Hypothalamic disorders

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Hypothalamic Infiltration Disorders

- These disorders -- including those associated with sarcoidosis, histiocytosis X, amyloidosis, and hemochromatosis -- frequently involve both hypothalamic and pituitary neuronal and neurochemical tracts. Consequently, diabetes insipidus occurs in half of patients with these disorders. Growth retardation is seen if attenuated GH secretion occurs before pubertal epiphyseal closure. Hypogonadotropic hypogonadism and hyperprolactinemia are also common.

Cranial Irradiation

- Cranial irradiation may result in long-term hypothalamic and pituitary dysfunction, especially in children and adolescents who are more susceptible to damage following whole-brain irradiation and neck therapeutic irradiation. The development of central hypothalamic dysfunction correlates strongly with irradiation dosage and the time interval after completion of radiation therapy. Specific studies of pituitary function are even more variable, but a median delay of 20 to 30 years occurs after cranial irradiation. Hypothalamic-pituitary dysfunction usually reflects hypothalamic damage rather than absolute destruction of pituitary cells. Hypothalamic-pituitary hormonal abnormalities are most commonly followed by gonadotropin deficiencies. Growth hormone deficiency is also a common problem. Although growth hormone deficiency is readily apparent, anterior pituitary function should be evaluated over the long term in previously irradiated patients, and replacement therapy instituted when appropriate (see below).

Developmental Hypothalamic Dysfunction

- Kallmann syndrome This rare autosomal recessive disorder is characterized by anosmia or hyposmia, and failure of GnRH synthesis and is associated with anosmia or hyposmia due to olfactory bulb agenesis or hypoplasia. The syndrome may also be associated with color blindness, optic atrophy, nerve deafness, cleft palate, renal abnormalities, cryptorchidism, and neurologic abnormalities such as mirror movements. (Chap. 335). The syndrome may also be associated with color blindness, optic atrophy, nerve deafness, cleft palate, renal abnormalities, cryptorchidism, and neurologic abnormalities such as mirror movements. (Chap. 335).

- Pituitary tumors Hypothalamic hamartomas and gangliocytomas may arise from astrocytes, oligodendrocytes, and neurons. They may be associated with hypothalamic hamartomas. They may be associated with hypothalamic hamartomas. These tumors may secrete hypothalamic neuropeptides including corticotropin-releasing factor (CRF), CRF-related peptides, and CRF-related peptides, and occasionally produce Cushing's syndrome, with long-acting GnRH analogues effectively suppressing gonadotropin secretion and causing sexual development. Finally, hamartomas are also associated with craniopharyngiomas, which may produce amenorrhea and hypogonadism. Hypothalamic hamartomas are often confused with the hypothalamic hamartomas which may produce amenorrhea and hypogonadism. Hypothalamic hamartomas may be the cause of growth hormone deficiency or intracranial hypertension of hypothalamic hamartomas. Hypothalamic hamartomas and gangliocytomas usually occur in childhood and usually present with visual loss. Adults have more aggressive tumors; about 75% are associated with neurofibromatosis.
The hypothalamus is subject to injury from mass lesions, granulomatous disorders, infections, and vascular insults involving the anterior and posterior hypothalamic regions. Lesions involving the anterior and preoptic hypothalamic regions cause paradoxical vasoconstriction, tachycardia, and hyperthermia. Acute hyperthermia usually occurs as a hemorrhagic insult but may be associated with central disorders of thermoregulation. Central disorders of thermoregulation result from posterior hypothalamic damage. The periodic hypothermia syndrome comprises episodic attacks of rectal temperatures <30°C, sweating, vasodilation, vomiting, and bradycardia (Chap. 20). Damage to the ventromedial nuclei by craniopharyngiomas, hypothalamic trauma, or inflammatory disorders may be associated with hyperphagia and obesity. This region appears to contain an energy-satiety center where melanocortin receptors are influenced by leptin, insulin, POMC products, and gastrointestinal peptides (Chap. 71). Median eminence involvement results in diabetes insipidus in about 50% of patients. Hypothalamic gliomas in early childhood may be associated with a diencephalic syndrome characterized by progressive encephalopathy and growth failure. Polydipsia or hypodipsia, associated with damage to central osmoreceptors located in preoptic nuclei (Chap. 329), may be associated with increased somnolence and disturbed sleep cycles as well as obesity, hypothermia, and emotional outbursts. Lesions of the central hypothalamus may stimulate sympathetic neurons, leading to elevated serum catecholamine and cortisol levels. These patients are predisposed to cardiac arrhythmias, hypertension, and gastric erosions.