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A RARE DIAGNOSIS IN 3RD TRIMESTER PREGNANCY OF FUNCTIONING LEFT PHAEOCHROMOCYTOMA AND PARAGANGLIOMA: A CASE REPORT

https://doi.org/10.15605/jafes.036.S65

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INTRODUCTION

Phaeochromocytoma or paraganglioma in pregnancy is extremely rare, with a frequency of 0.007% of all pregnancies. If unrecognized, it has high maternal and fetal mortality risk.

RESULTS

Malaysia

A 34-year-old G2P1 female at 34 weeks gestation with GDM on insulin was referred for further workup. She had previously been detected to have a left suprarenal mass (8.5 x 5.2 x 5.8 cm) by ultrasound during her first pregnancy, and underwent caesarean section (CS) at 36 weeks for severe pre-eclampsia with impending eclampsia. Postpartum, her anti-hypertensives were discontinued within 2 weeks. She subsequently defaulted further follow-up. She had paroxysmal symptoms of headache and palpitation once to twice a week during the current pregnancy. Blood pressure in the ward was <140/90. KUB ultrasound showed a heterogeneous mass with cystic component arising from the left suprarenal region (9.6cm x 7.7cm x9.6cm), with another smaller mass (6.0cm x 5.6cm) lateral to this. 24hour urine catecholamines revealed elevated epinephrine 7.55 times above the upper limit of normal (ULN) at 151.0 mcg/day (Normal: 0.5 -20.0) while norepinephrine and dopamine were raised 1.98 and 1.91 times above ULN respectively. The patient was then referred to an endocrine tertiary centre for expert multidisciplinary care. She was started on prazosin and underwent elective CS 1 week later at 36 weeks gestation, delivering a 2.7kg baby. An adrenal CT scan 3 weeks postpartum showed left suprarenal masses of mixed density and heterogeneous enhancement (9.4cm x 9.2cm x 8.3cm and 5.7cm x 6.4cm x 6.6cm). Her alpha-blocker was changed to Phenoxybenzamine 2 weeks prior to surgery. She underwent open adrenalectomy 3 months postpartum with excision of left paraganglioma (7 x 7 cm) and left phaeochromocytoma (10 x 10 cm) together with the normal-looking left adrenal gland.

CONCLUSION

In phaeochromocytoma and paraganglioma (PPGL) in pregnancy, multidisciplinary coordination is essential for effective management in terms of appropriate mode of delivery, timing of surgery, anaesthesia as well as adequate pre-operative medical preparation.

DROPOUT RATES AND RETENTION FACTORS OF A SINGLE-CENTRE WEIGHT MANAGEMENT CLINIC IN A TERTIARY HOSPITAL

https://doi.org/10.15605/jafes.036.S66

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INTRODUCTION

Weight management clinics are the mainstay of obesity care in Malaysia. The UiTM weight management clinic comprises a 7-visit programme over a 1-year period involving various specialties such as primary care physicians, endocrinologist, nutritionist, psychologist, and rehabilitation medicine. Each provides different aspects of obesity management. There is a high dropout rate for this clinic, thus, it is essential to recognize those who will benefit from it.

METHODOLOGY

This is a retrospective analysis of 145 patients attending the weight management clinic in UiTM from June 2018 to December 2020. All data were acquired through patients' medical records. Dropout rate is defined as the absence from 1 or more clinic visits at three monthly timepoints. Logistic regression analysis with SPSS version 22 was performed to identify factors predicting patients' retention to the programme.

RESULTS

A total of 145 patients attended the clinic. Dropout rate at 3 months was 37% (n=53), cumulative rate at 6 months was 48% (n=70) and 59% (n=86) at 12 months. 59 patients (41%) completed the programme. Mean age was 39.1 \pm 13.3 years and mean BMI 44.9 \pm 10.2 kg/m². Patients with dyslipidemia comprised 75% of the cohort (n=109). Patients with a baseline body mass index (BMI) of >40 kg/ m² and known dyslipidemia showed higher retention in the programme. Those with dyslipidaemia had a 4-fold increase in retention (OR 4.81 (CI 1.02,22.69)), p= 0.048) while those with baseline BMI of >40 kg/m² had a 5-fold higher retention (OR 5.53 (CI 1.37, 22.27) p=0.016).

CONCLUSION

There is a high dropout rate (59%) with only 41% retention in our weight management programme. There may be multiple factors associated with this occurrence. Our study showed that those with known dyslipidemia and those with baseline BMI of more than 40 kg/m² were more likely to complete the programme and benefit from it.

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SEMINOMA ARISING FROM TESTICULAR AND OVARIAN REMNANTS HERALDS THE EMERGENCE OF A RARE MALE OVOTESTICULAR DISORDER OF SEXUAL DEVELOPMENT

https://doi.org/10.15605/jafes.036.S67

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INTRODUCTION

Ovotesticular Disorder of Sex Development (OT DSD) or true hermaphroditism is a very rare subset of DSD and accounts for only 5% of cases. It has great phenotypic variability and poses diagnostic challenge to clinicians. It usually presents in childhood with ambiguous genitalia, characterized by histologic demonstration of ovarian and testicular tissues within the same individual.

RESULTS

We describe a 22-year-old man with OT DSD complicated by seminoma. He was delivered preterm with ambiguous genitalia and was lost to follow-up. He was raised as a female, at 14 years old, his teachers referred him due to concerns of male phenotype while participating in competitive sports. Phenotypically, he was a developed male with a micropenis, hypospadia, left scrotal cystic structure and empty right scrotum. Chromosomal analysis revealed 46XY and presence of SRY gene. Radiological imaging at age 16 showed fully developed Mullerian structures, with a single cervix and incomplete septate uterus, and an oval structure suggestive of testes at the left hemipelvis. Cysto-genitoscopy demonstrated normal urethra without prostatic urethra, opening at posterodistal bladder neck with blood clots likely representing the vagina. Laparoscopy identified tubulo-nodular structure inside the pelvis suggestive of vas deferens with suspicious early malignant changes. Wolffian remnant and bicornuate uterus were present with a right Fallopian tube with suspicious hydro-corpus. He was advised surgery, however, he defaulted again. He consulted again at age 22 due to pyuria, suprapubic pain and painless cyclical haematuria. Imaging studies demonstrated pyometra, bulky left ovary and bilateral undescended testes suspicious of malignant transformation. Exploratory laparotomy, gonadectomy, subtotal hysterectomy and left orchidectomy were performed. Histopathological examination revealed seminomas arising from testicular and ovarian remnants with suppurative inflammation in the uterus. He was provided testosterone replacement post operatively, and received chemotherapy (etoposide, bleomycin and platinum).

CONCLUSION

The complexity of the case exemplified the pivotal role of multidisciplinary input from various specialties.