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Case Study

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ACROMEGALY - A CASE REPORT

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ABSTRACT

GH hypersecretion is usually the result of a somatotrope adenoma but may rarely be caused by extrapitutary lesions. Protean manifestations of GH and IGF-1 hypersecretion are indolent and often are not clinically diagnosed for 10 years or more. This report details the case of a patient with an undiagnosed case of acromegaly in a 67-year-old woman, K/C/O Systemic hypertension who had complained of lower back pain, neck pain and bilateral knee pain over the course of several months. This patient was referred for a Magnetic Resonance Imaging (MRI) of brain which revealed changes suggestive of pituitary adenoma. The patient was referred to neurosurgery for endoscopic excision of adenoma.

KEYWORDS: Acromegaly, Pitutary Adenoma, Endoscopic Excision.

INTRODUCTION

- The pituitary gland integrates hormonal signals that control adrenal, thyroid, reproductive, growth, and metabolic functions.
- Distinct cellular compartments within the pituitary gland secrete highly specific trophic hormones in response to hypothalamic, intrapituitary, and peripheral hormonal and growth factor signals.
- Benign monoclonal adenomas can develop when specific types of pituitary cells proliferate and oversecrete their respective hormones.
- Acromegaly develops when somatotrophs (cells in the anterior pituitary gland that produce growth hormone) proliferate and oversecrete the hormone.
- Acromegaly is characterized by an acquired progressive somatic disfigurement, mainly involving the face and extremities, but also many other organs, that is associated with systemic manifestations.

CASE REPORT

- A 67-year-old female K/C/O Systemic hypertension, presented with the complaints of lower back pain since 10 years, insidious in onset, shooting type and was radiating to bilateral lower limbs, neck pain since 10 years, insidious in onset and radiating to bilateral upper limbs, bilateral knee pain since 10 years, insidious in onset, aggravates on walking up the stairs and relieves on taking rest and headache since 10 days, holocranial, dull aching type, not associated with vomiting, blurring of vision, photophobia, phonophobia.
- On general examination, pallor was present, no icterus, cyanosis, clubbing or lymphadenopathy. Her vital signs showed no fever, blood pressure of 150/90 mmHg, pulse of 102 beats/min; and respiratory rate of 18 breaths/min.
- Cardiovascular and Respiratory systems were normal. Per Abdomen examination was soft, non-tender, splenomegaly was present 2cm below the left costal margin. CNS examination was normal, no focal neurological deficits and field of vision was normal.
- The routine laboratory test results and urine analysis were within the normal limits except for hemoglobin of 9.1gm% and peripheral smear showed normocytic normochromic anemia and triglycerides of 330mg/dl.
- 2D ECHO showed sclerotic AV with trivial AR, grade-1 LVDD, mild MR, mild TR with mild PH, PASP-36mmHg, normal RV and LV function with EF-58%.
- X-Ray of PNS (water's view) showed mild opacification of left maxillary sinus, mild enlargement of B/L frontal sinuses and mild calvarial thickening.
- X-Ray of skull (lateral view) showed mild enlargement of frontal sinus, enlargement of sella turcica and enlargement and protrusion of lower jaw mandibular prognathism.
- X-Ray B/L hand showed mild hypertrophy of tufts of all distal phalanges, decreased carpo-metacarpal joint space, widening of base of all distal phalanges.

- X-Ray B/L foot showed mild hypertrophy of tufts of terminal phalanges of B/L great toes, hypertrophy of base of B/L terminal phalanges of great toes and base of all proximal phalanges, increased heel pan thickness bilaterally measuring 34mm on left side and 32mm on right side.
- X-Ray cervical spine showed anterior marginal osteophytes at multiple levels, reduction in anterior height of C3, C4, C5 and C6 and cervical lordosis is maintained.
- X-Ray LS spine showed anterior marginal and bridging osteophytes at multiple levels in lumbar and lower dorsal spine and lumbar lordosis is reduced.
- MRI Brain showed ~2.2 x 2.5 x 2.1 cm sized well defined isotense to grey matter lesion in sella turcica, the features are likely to represent adenoma of pituitary gland with compression of pituitary stalk.

Figure 1: The nose is widened and thickened, the cheekbones are obvious, the forehead bulges, the lips are thick and the facial lines are marked and macroglossia is noted. The forehead and overlying skin is thickened, sometimes leading to frontal bossing.

