

ACROMEGALY: A CASE REPORT WITH MRI FINDINGS

Akromegali: MRG Bulguları ile Birlikte Bir Olgu Sunumu

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ABSTRACT

ÖZ

Acromegaly is a rare metabolic disorder that develops with over secretion of growth hormone (GH) after the epiphyseal plates are closed. Acromegaly starts insidiously and progresses very slowly. Signs and symptoms of growth hormone elevation may not be noticed for years. Jaw enlargement, swelling of the hands and feet, prominent facial bones, facial roughness, nasal enlargement, tongue enlargement, deepening of the voice, snoring and sleep apnea, skin thickening, excessive and malodorous sweating are some of the clinical changes. Changes in acromegaly such as macroglossia, diastemas, class III malocclusion, condylar growth, mandibular enlargement and thickening of facial tissues are familiar for dentists. Dental clinicians can play an important role in the diagnosis of acromegaly or other systemic diseases with careful examination and history.

Akromegali, epifiz plaklarının kapanmasından sonra aşırı büyüme hormonu (GH) salgılanmasıyla gelişen nadir bir metabolik hastalıktır. Akromegali sessizce başlar ve çok yavaş ilerler. Büyüme hormonu artışı belirtileri ve semptomları yıllarca fark edilmeyebilir. Çene büyümesi, el ve ayaklarda şişlik, yüz kemiklerinde belirginlik, yüzde kabalık, burun büyümesi, dil büyümesi, sesin kalınlaşması, horlama ve uyku apnesi, cilt kalınlaşması, aşırı ve kötü kokulu terleme klinik değişikliklerden bazılarıdır. Makroglossi, diastemalar, sınıf III maloklüzyon, kondiler büyüme, mandibular genişleme ve fasiyal dokuların kalınlaşması gibi değişiklikler diş hekimliği ile yakından ilişkilidir. Diş klinisyenleri, dikkatli muayene ve anamnez ile akromegali veya diğer sistemik hastalıkların tanısında önemli bir rol oynayabilir.

Keywords: *Acromegaly, prognathism, growth hormone, pituitary adenoma*

Anahtar Kelimeler: *Akromegali, prognatizm, büyüme hormonu, hipofiz adenomu*



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INTRODUCTION

Acromegaly is a chronic endocrine disease that effects skeletal tissue and ends up in organ enlargement because of excessive GH secretion from somatotropic cells in pituitary tumors (1). If the growth hormone is over-secreted before epiphyseal plaques are closed, the disease is named gigantism. If it is over-secreted after epiphyseal plaques are closed, it is named acromegaly (2).

The disease was first described by Andrea Vega in 1864. The clinical features of acromegaly were revealed by Pierre Marie in 1886 (3). Acromegaly is a rare disease and its annual incidence is 3-4 new patients in a million. Males and females are equally affected. Acromegaly is diagnosed on the 4th decade of life which is about 4-10 years after the onset of the disease (4). This report emphasizes the role of dental clinicians in diagnosing of acromegaly.

CASE REPORT

A 35-year-old female patient was admitted to our clinic with the complaint of gingival bleeding and gap between her teeth. There was no history of systemic disease. The clinical image of the patient aroused suspicion. Extraoral examination revealed large facial features, thickening of both skin and soft tissues, speech disorders, growth in hands, pronounced supraorbital bulge and mandibular prognathia (Figure 1,2). Intraoral examination revealed macroglossia, diastemas in lower incisors and class III malocclusion (Figure 3). Panoramic radiography showed diastemas between mandibular anterior teeth, increased mandibular planar angle and bilateral mandibular enlargement (Figure 4). On lateral sefalogram, pneumatization of maxillary and frontal sinuses, enlargement of sella turcica attracted our attention (Figure 5).

Acromegaly was initially diagnosed as a result of all clinical and radiological findings and biochemical tests were requested. Growth hormone with OGTT, IGF-1 and prolactin levels were measured. In the glucose tolerance test, GH level was insufficiently suppressed whereas IGF-1 was significantly high (Table 1). The prolactin level was normal. Acromegaly was diagnosed as a result of all clinical, radiological and biochemical findings and the patient was referred to endocrinology department where the patient was diagnosed as pituitary adenoma (Figure 6).



Figure 1: Growth in hands.



Figure 2: Large facial features, skin and soft tissue thickening



Figure 3: Malocclusion, macroglossia, diastemas (in order)

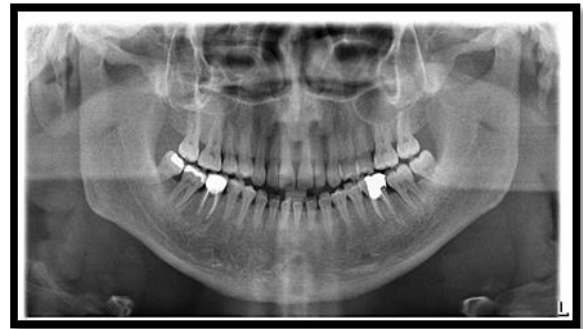


Figure 4: Panoramic radiography



Figure 5: Lateral cephalogram shows enlargement of sella turcica

Table 1: IGF-1 and GH values of patient.

