EP_A138

XANTHOMATOUS HYPOPHYSITIS PRESENTING WITH PROGRESSIVE HYPOPITUITARISM AND PITUITARY APOPLEXY

https://doi.org/10.15605/jafes.039.S1.149

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INTRODUCTION/BACKGROUND
Primary hypophysitis is a rare condition characterised by isolated inflammation of the pituitary gland and infundibulum. Among the histopathological subtypes, xanthomatous hypophysitis is rare.

CASE
We describe a 28-year-old female with xanthomatous hypophysitis (XH), its clinical course over 4 years, as well as the transsphenoidal surgery outcome.

She first presented 4 years ago with intermittent headaches and amenorrhea for four months. Investigations showed that she had central hypogonadism (FSH 0.9 IU/L, LH <0.1 IU/L, estradiol 77 pmol/L) and hyperprolactinemia (prolactin 2195 m IU/L). She continued to have central hypogonadism despite prolactin normalisation and was subsequently started on an oral contraceptive pill. A pituitary MRI showed a normal pituitary gland and no stalk lesion. One year later, she developed arginine vasopressin (AVP) deficiency, leading to desmopressin treatment. She was diagnosed with autoimmune hypophysitis, was started on prednisolone and 50 mg of azathioprine daily. However, she developed transaminitis after two months of treatment and it was withheld.

Two and a half years later, she developed central hypothyroidism and central adrenal insufficiency and was started on hydrocortisone and levothyroxine replacement. She was admitted to the ward two months later for pituitary apoplexy, and a repeat MRI showed an enlarging sellar cystic mass with mass effect and increasing T1-weighted hyperintensity within. The Humphrey visual field examination was normal. She underwent transsphenoidal hypophysectomy which revealed a sellar mass with a thickened capsule containing cheesy material. Histopathological findings were consistent with xanthomatous hypophysitis. Three months after surgery, she still had panhypopituitarism, and a repeat MRI revealed a pituitary gland within the sella with no evidence of recurrent disease.

CONCLUSION
Surgery is the treatment of choice for patients with XH who present with pituitary apoplexy. Close follow-up and monitoring of hormonal recovery and disease remission are essential to observe the clinical progression of this rare disease.

EP_A139

FERTILITY IN CONGENITAL ADRENAL HYPERPLASIA: A CASE REPORT

https://doi.org/10.15605/jafes.039.S1.150

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INTRODUCTION/BACKGROUND
Congenital adrenal hyperplasia (CAH), an autosomal recessive disorder affecting cortisol biosynthesis enzymes, results in virilization in affected females. Fertility challenges are often faced by females with CAH. We present two women with CAH who achieved successful spontaneous pregnancy and their pregnancy outcomes.

CASE 1
Ms. NAS, a 28-year-old Malay female, was diagnosed with 21-hydroxylase deficiency (21-OHD) at birth (presentation: ambiguous genitalia) and underwent corrective surgery at one year of age. She attained menarche at the age of 12 before having secondary amenorrhea for one year at 13 years old. She had normal menstrual cycles while on hydrocortisone 10 mg BD. Pre-pregnancy investigations were as follows FSH 4.04 IU/L, LH 2.22 IU/L, testosterone 2.22 nmol/L, dehydroepiandrosterone Sulphate (DHEA-S) 1.040 umol/L (NR 2.68-9.23). She safely delivered her baby via spontaneous vaginal delivery.

CASE 2
Ms. NN, a 22-year-old female, was diagnosed with CAH at 1 month of life presenting with a salt-losing crisis. She developed precocious puberty at 9 years old due to poor compliance to treatment. She was on triptorelin (Decapeptyl) sc for 2 years until the age of 12. She successfully conceived at the age of 21. Pre-pregnancy, she was treated with T hydrocortisone 5 mg/5 mg/7.5 mg TDS and T fludrocortisone 0.1 mg OD. Her hormonal level preconception were as follows: testosterone <0.1 nmol/L, 17 hydroxyprogesterone (17 OHP) 71.9 nmol/L (elevated), DHEA-S 0.349 umol/L (suppressed). Her pregnancy was uneventful, and successfully delivered her baby surgically after having transverse lie at term.