Figure 2a & 2b: As compared with the hand of a normal person (right), the hand of a patient with acromegaly (left) is enlarged in figure 2b, the fingers are widened, thickened and stubby, and the soft tissue is thickened.

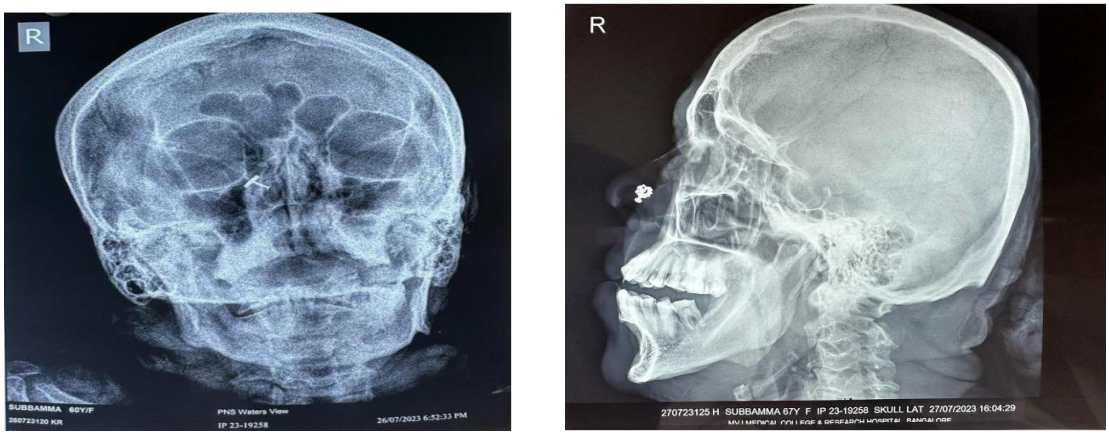

Figure 3: X-Ray of PNS showing enlarged frontal sinus and X-Ray of skull lateral view showing calvarial thickening and mandibular prognathism.

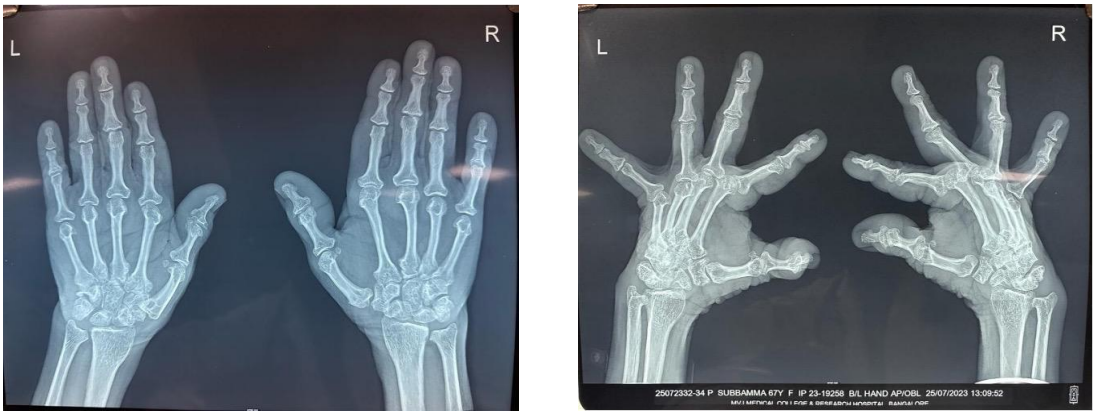

Figure 4a & 4b: X-Ray of hand AP and Oblique views showing mild hypertrophy of tufts of B/L distal phalanges, decreased carpo-metacarpal joint space and mild soft tissue hypertrophy.