Test name	Result	Unit
OGTT 75 gr. GH 30 minute	7.35	ng/mL
OGTT 75 gr. GH 60 minute	5.48	ng/mL
OGTT 75 gr. GH 90 minute	5.43	ng/mL
OGTT 75 gr. GH 120 minute	5.34	ng/mL
Somatomedin-C (IGF-1)	743	ng/mL

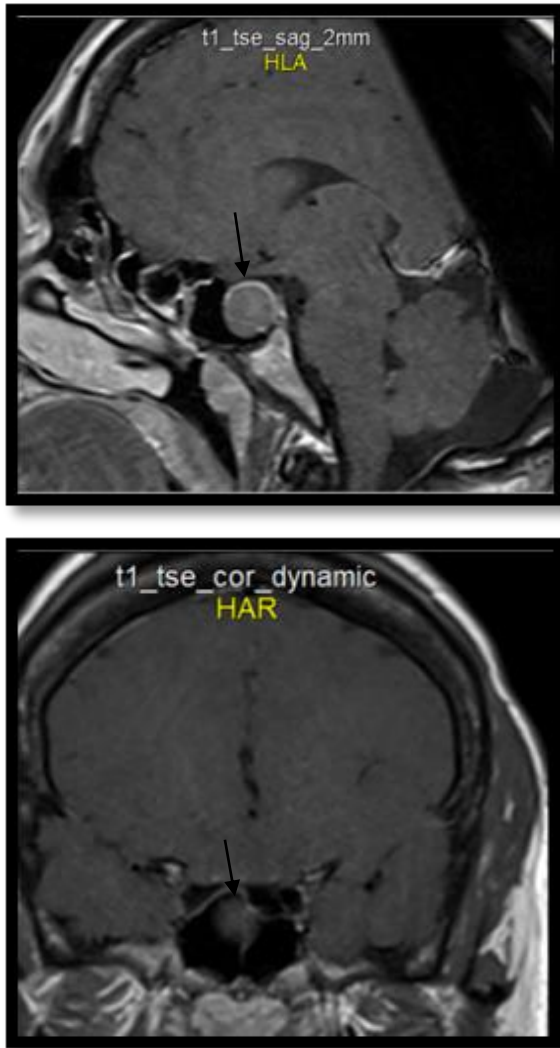


Figure 6: T1-weighted MR images showing pituitary adenoma (sagittal and coronal section)

DISCUSSION

Acromegaly diagnosis should be made with clinical and biochemical results. If the diagnosis is not clear, the measurement of insulin like growth factor -1 (IGF-1) level should be the first step. The level of IGF-1 is assessed according to age and sex. If the IGF-1 level is high, the GH value should be checked for a definitive diagnosis. Measurement of GH value during oral glucose tolerance test (OGTT) in biochemical analysis is accepted as the gold standard. GH levels drop during OGTT in a healthy person whereas in acromegaly patients the lowest GH value after OGTT

is higher than 1 ng / ml. Ninety percent of acromegaly patients have a value above 10 ng / ml (5).

Systemic symptoms of acromegaly are very diverse. The most common symptoms are diabetes mellitus, hypertension, heart and respiratory failure. Cardiovascular diseases are the first cause of death for acromegaly patients, followed by respiratory diseases and neoplasms. If the excessive secretion of growth hormone is controlled, the mortality rate of the patients decreases (6).

The most common clinical manifestations are face, hand and foot enlargement. The most prominent features of the face are the deep nasolabial sulcus, pronounced supraorbital bulge, enlarged lips and nose. Other features are macroglossi, swelling of nasopharyngeal tissues, arthralgia, sleep apnea, lethargy, menstrual irregularities and infertility (2). Mandibular growth can cause prognathism, malocclusion and diastemas (1,6). In our case, there was growth in the hands, macroglossia, significant supraorbital bulge, diastema, mandibular prognathism, enlargement in nose and lips. The increase in ring and shoe numbers may also be important in diagnosis of acromegaly (7).

Radiological examination reveals enlargement of sella turcica, prolonged ramus, enlarged paranasal sinuses, hypersementosis in posterior teeth and prognathism in patients (8,9). Our patient had all radiological findings except hypersementosis.

Treatment options include surgery, medical therapy, radiotherapy, and combinations thereof. Transsphenoidal surgery is accepted as the first treatment option. In medical therapy three kinds of agents are used: dopamine agonists, GH receptor antagonists, somatostatin analogs (10,11). Transsphenoidal surgery and medical treatment was planned by a neurosurgeon for our patient. To conclude, macroglossia, progressive dental malocclusion, diastemas, enlargement of facial features can be associated with acromegaly. Dentists

should have adequate knowledge of the systemic diseases and should review all patients with careful clinical and radiological examination.

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