



Fig. 17.14 The systemic features of Cushing's syndrome.

**Secondary hyperaldosteronism.** When renal glomerular filtration is reduced, for example through a fall in blood volume, the renin-angiotensin system (Fig. 17.12) stimulates aldosterone secretion from the zona glomerulosa in an attempt to correct this. This physiological response is known as secondary hyperaldosteronism, which is by far the commonest form of hyperaldosteronism.

**Diagnosis.** The diagnosis of primary hyperaldosteronism rests on two criteria: plasma aldosterone must be raised while renin is low. This is to distinguish it from secondary hyperaldosteronism, in which aldosterone levels are raised but are an appropriate response to high renin levels.

**Hypersecretion of sex steroids**

Some adrenal cortical adenomas secrete sex steroids, most commonly androgens. In Cushing's syndrome, quantities of androgens are occasionally secreted along with the glucocorticoids, causing virilisation of females, especially those with adrenocortical carcinomas.

Early congenital enzyme defects of the pathways of steroid synthesis may result in excess production of sex steroids. The most rare example is 'congenital adrenal hyperplasia' due to deficiency of the enzyme 21-hydroxylase, needed for the synthesis of both cortisol and aldosterone (Fig. 17.15).

Failure of cortisol production leads to increased ACTH secretion, resulting in hyperplasia of the adrenal cortex. The production of androgens occurs before the metabolic block caused by the enzyme deficiency, and their excessive secretion results in masculinisation of females and precocious puberty in males. 21-Hydroxylase deficiency is serious because deficiency of mineralocorticoids causes life-threatening salt loss unless replacement therapy is given.

**Tumours**

**Adenoma.** In addition to those 'functioning' adrenal cortical adenomas that present by causing Cushing's or Conn's syndromes, a clinically unsuspected 'non-functioning' adenoma occurs in about 2% of adults. The adenoma is a pale yellow circumscribed nodule, perhaps 20–30 mm in diameter (Fig. 17.16). The cells have clear cytoplasm owing to their high lipid content (Fig. 17.17).

**Carcinoma.** Adrenal cortical carcinoma is rare; these tumours are usually hormone-secreting, with a tendency to produce androgens. They are commonly large (over 100 g) and exhibit invasive growth. Examination of the adjacent adrenal cortex and that of the opposite gland may give a clue as to the function of the neoplasm; glucocorticoid-secreting tumours will suppress ACTH, resulting in atrophy of the non-neoplastic adrenal cortex.

# General And Systemic Pathology 5th Edition

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