Laparoscopic treatment of Cushing’s syndrome in a woman in late pregnancy – a case presentation

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Abstract

Hypercortisolaemia during pregnancy constitutes a serious threat to life of the mother and fetus and may be associated with adrenocortical carcinoma. The objective of this study is to present the usefulness of laparoscopic procedures in treating adrenal tumours in such cases. One 21-year-old woman, 24 weeks pregnant, with hypertension and Cushing’s syndrome due to a left adrenal tumour, underwent laparoscopic adrenalectomy followed by hydrocortisone replacement. Spontaneous delivery occurred at the 37/38th week of gestation. At 3 months postpartum the function of the remaining adrenal gland was found to be normal. Similarly, imaging tests, abdominal CT scan and chest X-ray revealed no abnormalities. Pregnancy is not a contraindication for performing complicated laparoscopic procedures unless they are planned in advance and done by an experienced team.

Key words: Cushing’s syndrome during pregnancy, laparoscopic adrenalectomy, adrenal tumour, pregnancy.

Introduction

Cushing’s syndrome during pregnancy is rare due to inhibition of gonadotropin-releasing hormone release by elevated cortisol resulting in anovulation. Hypercortisolaemia in pregnancy can lead to hypertension, diabetes, cardiomyopathy and other disorders constituting a risk to both mother and fetus. The maternal mortality rate has been found to be 5% [1]. The fetal mortality rate is as high as 25% [2]. About 2 in 3 pregnancies are complicated and the risk of perinatal mortality is markedly increased.

The prevalent cause of hypercortisolaemia in pregnancy is cortisol-secreting adrenocortical tumours (zona fasciculata). Additionally, the levels of adrenal androgens can be increased (zona fasciculata and reticularis). Cortisol can also be secreted by adrenocortical carcinoma, which should be considered in the differential diagnosis of adrenal tumours.

Case report

A 21-year-old woman, 24 weeks pregnant, presented with hypertension and overweight. An adrenal mass was demonstrated on ultrasound scan. She was admitted to an endocrinological clinic for further investigation and treatment.

She has had a 6-year history of untreated hypertension. The highest arterial blood pressure was 180/120 mmHg. Shortly before admission to the clinic she started antihypertensive therapy with
methyldopa. After admission, taking into account the possibility of pheochromocytoma, dopesty was replaced with the α-blocker doxazosin. Results of biochemical tests, normal diurnal methoxycatecholamines and normal plasma adrenaline, noradrenaline and dopamine levels excluded this possibility. Increased diurnal cortisol metabolite excretion, together with elevated plasma cortisol, loss of diurnal cortisol rhythm and total suppression of adrenocorticotropic hormone (ACTH), was consistent with Cushing’s syndrome due to primary adrenal hypercortisolaemia. The magnetic resonance imaging (MRI) confirmed the presence of a 57 mm × 40 mm left adrenal mass with smooth boundaries and no signs of local invasion situated on top of the left kidney and adjacent to the pancreas. The chemical shift MRI was suggestive of adrenocortical carcinoma as it demonstrated insufficient intracytoplasmic lipid for a diagnosis of an adrenal adenoma. Considering the above findings together with the risks associated with hypercortisolaemia and the young fetal age (being a contraindication for inducing labour), the decision about surgical treatment was reached by an interdisciplinary team. The patient was scheduled for laparoscopic adrenalectomy.

She was placed in the lateral decubitus position. Three 10 mm trocars and one 12 mm trocar were inserted at the usual sites below the left costal margin. The initial visual trocar was inserted directly (without the use of a Veress needle) following insufflation. Carbon dioxide was used for insufflation of the peritoneal cavity not exceeding the limits of intraabdominal pressure of 10 mmHg. The left adrenal vein was isolated, clipped and divided. A wide excision of the adrenal gland with the peri-adrenal fat was performed using a harmonic scalpel. The specimen was extracted using an Endo-bag. The operation time was 160 min. The blood loss was 100-150 ml. The fetal pulse monitored during the operation was normal. The postoperative biochemical suppression was managed with hydrocortisone replacement. Histopathology revealed an adrenocortical adenoma. The patient was discharged in good condition. At 37/38 weeks gestation she spontaneously delivered a healthy baby (neonatal transient jaundice was the only finding). At 3 months postpartum control tests showed normal function of the remaining adrenal gland and well-controlled hypertension. Imaging tests: abdominal computed tomography (CT) scan and chest X-ray showed no abnormalities.

Discussion
The diagnosis of hypercortisolaemia is made difficult by the physiological changes of pregnancy. During normal pregnancy, the cortisol levels can be transiently increased as high as 3-fold [3]. Additionally, pregnancy is associated with weight gain, and often glucose intolerance and hypertension. These symptoms can be a result of oversecretion of adrenocortical hormones. Therefore, adrenocortical adenomas constitute a challenge in terms of diagnosis and treatment. The decision about the treatment is made by an interdisciplinary team consisting of surgeons, gynaecologists, endocrinologists and anaesthesiologists, as well as other relevant clinical specialists.

Laparoscopic adrenalectomy is the mainstay of treatment for adrenal tumours. As a minimal invasive method it allows early postoperative mobilization, minimizes postoperative pain and the risk of complications related to wound healing, and substantially reduces the intraoperative blood loss. Pneumoperitoneum is the main risk, especially in pregnant patients. Until recently this has constituted the major contraindication to laparoscopy because increased intra-abdominal pressure can reduce placental blood flow, whereas absorption of the carbon dioxide used for insufflation can lead to fetal acidosis [4]. However, recent experience of laparoscopic adrenalectomy, cholecystectomy [5] or appendectomy suggests that, with the use of special precautionary measures, these procedures can be performed safely in pregnant women [6]. These measures include maintenance of pneumoperitoneum below 12 mmHg as well as avoidance of direct contact between the surgical instruments and the uterine muscle during the operation [7]. The progress in laparoscopic techniques also allows one to take care about cosmetic results due to e.g. SILS [8, 9].

Conclusions
Laparoscopic adrenalectomy in pregnant women is an effective and relatively safe procedure. Diagnosis and treatment of adrenal tumours in pregnant women requires interdisciplinary work.

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