ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 03 | March - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

ACROMEGALY UNVEILED DURING A NASAL RECONSTRUCTION SURGERY: A CASE REPORT

Endocrinology

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ABSTRACT

Acromegaly is a condition of excessive somatic growth and distorted proportion due to hypersecretion of growth hormone(GH) and insulin-like growth factor 1 (IGF-1). Insidious clinical manifestation of GH excess as a result of GH-secreting pituitary adenoma renders acromegaly a disease with typically delayed diagnosis. We report a 29-year old male planned to undergo reconstruction of the nose. The patient was referred for medical fitness. Given the clinical features consistent with acromegaly the patient was evaluated further for diagnosis. Laboratory investigation showed raised IGF-1and non-suppressed GH post 75gms glucose, amid normal cortisol, corticotropin(ACTH), prolactin, testosterone, and thyroid function tests. He did not have dysglycemia. Magnetic resonance imaging (MRI) of the pituitary revealed a pituitary macroadenoma consistent with acromegaly. This case highlights the notable absence of recognizing the clinical presentation of acromegaly in this patient by his medical care physician and surgeons, and therefore the importance of thorough history taking, attention, and observation in making a new diagnosis that has the potential to alter a patient's health care and alleviate impending complications, morbidity and/or mortality.

KEYWORDS

Acromegaly, IGF-1, Growth hormone , pituitary macroadenoma

INTRODUCTION

Acromegaly was described more than 120 years ago[1] and later ascribed to pituitary secretion and adenomas[2]. It denotes enlargement of soft-tissue and osseous tissue in acral areas. It is a rare condition, the clinical profiles associated at times being non-specific and the manifestations of the disorder are insidious, several patients go undiagnosed for several years after the onset of initial signs and symptoms. Timely screening, diagnosis, and prompt treatment are vital as it carries up to a 72% increase in all-cause mortality when compared to the general population[3]. Changes in appearance, subtle, bring only 13% of patients who have acromegaly to seek medical attention[4,5], despite these changes account for 98% of presenting features[6]. In this paper, we report a case of acromegaly unveiled in a patient planned to undergo reconstruction of the nose by the team of otorhinolaryngologist and plastic surgeons.

Case presentation

A 29-year-old-male, presented with enlargement of the nose for 3 years. The patient was apparently alright 3 years back when he noticed enlargement of the nose which has progressed gradually over the years to the current state. The enlargement of the nose was associated with hyperpigmentation of the face, however, the patient did not seek any medical attention for the same. At present, the patient complains of difficulty in breathing through the nose or nasal congestion, pain, tenderness, and swelling around the eyes, cheeks, nose, and forehead. This was accompanied by at times by yellowish-green nasal discharge at times for which he took antibiotics from the family physician. There was no history of headache, postnasal drainage, reduced sense of smell and taste. No history of snoring. Computed tomography paranasal sinuses revealed few polyp/retention cyst noted in bilateral maxillary and right frontal sinuses and right ethmoid air cells, measuring up to 1.0 x 1.7 cm in the left maxillary sinus. The nasal septum is deviated to the right with mild mucosal hypertrophy in the left inferior turbinate. There was a history of acral enlargement for the past 8 years, however, went unnoticed. There was no history of visual disturbances. No complaints of joint pains and backache. A progressive deepening of voice was observed. On examination, his height was 176cm and weight 80kg with a body mass index of 25.8kg/m^2. The blood pressure was 130/80mm of Hg. He had a florid presentation of acromegaly and had no goiter (Table 1).

Table 1 Florid Features of patient suggestive of acromegaly

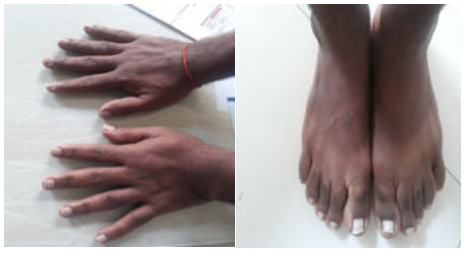
Frontal Bossing
Prognathism
Thick fleshy lips
Macroglossia
Hyperpigmentation
Widened and enlarged nose
Enlarged hands and feet
Punchinello rib cage

Skin thickening was noticed mainly on the face , hands and feet (Figure1a, b respectively).

Figure 1 Facial Photographs

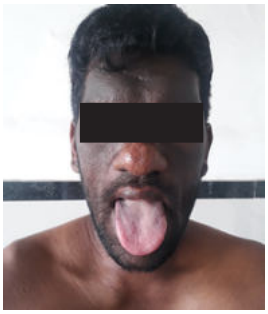
a) Frontal **Lateral**

b) Hands and Feet



The patient had macroglossia (Figure 2)

Figure 2 Macroglossia



There were no skin tags. His hands were warm and moist. There was diffuse hyperpigmentation. He had hyperhidrosis and seborrhea.

There was deformation of rib cage, leading to the classical punchinello aspect(Figure 3)

Figure 3 Punchinello rib cage

Serum blood glucose, electrolytes, renal and liver function tests were normal. He has mild hyperphosphatemia, though his calcium profile was normal. Hormonal workup showed normal thyroid function tests, 0800-hour cortisol, ACTH, prolactin, and testosterone. His insulin-like growth factor 1 (IGF 1) was elevated and growth hormone (GH) was not suppressed following the glucose tolerance test (Table 2).

