RADIOLOGICAL ASSESSMENT OF ACROMEGALY

Thesis

Submitted in partial fulfilment
of Master Degree in Radiodiagnosis

BY
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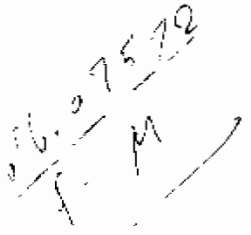

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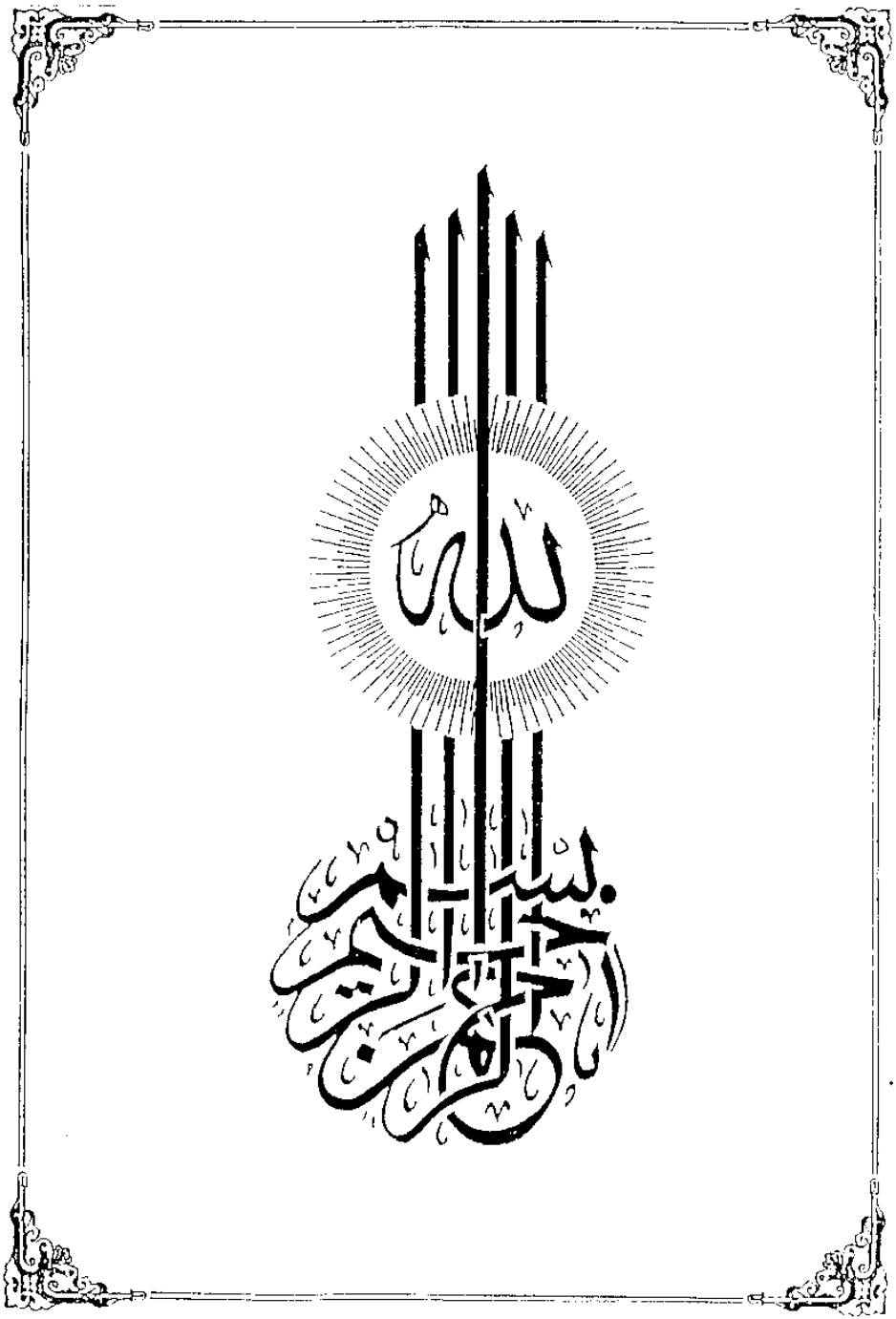

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CONTENTS

Introduction and Aim of Work

INTRODUCTION

Hyperactivity of the anterior lobe of the hypophysis results in gigantism when the condition affects the adolescent and in acromegaly when it occurs in the adult. In 1896, Marie described a condition marked by obesity, coarsening of the features, broadening of the nose, thickening of the lips, prognathism, and increased size of the hands, feet and head which he designated as acromegaly. In the following year, Minkowski associated the disease described by Marie with a tumor of the pituitary. Five years later, Massolongo indicated the relation between acromegaly and gigantism and Benda 1900 showed that these conditions were associated with the eosinophilic cells of the pituitary.

AIM OF WORK

This piece of work aims to describe the relevant radiographic changes which occur in acromegaly and their value in the diagnosis of the condition. Besides a brief study of the radiolgoical means of assessment of pituitary adenomas and their relative merits in this respect are presented.

Pathology

PATHOLOGY

Definition:

Acromegaly is a chronic disease of middle life characterized by overgrowth of bone, connective tissue and viscera in response to prolonged and excessive secretion of adenohypophyseal growth homone. The term acromegaly denotes the typical enlargement of acral or distal parts of the body, the hands, feet, face and head (Wyngaarden and Smith 1980).

Incidence and Prevalence:

Acromgaly is a rare disorder with about 300 new cases per year being discovered in the United States. The sexes are affected equally. Acromegaly starts most commonly in the third and fourth decades . A few patients trace the start of acromegaly to adolescence. Some 30 acromegalics children have been reported. (Wyngaarden and smith 1980).

