International Journal of Medical Research Professionals
P-ISSN: 2454-6356; E-ISSN: 2454-6364
DOI: 10.21276/ijmrp

IJMRP

Case Report.

A Rare Case of Acromegaly Among Yamani Male

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ABSTRACT

Acromegaly is a rare disease caused by an over growth hormone secretion, mostly as a result of pituitary adenoma. A 30 year old Yamni male presented with increased stature growth and excessive enlargement of feet and hands since childhood with prominent supraorbital ridges. Examination revealed that there were gross acromegalic features such as enlarged body parts, prominent supraorbital ridges, coarsening of facial feature along with frontal bossing, prognathism and macroglossia, separated teeth and deep voice. MRI of the sella with IV contrast using different pulse sequence in different planes showed enlarged pituitary gland. Post-surgical MRI report showed a known case of large pituitary macroadenoma and clinically acromegaly with complete transphenoidal resection. Post-operative laboratory investigation revealed that growth hormone was 2.3 ng/ml. Conclusively, early recognition and treatment of disease helped in better prognosis of disease and avoiding serious the complications.

Keywords: Acromegaly, Growth Hormone, Pituitary Adenoma.

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Article History:

Received: 02-01-2017, **Revised:** 06-01-2017, **Accepted:** 12-01-2017

Access this article online	
Website: www.ijmrp.com	Quick Response code
DOI: 10.21276/ijmrp.2017.3.1.040	

INTRODUCTION

Acromegaly is disease caused by an over growth hormone secretion, mostly as a result of pituitary adenoma. Pituitary adenomas are never became malignant, however they are accompanied by high morbidity and mortality rates.[1,2]

Acromegaly is rarely presented clinically as it occurs in a prevalence of 50-70 cases/million and in an incidence of 3/1000,000 cases/year.[3]

Acromegaly is manifested clinically in many body systems including visceromegaly (goiter, macroglossia, hepatomegaly and splenomegaly), cardiovasular system (ventricular hypertrophy, cardiomyopathy, hypertension and congestive heart failure), gastro-intestinal system (Colon polyp), respiratory system (sleep apnoea and upper airway obstruction as a result of macroglossia), metabolic disorders (diabetes mellitus, impaired glucose tolerance), muscluloskeletal system (prognathism, acral enlargment, frontal bossing arthralgia, myopathies), skin (acnthosis nigricans and oily skin), neurological disorders (Carpal Tunnel Syndrome, headache, aneurysm, and local effect due to tumous.[4]

Diagnosis of acromegaly is based mainly on clinical presentation as well as biochemical investigation and radiography. The biochemical diagnosis of acromegaly is achieved by increasing in the level of growth hormone and insulin like growth factor. Normal growth hormone level excludes the diagnosis of acromegaly in most of the cases. Visual field assessment is necessary in cases of macroadenoma which are close to optic chiasm. Additionally, x-ray of limbs and skull and chest are needed to confirm the diagnosis. Colonoscopy is needed to look for polyp. Echocardiography is also indicated if patient has clinically cardiac involvement.[5]

Transsphenoidal surgery is the treatment of choice in case of acromegaly due to pitutary adenoma. Also, medical treatment can be used including somatostatin analogs, dopamine agonist and GH receptor blocking agents. Surgery is superior to medical treatment as it gives rapid relief of symptoms and signs and stops the organ damage due to hormone excess.[2]

CASE REPORT

A 30 year old Yamani male presented with increased stature growth and excessive enlargement of feet and hands since childhood with prominent supraorbital ridges, discovered accidently by his father's doctor. Additionally, he had frontal headache since long time. There were no voice changes, joint pain, muscular pain, excessive sweating, visual disturbance, erectile dysfunction, numbness or tingling sensation. No history of night sweating, fever or fatigue, weight loss or loss of appetite.

Medical history was not significant. Family history was contributory for similar episode in his cousins. No history of blood transfusion, recent traveling, he doesn't take any medication and no history of allergy.

Examination revealed that patient was fully conscious and oriented without any neurological deficits. Blood pressure was

normal. Height 180 cm, weight 115 kg, body mass index (BMI) was 35 49 kg/m^2. There were gross acromegalic features such as enlarged body parts, prominent supraorbital ridges, coarsening of facial feature along with frontal bossing, prognathism and macroglossia, separated teeth and deep voice. There was no pallor, cyanosis, clubbing or lymphadenopathy.

Cardiac examination revealed normal first heart sound and second heart sound with no added sound or murmur. Chest examination showed bilateral vasicular breathing with no added sound while abdominal examination showed soft and lax with audible bowel sound with no organomegaly. Motor and sensory neurological systems were intact.

