CR-GE-39

SELECTIVE ACTH SAMPLING IN LOCALIZING SOURCE OF ACTH IN VON HIPPEL LINDAU DISEASE WITH PANCREATIC NEUROENDOCRINE TUMOUR AND RENAL CELL CARCINOMA

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INTRODUCTION

Crushing's sundrome (CS) in a patient with VHL has been attributed to a number of possible causes; pancreatic NET and renal cell carcinoma. The precise location of ectopic ACTH aid enormously in the management of VHL.

CASE

A 31-year-old woman with Type 2 diabetes and family history of VHL presented with florid features of CS in her second trimester of pregnancy. Investigations supported a diagnosis of ACTH dependent CS. She underwent emergency caesarean section due to pre-eclampsia at 28 weeks gestation. MRI pituitary was normal. CT abdomen showed an enlarged pancreas, almost completely cystic and a right renal mass (3.7x2.7x4 cm). Serum chromogranin A was elevated. Twenty-four hour urinary free metanephrine was normal. Selective ACTH sampling was done with bilateral IPSS to elicit source of ACTH. Increased gradient of ACTH level compared to the periphery was detected from the hepatic vein that drains the pancreas (hepatic vein:138.92 pg/mL, IVC:115.12 pg/mL, renal vein:100.2 pg/mL). Total pancreatectomy and right nephrectomy were performed. A week after surgery, am cortisol was 103 nmol/L. HPE identified a solid tumour (16X12X12X mm) at the pancreatic tail which stained positive to Chromogranin A, synaptophysin and ACTH with mitoses of 0-1/10hof and a Ki67 index of 2%. The renal mass was a Grade 1 clear cell renal cell carcinoma. Two months later, there was resolution of cushingoid features.

CONCLUSION

When managing VHL with CS, there is always a possibility of more than one source of ACTH production. The use of selective ACTH sampling may be considered where functional imaging (DOTATATE) is unavailable to delineate the cause.

KEY WORDS

ectopic cushing's von hippel lindau, acth dependent cushing's

CR-GE-40

RECURRENT CAVERNOUS SINUS THROMBOSIS – AN UNUSUAL COMPLICATION OF CUSHING'S DISEASE

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INTRODUCTION

Cushing's disease increases risk of venous thromboembolism. However, its association with cavernous sinus thrombosis (CST) is rarely reported. We report a case of Cushing's disease complicated with recurrent bilateral CST.

CASE

She first presented at age 14 with pituitary apoplexy. Initial investigations showed non-functioning pituitary adenoma. Transphenoidal surgery was done. Left CST was diagnosed 6 months post-surgery when she had severe headache. This prompted further investigations which revealed Cushing's disease. She was started on warfarin, but developed right CST within 2 months and was switched to enoxaparin. Her cortisol levels remain elevated on surveillance and one year later, a recurrent pituitary macroadenoma was seen on MRI encroaching the left cavernous sinus with left CST. She subsequently underwent petrosal craniotomy for tumour debulking. Enoxaparin was stopped post-surgery, but 6 months post-surgery her MRI now shows presence of right pituitary macroadenoma with right CST with no normalisation of cortisol.

CONCLUSION

CST is a rare complication of Cushings and may occur due to hypercoagulablity due to hypercortisolism and compression from tumour extension from the pituitary fossa.

KEY WORDS

cushings, cavernous sinus thrombosis, recurrence