

70 Years of Undiagnosed Acromegaly & Treatment with Cabergoline

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Abstract: *Acromegaly is a rare disorder with an incidence of 3-4 cases per million per year worldwide. It is caused by excessive growth hormone (GH) secretion, usually resulting from a benign GH-secreting pituitary adenoma. The effects of GH hypersecretion progress slowly so that the course of the disease is usually insidious and the changes may not be noticed for years. Ultimately, if left untreated, can lead to bony disfigurement and serious life threatening complications. We present this case to show the uncommon but distinct manifestations which have been missed clinically for years by other specialities leading to massive disfigurement and complications*

Keywords: Acromegaly, Dermatology, Cabergoline, Growth Hormone

1. Introduction

Acromegaly is a rare disorder with an incidence of 3-4 cases per million per year worldwide. It is caused by excessive growth hormone (GH) secretion, usually resulting from a benign GH-secreting pituitary adenoma. The effects of GH hyper-secretion progress slowly so that the course of the disease is usually insidious and the changes may not be noticed for years. Ultimately, if left untreated, can lead to bony disfigurement and serious life threatening complications. We present this case to show the uncommon but distinct manifestations which have been missed clinically for years by other specialities leading to massive disfigurement and complications.

2. Case Report

We report a case of a 72 year old man with an undiagnosed disorder since past 30 years with coarse facial features, deep seated furrows and supraorbital ridging over the forehead (frontal bossing), thickened skin over the eyelids causing mechanical ptosis, rhinophymatous nose, cutis vertis gyrata over the scalp, macroglossia, enlarged hands and feet, multiple joint pains and multiple pedunculate lesions over the face, neck and back.

On the basis of his coarse facial features, differential diagnosis of Patchydermoperiostosis and Acromegaly were given.

Ophthalmic examination revealed mechanical ptosis along with increased intraocular pressures leading to glaucoma and optic atrophy. Patient had a past history of trabeculectomy of the right eye which led to a cystic bleb formation adherent to cornea

On further investigations The serum levels of Growth hormone and Insulin like growth factor 1 (IGF-1) were raised. Growth hormone was 6.38ng/mL (n= 0-3) and IGF-1 was 1036 ng/mL (n=59-177). Blood sugars, Thyroid profile, Liver and Renal function tests were normal. Patient's blood pressure was 150/90 mmHg.

The X-ray scan of the skull showed enlarged pituitary fossa (sella turtica) with thickening of skull borders and a prominent jaw and a protruding mandible. X-ray of the feet showed enlarged heel pad thickness (30mm) with mild tufting of the terminal phalanges. Chest X-ray showed cardiomegaly. ECG was within normal limits. MRI brain could not be performed because of patient's THR prosthesis (old metals were used). Thus to rule out prolactinoma, prolactin levels were tested which came out within normal limits. Later CT Brain confirmed a 1cm pituitary macroadenoma.

Pachydermoperiostosis was ruled out with no features of clubbing and periostosis in radiological films. Insulin dependent pseudoacromegaly was ruled out by normal fasting and post prandial blood sugars.

Skin biopsy of his forehead folds showed focal hyperkeratosis and parakeratosis with the dermis showing increase in dermal collagen

The patient was diagnosed as acromegaly. Our patient had uncontrolled hormonal levels and was further evaluated and managed by an Endocrinologist. Patient was started on T. Cabergoline (0.5mg) (dopamine agonist) twice weekly with diet precautions since patient could not afford Somatostatin therapy. After 4 months of the therapy his growth hormone levels significantly came down to 5.12ng/ml and his IGF levels came down to 936ng/ml without any complications.

3. Photos



Figure 1: Showing coarse facial features, Frontal bossing, enlarged nose & macroglossia

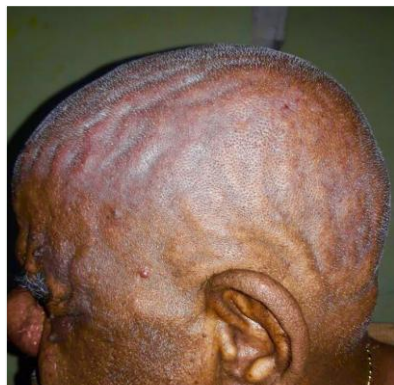


Figure 2: Cutis Verticis gyrata



Figure 3: X-Ray skull showing prominent thickening of skull borders, enlarged sella turcica and mandibular prognathism

4. Discussion

Acromegaly is not a common disease. The term "acromegaly" was coined by Pierre Marie in 1886, [1] though he was not the first person to describe this clinical entity and in fact he had refuted the idea of pituitary hyperfunction. Minkowski in 1887, when describing a patient with headache and hemianopia, was the first to suggest that pituitary dysfunction caused acromegaly.[1]

Excessive secretion of growth hormone starting after or around the time of closure of epiphyses of the long bones results in acromegaly. Gigantism results when the excessive secretion of growth hormone begins before the epiphyses close. However, patients with gigantism almost always develop the clinical features of acromegaly because overproduction of growth hormone continues long after closure of the epiphyses of the long bones. The lesions responsible

for the excessive production may be hypothalamic, hypophyseal or ectopic in origin producing growth hormone or growth hormone releasing factor.[1]

Growth hormone exerts its effects directly on all or almost all tissues of the body ⁽⁴⁾

It has been postulated that post pubertal overproduction of GH leads to highly disproportionate growth of the jaws and facial bones, This result in enlargement of the ascending ramus and prominence of the mandible compared to maxilla, which is a quiet noticeable characteristic of an acromegalic patient ⁽⁹⁾. As a sequelae to this mandibular growth, macroglossia occurs. Along with this, the soft palate also shows enlargement which can lead to sleep apnea.

Radiographic features are striking, with the enlargement of sella turcica being the important feature in acromegaly patient. The other features are, prominent supraorbital ridges, hypercementosis, large pulp chambers, and increase in size of mandible, interdental spacing, hand and wrist, shows tufting of fingers. Patients with acromegaly usually exhibit enlargement of all parts of the neuro cranium and orofacial bones except the maxilla ⁽⁵⁾

Medical therapy is appropriate for patients not achieving a surgical cure and for those for whom a surgical approach is not an option or who prefer medical therapy over surgery.⁽⁶⁾⁽⁷⁾

Medical therapies for management of acromegaly include dopaminergic agonists, somatostatin analogues, and growth hormone receptor antagonists. Surgical therapy includes transsphenoidal approach and transnasal endoscopic approach frontotemporalcraniotomy. ⁽¹⁰⁾ Radiotherapy includes gamma knife. Remission of the tumour after a period of time is to be evaluated which can be an important factor in giving a prognosis to the treatment. In 2 major studies, it was concluded that cabergoline adjuvant normalizes IGF-I in about 50% of cases⁽²⁾⁽³⁾ which promoted us to safely use the drug and the results were significant.

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Author Profile



Dr Isha Singh received her M.B.B.S degree from Maharashtra University of health sciences and is currently pursuing her M.D degree in Dermatology from MGM university. Currently posted as the chief resident and working in MGM hospital in Navi Mumbai.