



CUSHING'S DISEASE

Shrushti Dalal^{1*}, Dr. Yashesh Dalal² and Dr. Chetan Trivedi³

^{1*}Final Year M.B.B.S Student, G.M.E.R.S Gotri, Vadodara, Gujarat, India.

²M.B.B.S, M.S General Surgery, M.Ch. Consultant Neurosurgeon, Srushtil Hospital, Vadodara, Gujarat, India.

³M.B.B.S, M.D. D.M Neuro, Consultant Neurologist, Alkapuri Vadodara, Gujarat, India.

***Corresponding Author: Shrushti Dalal**

Final Year M.B.B.S Student, G.M.E.R.S Gotri, Vadodara, Gujarat, India.

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ABSTRACT

The term "Cushing's disease" is reserved for Cushing's syndrome caused by excessive secretion of corticotropin by pituitary corticotroph tumors.^[2] It is the most common form of the syndrome. Cushing's disease, if not treated, together with the eventuality chronically raised glucocorticoid levels can lead to severe metabolic disturbances including, diabetes mellitus, hypertension, obesity, muscle wasting and osteoporosis. We present the case of a 19 year old Asian female presenting with headache since a month, diagnosed with a pituitary adenoma on CT scan and treated by microendoscopic transsphenoidal surgery.

KEYWORDS: Cushing's disease, Transsphenoidal microsurgery, Pituitary tumours.

INTRODUCTION

Cushing's syndrome is attributed to the clinical manifestations induced by chronic exposure to excess of glucocorticoid levels.^[3,4] There are three pathological conditions that can cause the chronic overproduction of endogenous cortisol in man: the most usual is Cushing's disease where adrenocorticotrophic hormone (ACTH) is overproduced by a pituitary corticotroph adenoma, occasionally ACTH can be produced in an 'ectopic' manner by a non-pituitary tumour, lastly cortisol can be directly over-secreted by one or (rarely) the two adrenals that have become tumourous, either benign or malignant. Cushing's disease is the most common cause of spontaneous Cushing's syndrome, occurring in 59–69% of Cushing's patients.^[5] It results from the hypersecretion of adrenocorticotrophic hormone (ACTH) by a pituitary corticotroph adenoma. The finest treatment option of Cushing's disease is when the causative corticotroph adenoma can be completely removed by the trans-sphenoidal approach, with sufficient skill to retain the normal anterior pituitary function.^[6]

CASE REPORT

We present the case of a 19 year old Asian female, with a complain of headache since the past 1 year, which was diffuse in nature, spontaneous in origin, dull aching type, not relieved by taking medications, no aggravating and relieving factors, no diurnal variation and no radiation of pain. There was no associated complaint of any fever, visual disturbance, sinusitis, vomiting, giddiness, stress or anxiety. There was no history of any trauma. She also complained of excessive gain of weight, especially in the

trunk region, approximately 26.45 pounds in the past 6 months. She had no menstrual abnormality. On examination, she was conscious, cooperative, well oriented to time, place, and person. She was noted to have truncal obesity. Her vitals were stable. She had a moon face and purple striae over her abdomen and acne all over the face. She had hyperpigmentation on knuckles and face. There was no muscle wasting. Her random blood sugar was high 176.5 mg/dl, her serum tested negative for HIV and HBSAg, PT, APTT were within the normal range, Serum creatinine, ALT, Na⁺, K⁺ were all within the normal range. Hb was 10.6 gm%, WBC count was low with 12,000 cu/mm. Her hormonal assay showed FSH 1.73 mIU/ml, LH 0.74 mIU/ml, Prolactin 35.95 n/ml, Growth hormone 0.2 ng/ml. Her T3, T4 and TSH was normal. Plasma 8am cortisol level was 23.86 mcg/dl. A.C.T.H level was 34.4 pg/ml.

Her perimetry assessment was normal. CT scan showed a small well defined, hyperdense soft tissue in the sella, extending up to suprasellar region, 1.4x1.6x1.2 cms in measurement. MRI revealed a haemorrhagic pituitary adenoma.

She was treated by complete excision of the pituitary adenoma through micro-endoscopic transsphenoidal surgery.

Patient was advised to continue long term surgical and endocrinological follow up.

DISCUSSION

Cushing's disease or pituitary- dependant Cushing's syndrome, is a severe endocrine disease caused by a

corticotroph pituitary tumour and associated with an increased morbidity and mortality.^[7]

Transsphenoidal selective microadenectomy by an experienced surgeon is the treatment of choice for the vast majority of patients with Cushing's disease. However, it provides long-lasting remission in only 49–59% of cases, emphasising the need for other effective treatments to lower ACTH in Cushing's disease.^[1]

Patients with persistent or recurrent Cushing's disease require additional treatments, including pituitary radiotherapy, adrenal surgery, and/or medical therapy.^[8,9] Pituitary radiotherapy is effective in controlling cortisol excess in majority of the patients, but it is associated with an appreciable risk of hypopituitarism.^[10] Adrenal surgery is followed by an accelerated and clear-cut control of cortisol excess in nearly all patients, but it induces adrenal insufficiency.^[11,12] Medical therapy has recently acquired an important role compared to the past. Currently, it is used as a presurgical treatment, particularly for severe disease, or as postsurgical treatment, in cases of failure or incomplete surgical tumour resection, or as a bridging therapy before, during and after radiotherapy while awaiting for the results or in selected cases, as the primary therapy, mainly when surgery is not an option.^[13,14]

CONCLUSION

The treatment of choice for patients with Cushing's disease is transsphenoidal microadenectomy, if a clearly circumscribed microadenoma can be identified and resected.^[15,16] Among experienced hands, the cure rate is about 80 percent after the initial surgery; second operations tend to be less successful.

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