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ITSENKO-CUSHING SYNDROME: A SET OF SYMPTOMS THAT OCCUR ON THE BACKGROUND OF HYPERCORTISOLISM

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Annotation

Cushing syndrome is a hormonal disorder caused by prolonged exposure to inappropriately high levels of plasma glucocorticoid (also referred to as cortisol) hormones. Glucocorticoid hormones maintain glucose regulation, suppress the immune response and are released as part of the body's response to stress. The production of cortisol from within the cortex of the adrenal glands is regulated by a small gland just below the brain called the pituitary gland.

Keywords: adrenocortical hyperplasia, adrenocorticotrophic hormone, purpura, moon face, serum cortisol, corticotrophin.

There are several causes of Cushing syndrome:

- Adrenocortical hyperplasia (overgrowth of adrenal cortex) secondary to pituitary overproduction of adrenocorticotrophic hormone (ACTH)
- Benign or malignant adrenal tumours that release excess glucocorticoids into the blood
- Ectopic ACTH sydrome – secretion of ACTH by malignant or benign tumours arising in structures other than the pituitary or adrenal glands
- Exogenous steroid administration – usually from administration of high doses of systemic corticosteroids such as prednisone for blistering diseases, asthma, arthritis and other inflammatory diseases.

The most distinctive features of Cushing syndrome are the deposits of fat over the clavicles and back of the neck (buffalo hump), the rounded puffy face (moon face, tomato face) with the contrasting slender wasted limbs. Bones are weakened, and activities such as bending, lifting or rising from a chair may lead to backaches, rib and spinal fractures. Many people suffer severe fatigue, weak muscles, high blood pressure, high blood sugar, irritability, anxiety and depression.

Signs and symptoms appearing in the skin include:

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- Easy bruising (purpura) and purple striae of the skin over the abdomen, buttocks and thighs
- Telangiectatic cheeks (broken capillaries)
- Fragile skin and poor wound healing
- Acne and hirsutism (excessive hair growth in women on their faces, necks, chests, abdomens and thighs)
- Women may also show clitoral hypertrophy and male-pattern baldness

Although the diagnosis of Cushing syndrome may be made clinically, it is important to confirm it and explain the reasons for it. Laboratory tests may include:

- Serum cortisol, measured at a specific time of day or night
- Salivary cortisol test, usually done late at night
- 24-hour urine cortisol to determine overall cortisol production
- Dexamethasone suppression test: dexamethasone should suppress cortisol secretion but this does not occur in Cushing syndrome
- Corticotrophin (ACTH) stimulation test: ACTH should stimulate cortisol production but in Cushing syndrome there may be no response

Imaging to locate a tumour may include CT scan, MRI or ultrasound.

Treatment depends on the cause of the glucocorticoid excess and may include surgery, radiation, chemotherapy or drug therapy.

Exogenous steroid administration:

- Reduce the dose of corticosteroid drug to lowest dose adequate for control of the original disease
- Once control established, consider increasing the dose of corticosteroids and giving on alternate days to lessen side effects.

Pituitary tumours:

- Most commonly treated by surgical removal of the tumour, known as transsphenoidal hypophysectomy
- Where surgery has failed or as an alternative to surgery, radiation therapy to the pituitary gland over a 6 week period

Adrenal tumours: surgery is the mainstay of treatment for benign as well as cancerous tumours of the adrenal glands. Prognosis is poor for cancerous tumours.

Ectopic ACTH syndrome: removal of cancerous tissue that is secreting ACTH through surgery, radiotherapy, chemotherapy, immunotherapy, or a combination of these treatments.

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