Causes:

Growth hormone hypersecretion is usually associated with a pituitary tumor which contains eosinophilic staining granules in about 80 per cent of patients and chromophobic granules in the remainder, using electron microscopy together with special cytochemical and immunochemical staining techniques. However almost all the chromophobe tumors associated

with acromegaly can be demonstrated to have GH-containing secretory granules. The presence or absence of abundant GH secretory granules in tumor cells does not correlate with plasma GH levels but is merely a reflection of the capacity of hormone storage as contrasted to synthesis and release. The absence of storage granules does, however, indicate a relatively less differentiated tumor, which in turn is reflected in its growth rate. Thus eosinophilic adenomas tend to be smaller, grow more slowely and permit signs and symptoms of GH hypersecretion to develop for a prolonged period, whereas chromophobe tumors frequently exhibit a more rapid growth rate and produce symptoms of an expanding tumor mass, with those of GH hypersecretion being less pronounced (. (Felig 1981).

Although cases of acromegaly have been reported showing only hyperplasia of the gland involving chiefly the acidophilic elements, by far the most common finding in this condition is an acidophil adenoma.

The size of the tumor varies considerably; in some instances it is small and embeded within a normal or small anterior lobe. Occasionally, a small asymptomatic adenoma may be found, on routine examination, without any features of acromegaly being noted. Presumably such a tumor is not putting out excessive quantities of GH.

Usually, the tumor in acromegaly is large enough to erode and enlarge the walls of the sella turcica on X-ray examination of the skull. Extension upward into the cranial cavity and downward into the sphenoidal sinuses is common. Lateral extension of the tumor through the wall of the cavernous sinus occasionally occurs, producing a syndrome of paralysis of one or more of the third, fourth, fifth and sixth cranial nerves that traverse the sinus. Lateral spread may extend to involve the temporal lobe. Extension upward to the diaphragma sellae usually affects the optic chiasm. Occasionally, the position of the chiasm may be sufficiently anterior or posterior to the diaphragma so that the optic pathway may not be compressed by the growing tumor. Forward progression sometimes involves the frontal lobe. Upward extension may fill the third ventriclewith tumor and may obstruct the foramen of Monro on one or both sides as well as the aqueduct of sylvius, leading to marked dilatation of the ventricle (Netter 1965).

There is considerable controversy as to whether acromegaly is a primary pituitary disease due to autonomous tumor formation or is of hypothalamic (or other CNS) origin, occurring as a consequence of excessive secretion of GH. releasing factor or possibly decreased secretion of somatostatin.

The evidence for a hypothalamic etiology follows:

(1) GH secretion in most acromegalics is not autonomous but reponds to stimuli mediated through the hypothalamous such as glucose, Insulin hypoglycemia and arginine. This implies that the somatotrophs even though neoplastic are capable of responding to hypothalamic signals.

(2) Hypothalamic tumors have been associated with GH hypersecretion and acromegaly, suggesting that over-production of a GH-relasing factor may have been involved (Fehg 1981).

(3) Plasma from patients with acromegaly has been reported to contain GH-releasing activity when tested in vitro.

(4) Acromegaly caused by GH-secreting pituitary tumors is reversible following removal of bronchial carcinoid or islet cell tumors which contain a GH-relasing factor. Thus, GH hypersecretion and even pituitary tumors can develop in response to prolonged GH-relasing factor stimulation.

(5) In addition, GH responses to glucose suppression and insulin stimulation, when examined following removal of the GH-secreting adenomas, generally remain abnormal even when basal levels of GH are normal.

(6) Finally, GH secretion in acromegaly is altered by neuro-pharmacologic agents (alpha-adrenergic antagonists and beta-adrenergic agonist stimulate GH-secretion) which are believed to act within the central nervous system (CNS). These arguments suggest a hypothalamic etiology for GH hypersecretion but do not preclude the eventual development of an autonomous pitiutary tumor ((Felig 1981).

Acromegaly associated with bronchial carcinoid or pancreatic islet cell tumor has been described in small number of patients. In at least five of these patients GH secretion was restored to normal or the clinical signs and symptoms of acromegaly subsided after the extrapituitary tumor was removed without any therapy being directed towards the pitiutary gland.

Extracts from such an adenoma were found to contain a potent substance capable of releasing GH from dispersed pituitary cells in culture.

This GH relasing hormone (GHRH) has been shown to be a protein with a molecular weight of 6000 Elaboration of GHRH by a peripheral tumor may therefore have the potential of leading to a pitiutary tumor. The presence of

an extrapitiutary tumor should be excluded in any patient with acromegaly. (Wyngaarden and smith 1980).

The excessive secretion of growth hormone affects virtually all organs and tissues. The response of cartilage and bone is obvious and distinctive. The histologic appearance of chondrogenesis and osteogenesis tends to be disorderly.

The cartilaginous and bony response occurs at points of special sensitivity, e.g mandible zygoma, ribs and clavicle. The uneven growth is thought to be due to the effect of pressure or of muscular traction. Increased chondrogenesis is most obvious in hypertrophy of the costal cartilages, which contributes to the increased circumference of the thorax. Acromegalic arthritis affecting chiefly the large joints and the spine resembles osteoarthritis and results from proliferation of deep layers of joint cartilages with thinning of articular cartilages. Hypertrophy of nasal and aural cartilage accounts for part of the enlargement of nose and ears. Osteogensis from the thickened periosteum is accelerated, absorption of bone is also rapid, late in the disease osteoporosis may be striking. There is overgrowth of the supraorbital ridge The anterior temporal ridge advances, with forward growth of the lateral portions of the orbit, expansion of the frontal sinuses, and forward displacement of the zygoma. Over growth