Head and Neck Manifestations of Endocrine Disorders

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KEYWORDS

• Endocrine disorders • Pituitary • Thyroid • Parathyroid • Adrenal • Diabetes

KEY POINTS

- The endocrine system produces hormones that regulate essential body functions.
- Endocrine dysfunctions can lead to various forms of disease that negatively impact the patients' well-being and quality of life.
- Endocrine disorders have distinguishable manifestations, especially in the head and neck region, in which the dentist should be familiar with.
- Dental management of patients with endocrine disorders requires special knowledge and considerations.

Introduction

The endocrine system is a collection of glands that synthesizes and releases its hormones directly into the bloodstream. These hormones are responsible for fundamental and essential body processes, such as metabolism, tissue growth, homeostasis, sexual development, reproduction, immune regulation, cognitive function, and emotional stabilization. The endocrine glands are highly controlled by the nervous system and by the sensitive biofeedback mechanism that maintain well-balanced levels of hormones throughout the body. Any shift in this balance can significantly disturb body function and lead to disease manifestation that can negatively impact the individual's health and well-being.

Pituitary gland disorders

The pituitary gland is commonly referred to as the '"master gland" owing to its essential involvement in the functions of several organs. It occupies the sella turcica, a bony compartment of the sphenoid bone, and receives its stimulation directly from the hypothalamus.[1]

The intermediate and posterior lobes of the pituitary have limited function of storing and secreting melanocyte-stimulating hormone and antidiuretic hormone, respectively. In contrast, the anterior pituitary lobe is responsible for synthesizing and secreting several hormones:

- Growth hormone (GH),
- Puberty hormones (gonadotropins),
- Thyroid-stimulating hormone,
- Prolactin, and
- Adrenocorticotrophic hormone (ACTH).

Hypopituitarism

Etiology and epidemiology

Several etiologic factors can be responsible for pituitary hormone deficiency. Conditions affecting the pituitary gland itself (eg, genetic, neoplastic, infectious, iatrogenic, and traumatic) evolve into primary hypopituitarism, whereas dysfunction of the hypothalamus leads to secondary hypopituitarism.[2] Owing to the multifunctional activity of this gland, hypopituitarism can affect individuals' growth, puberty, thyroid, and adrenal functions. GH and gonadotropins have the highest susceptibility for deficiency among all pituitary hormones with the former being highly linked to head and neck manifestations.[3] Several earlier studies have estimated the prevalence of GH deficiency in pediatric population to fall between 1:1800 and 1:30,000.[4–7]

Clinical manifestations

GH deficiency during childhood is known as pituitary dwarfism. The condition is known for the low growth velocity, short stature, and abundant clinical features. In contrast, GH deficiency during adulthood has been associated with less dramatic effect and nonspecific clinical features. The manifestations of hypopituitarism in the head and neck region are summarized in Table 1.[8–14]

Differential diagnosis

Idiopathic short stature, hypothyroidism, osteogenesis imperfecta, and other skeletal dysplasia are included in the differential diagnosis of hypopituitarism.

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Table 1 Head and neck manifestations of the onset of hypopituitarism during childhood and adulthood

Childhood hypopituitarism	Adulthood hypopituitarism
Hypognathic maxilla and mandible that run proportionally with smaller facial features	Immobile expression
Smaller size of the jaws with normal sized teeth. This results in dental crowding and malocclusion.	Thin lips, scant eye brows, and loss of eyelashes
Exfoliation pattern of deciduous dentition is delayed with consequent retardation in the eruption of permanent teeth.	No specific oral manifestations have been described.
Agenesis of third molars (common) and maxillary incisors (rare).	
Amelogenesis imperfecta (very rare).	

Hyperpituitarism

Etiology and epidemiology

Excessive production of GH is the most abundant form of hyperpituitarism. The disorder is known as acromegaly if it occurs during adulthood (around the age of 40), whereas gigantism is the childhood counterpart of the same disorder (Figs. 1 and 2). The incidence of acromegaly is estimated to be 3 to 4 cases per million per year,[15] although there have been only 100 reported cases of gigantism in medical literature.[16] GH hypersecretion may be caused by:

- Primary pituitary adenomas, and
- Secondary pituitary hypersecretion owing to excessive production of hypothalamic GH releasing hormones or genetic disorders, such as multiple endocrine neoplasia type-1, McCune-Albright syndrome or Carney complex.

Clinical manifestations

Gigantism is characterized by generalized overgrowth of body parts while maintaining symmetry and proportionality. Acromegaly, in contrast, is characterized by progressive acquired disfigurement involving the face and extremities. The craniofacial characteristics of both conditions are summarized in Table 2.[17–22]

Differential diagnosis

Familial tall stature, cerebral gigantism (Sotos syndrome owing to NSD1 gene mutation), estrogen receptor mutation, and Weaver syndrome are included in the differential diagnosis for hyperpituitarism.

Treatment considerations

Early intervention by endocrinologists to correct the underlying hormonal disorder is required to prevent further growth

Fig. 1 Head and neck manifestations of pituitary gigantism. (*From* Kleinschmidt-DeMasters K. Pituitary gland. In: Rosai J, editor. Rosai & Ackerman's surgical pathology. 10th edition. Elsevier; 2011:2441–65; with permission.)

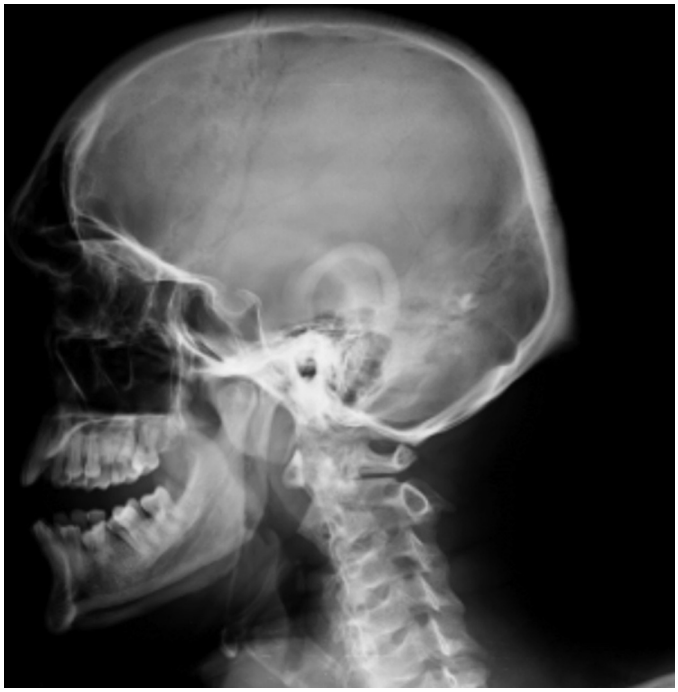

Fig. 2 Lateral skull radiograph in a patient with acromegaly. Note the prognathism of the mandible and the increased and interdental spacing between teeth. (*From* Farishta F, Hadi MS. Acromegaly: a case study. J Diabetes Metab 2015;6:621; with permission.)