Laboratory investigations showed that complete blood count, liver function test, renal function test and chemistry were within normal levels. Thyroid-stimulating hormone was 1.05 uiU/ml, Adrenocorticotropic hormone (pm) was 18.5 pg/ml, Adrenocorticotropic hormone (am) was 36.0 pg/ml, growth hormone ≥40 ng/ml, follicle-stimulating hormone was 2.7 mIU/ml, Luteinizing hormone was 1.4 mIU/ml and prolactin was 26.5 ng/ml.

Skull scanoscopy demonstrated marked enlargement of sella turcica (ballooned sella) and erosion of the dorsum. Coronal and axial CT slices showed intra and parasellar mass measures 1.7X2.2 cm, resulted in marked erosion of the dorsum of sella as well as pituitary fossa enlargement. Sphenoid sinus in deformed. CT findings suggested of macroadenoma of the pituitary gland.

MRI of the sella with IV contrast using different pulse sequence in different planes showed enlarged pituitary gland with a well-defined lobulated mass lesion having homogenous signal intensity causing bulge to the supra sellar cistren and laterally to the cavernous sinus. Overall dimension of the lesion measured about 3x3x2 cm in cranio-caudal, transverse and anterio-posterior diameters. The carotid portion in the cavernous sinus appears patent and present normal signal void. Sagging of the sellar floor with no evidence of intra sphenoid extension. The mass present is intense in T1 and T2 and present homogenous post contrast enhancement. No evidence of supra sellar hydrocephalic changes. Normal size of ventricles with no midline shift. Normal cervico-medullary junction.

Patient admitted to the hospital on 25/ 3/ 2005 as a case of large pituitary macroadenoma and clinicaly acromegaly, after lab investigation, imaging (CT and MRI) undergo complete transphenoidal resection. Post-surgical MRI report revealed a wide empty sella (post-surgical) with no signs of rest tumor or recurrence and discharge home on 29/3/2005.

Pathological report showed tiny pieces of grayish soft tissue, collectively measuring 0.4 X 0.3 cm. Microscopical examination revealed pieces of tumour tissue formed of epithelial cells with regular rounded nuclei, chromophobic cytoplasm arranged in sinusoidal and trabecular pattern. The tumour is moderately vascular. No evidence of malignancy. Post-surgical MRI report showed a known case of large pituitary macroadenoma and clinically acromegaly with complete transphenoidal resection. The current control e examination revealed wide empty sella (post-surgical) with no sign of rest tumor or recurrence. Post-operative laboratory investigation revealed that growth hormone was 2.3 ng/ml, follicle-stimulating hormone was 3.2 mIU/ml, luteinizing hormone was 0.8 mIU/ml and insulin-like growth factor was 200 ng/ml.

DISCUSSION

The present care was diagnosed at age of 30 years. It has been documented about 8-10 years of delay is commonly observed from the onset of symptoms to diagnosis of cases of acromegaly.[6] In order to have good prognosis of disease, early diagnosis and management is fundamenta.[6]

Acromegaly is seen equally in both genders.[7] The present case is male patient. Main presenting features of the present case were increased stature growth and excessive enlargement of feet and hands since childhood with prominent supraorbital ridges. In other cases, the main features were weight gain, sweating, headache and sometimes joint pain, which had been present for at least 4 years before diagnosis.[8]

The present case in accordance with others presented with multisystem involvement; cardiovascular, endocrinal, musculoskeletal, cutaneous, neurological and psychiatric troubles.[8,9]

In the present case, diagnosis was confirmed by clinical picture, elevatedIGF-1 levels and MRI brain showing pituitary macroadenoma. It has been reported that the key diagnosis of acromegaly is bases on a combination of signs and symptoms of the condition with biochemical investigations.[10] A pituitary MRI should be performed after biochemical testing to confirm the presence of a pituitary macroadenoma. If the pituitary tumor is diagnosed incidentally, and if acromegaly is suspected based on clinical feaure, IGF1 level should be measured.[11]

It has proven that unless GH levels are controlled, survival is reduced by an average of ten years compared with an age-matched control group.[12]

This patient was detected in a relatively early phase without fatal complications. The objective of therapy was to control growth hormone over secretion. Hence, a surgical resection of the GH secreting adenomas was the initial treatment in the present case. Transsphenoidal surgical resection was performed and Growth hormone level retained to its normal level post-operatively.

Conclusively, although acromegaly is a rare disease, it should be in the differential diagnosis of patients with clinical signs and symptoms suggestive of acromegaly, since early recognition and treatment of disease helped in better prognosis of disease and avoiding serious the complications and consequently better quality of life for patients with the disease.

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Source of Support: Nil. **Conflict of Interest:** None Declared.

Cite this article as: Bushra Mohammad Alqurashi, Lama Khalid Bahatheq, Maram Abdullah Almalki. A Rare Case of Acromegaly Among Yamani Male. Int J Med Res Prof. 2017; 3(1):200-02. DOI:10.21276/ijmrp.2017.3.1.040