperemesis gravidarum that had required treatment with ondansetron. The results of her routine prenatal investigations were unremarkable.

At the time of presentation, the patient had normal vital signs and was afebrile. Her physical examination revealed moderate epigastric tenderness. The findings on peripheral neurological and musculoskeletal examination were unremarkable. Her initial investigations revealed a normocytic anemia (hemoglobin concentration 90 g/L, mean corpuscular

volume 92.9 fL), thrombocytopenia (platelet concentration 99×10^9/L), and mild elevation of hepatic transaminase levels (serum AST 148 U/L, serum ALT 191 U/L). Urinalysis was negative for protein or blood. She was admitted to hospital with a provisional diagnosis of HELLP syndrome, and was immediately transferred to a tertiary care centre because of the gestational age of her fetus.

Following transfer, the patient underwent rehydration and subsequently showed some improvement. A critically high serum calcium level of 3.25 mmol/L was identified (normal 2.10 to 2.55 mmol/L), and serum phosphorus was low at 0.58 mmol/L (normal 0.81 to 1.45 mmol/L). After consultation with an endocrinologist, further evaluation showed elevated 24-hour urinary calcium excretion of 9.01 mmol/day (normal < 7.5 mmol/day) and an elevated parathyroid hormone level of 23.8 pmol/L (normal 0.8 to 5.7 mmol/L). This confirmed the diagnosis of primary hyperparathyroidism.

Conservative medical management with intravenous fluids and administration of a loop diuretic was initiated. An obstetrical ultrasound examination showed asymmetric fetal growth restriction (biparietal diameter 33+0 weeks; abdominal circumference 29+4 weeks; femoral length 29+5 weeks) in the third percentile for weight. Obstetrical Doppler studies showed an elevated systolic/diastolic ratio of 3.7 with normal end-diastolic flow. Biophysical profile was not assessed. Amniotic fluid index was normal at 17 cm. A maternal neck ultrasound examination showed a $1.7 \times 1.3 \times 1.3$ cm lobulated hypoechoic vascular mass which was thought to represent a parathyroid adenoma. After discussion with pediatric, otolaryngology, and endocrinology consultants, a decision was made to remove the neck mass and to deliver the baby concurrently. Two doses of betamethasone 12 mg were given intramuscularly 24 hours apart to assist with fetal lung maturation before delivery.

Caesarean section was performed under general anaesthesia at 32+3 weeks' gestation, and a vigorous live infant with ambiguous genitalia was delivered. The neonate was transferred to the NICU immediately after delivery. Following delivery, the patient was repositioned and prepared for a left parathyroidectomy. A $3.4 \times 2.2 \times 1.4$ cm mass was removed from the left superior parathyroid region of the neck (Figure). Histological assessment of the mass identified vascular invasion, coagulative necrosis, macronuclei, and cellular atypia, consistent with an atypical parathyroid adenoma.

Serum bound and ionized calcium levels were followed closely postoperatively, and oral calcium and vitamin D supplementation was provided. Neither the mother nor the baby demonstrated hypocalcemia postoperatively.

The infant was transferred to the nearest quaternary care hospital (the Hospital for Sick Children in Toronto) for endocrinological and urological evaluation. He was identified as a male (karyotype 46,XY) with hypospadias and a bifid scrotum. At three weeks of life, he underwent emergent hernioplasty for an incarcerated inguinal hernia which was later complicated by postoperative cellulitis. He recovered well on a short treatment course of broad spectrum antibiotics. He was discharged from the NICU in good health at age seven weeks.

Given the atypical nature of the maternal parathyroid tissue and the mother's age, a left hemithyroidectomy and lateral compartment inspection were performed at three months postpartum. The histology of this tissue was unremarkable.

DISCUSSION

Primary hyperparathyroidism occurs in 0.1% of the general population and is two to three times more common in females than in males.[1] The classical manifestations of primary hyperparathyroidism are those directly related to the hypercalcemic state; nephrolithiasis, musculoskeletal weakness and fatigue, mild psychiatric disturbances, and rarely osteitis fibrosa cystica can all be seen in patients with hypercalcemia.[2–5]

It is likely that many cases are undetected in pregnancy because of the calcium-shunting effect of the maturing fetus. Most commonly, primary hyperparathyroidism in pregnancy presents in the postpartum period with neonatal tetany.[6] Once delivered, the neonate no longer has access to calcium-rich maternal serum and is unable to mobilize calcium from bone because of suppression of parathyroid gland function. The consequent hypocalcemia leads to tetany. Neonatal hypocalcemia typically presents at five to 14 days after delivery, once transferred maternal calcium stores have been depleted.[7] Treatment is straightforward using oral calcium supplementation. Stillbirth and abortion, with an incidence as high as 17%, are other complications that have been reported.[8]

Maternal complications of primary hyperparathyroidism are similar to those found in the non-pregnant patient. One review of 16 cases of primary hyperparathyroidism found an incidence of preeclampsia of 25% among the women studied.[4] Case reports identifying this coincidence have also been published, although none have proposed a mechanism for the association with preeclampsia.[9–11] Other less common but serious complications include pancreatitis (up to 13%), pyelonephritis, and hypercalcemic crisis.[12] Fortunately, the last two are quite