Table 2 Serum hormonal profile

Hormone	Observed value	Normal Range
0800 h Cortisol	9.46	3-21 g/dl
ACTH	51.4	7.2-63.8 pg/mL
Prolactin	6.96	4.04-15.2 ng/mL
Free T4	0.986	0.93-1.7 ng/dl
TSH	1.03	0.54-4.5IU/mL
Total Testosterone	439	249-836 ng/dL
IGF 1	749	88-537 ng/mL
GH at 0 min	50	0-3 ng/mL
GH at 60 min	39.7	<1 ng/mL
GH at 120 min	37.6	<1 ng/mL

Perimetry showed no visual field involvement. 2-D echocardiography was normal with a left ventricular ejection fraction of 60%.

X-ray skull showed calvarial thickening with frontal bossing and enlarged frontal sinus,

The pituitary fossa appeared voluminous. There is prognathism of mandible(Figure 4).

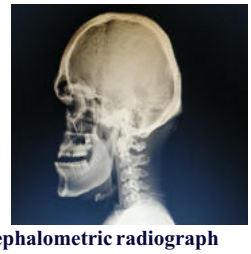
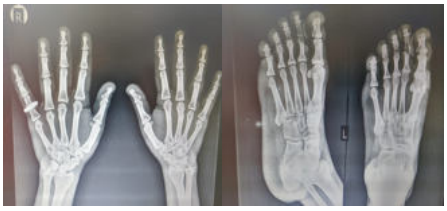


Figure 4 Lateral cephalometric radiograph

X-ray hands and feet showed terminal phalanges tufts having a " spade appearance" -the spade phalanx sign likely suggestive of acromegaly(Figure 5).

Figure 5 X-ray of hands and feet



MR imaging revealed 18x20x11mm well defined intensely enhancing sellar mass lesion with local extension to suprasellar cistern without reaching the optic chiasma, suggestive of a pituitary macroadenoma (Figure 6).

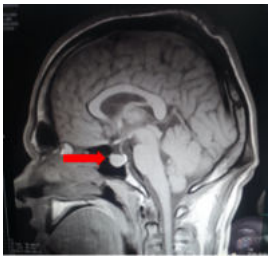


Figure 6 MRI Brain (Plain and Contrast)

Our patient underwent a uneventful trans-sphenoidal pituitary surgery. He did not have cerebrospinal fluid rhinorrhea, but had polyuria which resolved after 3 days. Serum IGF-1 and pituitary dedicated MRI will be repeated three months later.

DISCUSSION:

Acromegaly is an uncommon disease caused by GH hypersecretion, mostly from a pituitary adenoma, driving IGF-1 overproduction[7]. The diagnosis is delayed owing to the insidious onset of clinical manifestations. The main clinical features are broadened extremities (hands and feet), widened thickened and stocky fingers, and thickened soft tissue. The facial features attribute and included a wide and thickened nose, notable cheekbones, forehead bulging, thick lips, and pronounced facial lines. The forehead and the overlying skin is thickened, sometimes resulting in frontal bossing. There is a proneness towards mandibular overgrowth with prognathism, maxillary widening, tooth separation, and jaw malocclusion[8].

Acromegaly is associated with increased mortality. According to series published in the 1980–the 1990s, about 60% of patients die from cardiovascular disease, 25% from respiratory complications, and 15% from cancer. If left untreated, patients with acromegaly would die about 10 years earlier than healthy subjects[9].

In our patient, the diagnosis of acromegaly was reviewed due to the classical features and manifestations at first presentation. Widened and thick nose with recurrent nasal congestion was the main reason for the patient to seek an otorhinolaryngologist. Enlargement of the lips and the tongue, frontal bossing, prognathism, macroglossia and increased size of hands and feet were present in our patient. All these changes have often been described in the literature as cosmetic changes associated with GH adenomas[10]. The classical Punchinello deformation of the rib cage was observed in our patient[7]. Diffuse hyperpigmentation in a patient with acromegaly can occur due to direct effect of GH melanocytes as observed in our patient. The optic chiasma was not involved, as in our patient, there weren't any vision related complaints, and confirmed by assessment of visual field and acuity. His lateral cephalometric radiograph displayed enlargement of the sella turcica caused by the adenoma expansion of the pituitary gland a noticeable indication that is detectable in almost every patient with acromegaly[11]. Measurement of GH response to a glucose load is the gold standard diagnostic test. An increase in the serum concentration of IGF-I, the main GH dependent growth factor, confirms this diagnosis1[2-13]. MRI has emerged as the imaging modality of choice for evaluating pituitary glands[14]. MR imaging in our patient revealed 18x20x11mm well defined intensely enhancing sellar mass lesion with local extension to supra-sellar cistern without reaching the optic chiasma, suggestive of a pituitary macroadenoma. The trans-sphenoidal adenectomy approach is considered as the first-line treatment of acromegaly[15]. Though safe removal of all tumor tissue can be difficult., depending on the tumor size, long-term remission is

achieved in 50% to 67% of patients, but microadenomas are more responsive to cure[9,16]. Preoperative GH concentration seems to be the best measure for successful treatment and a perpetual decline of GH and IGF-I[17].

CONCLUSION

Although acromegaly is a rare disease, its clinical presentation is salient and can barely be misconstrued. This case highlights the need to watchful, as diagnosis and treatment will ward off the patient from imminent complications due to morbidity and or mortality.

Ethics Approval: Not applicable
Consent Publication: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of interests The authors declare that they have no competing interests.

Funding: None
Author Contributions: Author and co-authors have equally contributed to the manuscript.

Acknowledgements: None

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