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A Rare Case of Co-Existence Pituitary Macroadenoma with Mayer-Rokitansky-Kuster-Hauser (MRKH) Syndrome

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INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome or Müllerian agenesis is a congenital failure of the Müllerian duct to develop, resulting in complete or partial absence of the cervix, uterus, and vagina. It can be isolated (MRKH type I) or associated with renal, vertebral, auditory and cardiac defects (MRKH type II). It is not known that pituitary disease has any association with this condition.

We report a patient who has MRKH type I and a pituitary macroadenoma, diagnosed concomitantly.

CASE

A 17-year-old lady was referred with primary amenorrhoea, occasional headache, nausea and lethargy. She denies any reduction or loss of vision. On clinical assessment, she had normal secondary female sexual characteristics, with Tanner stage 3. There was no significant family history. Hormonal investigations showed high prolactin level; 4200 miU/L (post dilutional). Estradiol was low, 37 pmol/l (93–1400 pmol/l), as well as FSH 2.38 U/L, LH 0.96 U/L, progesterone <0.3 nmol/L and testosterone <0.45nmol/L. Cortisol level 206 nmol/L, FT4 7.5 pmol/L with TSH 5.091 mIU/L. MRI of pituitary showed 2.7 cm (AP) x 3.7 cm (W) x 4.6 cm (CC) pituitary macroadenoma, with mass effect, infiltration into left cavernous sinus and encasement of cavernous portion of left ICA. MRI pelvis showed absent uterus, cervix and 2/3 upper vagina confirming Müllerian hypoplasia. Cytogenetics showed 46XX. Diagnosis of Mayer Rokitansky Kauser Hauser Syndrome and panhypopituitarism secondary to pituitary macroadenoma was made. She was treated with hydrocortisone, levothyroxine and cabergoline. Follow up MRI has shown reduction in tumour size. However, she has remained amenorrhoeic.

CONCLUSION

The estimated prevalence of MRKH syndrome is one in 4500 female births. The etiology of MRKH syndrome remains unclear. There is no known association with pituitary disease. To the best of our knowledge this is the first case of co-existing MRKH syndrome and pituitary adenoma reported from the ASEAN world.

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The Hiding Giant: A Case of an Incidental Functioning Metastatic Adrenal Carcinoma

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INTRODUCTION

The majority of incidental adrenal tumours diagnosed by imaging are non-functioning and small in size. However, in this case, a follow through from a CT angiogram of lower limbs led to the findings of a giant functioning metastatic adrenal carcinoma.

CASE

A 59-year-old lady with hypertension and T2DM on insulin was admitted for seizure with hypoglycaemia and hypokalaemia secondary to poor oral intake due to Left Lower Limb Necrotising Fasciitis. A CT angiogram of the left lower limb found necrotic aortocaval lymph nodes. Upon follow up a month later, to rule out either TB or malignancy, a CT abdomen/pelvis was done and showed a huge heterogenous suprarenal mass (10 x 11.6 x 8.7 cm) with metastases to the lymph nodes and lungs. On examination, patient was cushingoid and generally weak. Hormonal screen suggests Cushing's syndrome and hyperandrogenism. A further CT adrenal protocol supports the diagnosis of adrenocortical carcinoma with referrals made for surgical intervention in Putrajaya Hospital but patient died of Severe Hospital Acquired Pneumonia with Upper GI Bleeding before surgical review.

CONCLUSION

Even though her admission was protracted there was no early identification of Cushing syndrome until CT abdomen/pelvis was done. Therefore, clinical suspicion of cortisol hypersecretion is crucial once there is hypertension with hypokalaemia. Hyperandrogenism, is extremely rare in adrenal carcinoma. However, this patient exhibited raised DHEAS levels combined with increased testosterone which support a diagnosis of a malignant adrenocortical tumour. This patient did not receive any treatment for her cancer. Should she have survived, surgical resection would be suggested despite lacking data for its benefit in metastatic disease. However, there is evidence that adrenalectomy coupled with chemotherapy may provide better survival.