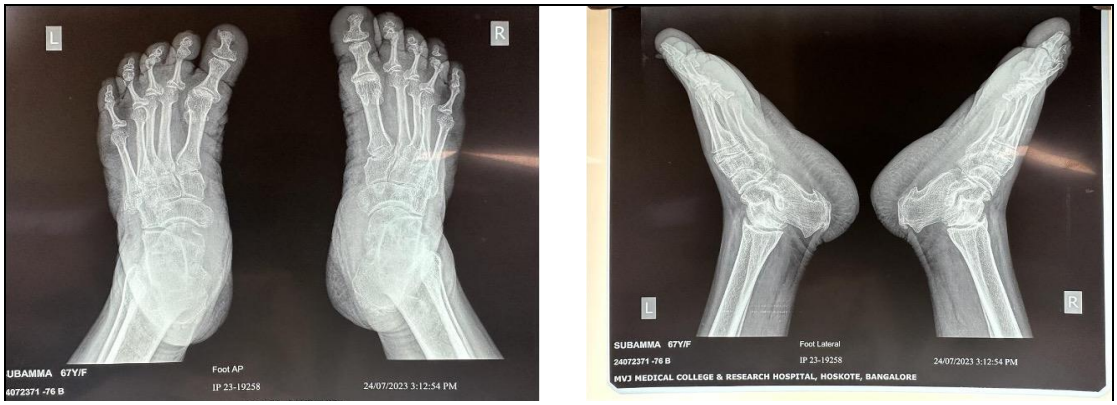

Figure 5a & 5b: X-Ray of feet AP and Oblique views showing increased heel pad thickness measuring 34mm on left side and 32mm on right side.

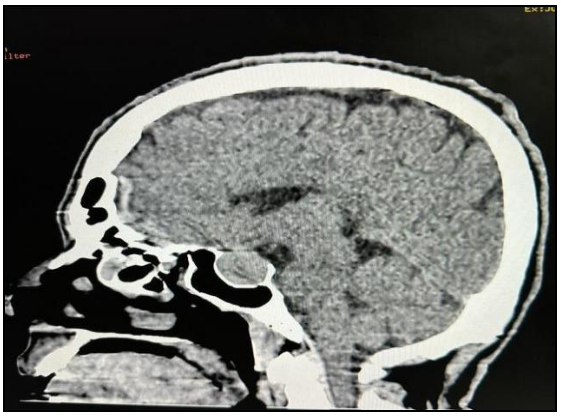

Figure 6: MRI of Brain showingadenoma of pitutary gland with compression of pitutary stalk.

DISCUSSION

- GH hypersecretion is usually the result of a somatotrope adenoma but may rarely be caused by extrapituitary lesions(i.e, pancreatic islet cell tumor, lymphoma).
- In addition to the more common GH-secreting somatotrope adenomas, mixed mammosomatotrope tumors and acidophilic stem cell adenomas secrete both GH and PRL.
- In patients with acidophilic stem cell adenomas, features of hyperprolactinemia (hypogonadism and galactorrhea) predominate over the less clinically evident signs of acromegaly.
- They present clinically as Acral bony overgrowth results in frontal bossing, increased hand and foot size, mandibular enlargement with prognathism, and widened space between the lower incisor teeth.
- Soft tissue swelling results in increased heel pad thickness, increased shoe or glove size, ring tightening, characteristic coarse facial features, and a large fleshy nose.
- Other commonly encountered clinical features include hyperhidrosis, a deep and hollow-sounding voice, oily skin, arthropathy, kyphosis, carpal tunnel syndrome, proximal muscle weakness and fatigue, acanthosis nigricans, and skin tags.

- Generalized visceromegaly occurs, including cardiomegaly, macroglossia, and thyroid gland enlargement.
- Cardiovascular system involvement may be in the form of Cardiomyopathy with arrhythmias, Left ventricular hypertrophy, Decreased diastolic function and hypertension ultimately occur in most patients if untreated.
- Respiratory system involvement may be in the form of Upper airway obstruction with sleep apnea occurs in >60% of patients, and is associated with both soft tissue laryngeal airway obstruction and central sleep dysfunction.
- Diabetes mellitus develops in 25% of patients with acromegaly, and most patients are intolerant of a glucose load (as GH counteracts the action of insulin).
- Acromegaly is associated with an increased risk of colon polyps and mortality from colonic malignancy; polyps are diagnosed in up to one-third of patients.
- Unless GH levels are controlled, survival is reduced by an average of 10 years compared with an age-matched control population.
- Age-matched serum IGF-1 levels are elevated in acromegaly and in our case it IGF-1 levels are 506.0 ng/ml.
- The diagnosis of acromegaly is confirmed by demonstrating the failure of GH suppression to <0.4 µg/L within 1–2 h of an oral glucose load (75 g).
- Prolactin should be measured, as it is elevated in ~25% of patients with acromegaly. Thyroid function, gonadotropins, and sex steroids may be attenuated because of tumor mass effects.
- The goal of treatment is to control GH and IGF-1 hypersecretion, ablate or arrest tumor growth, ameliorate comorbidities, restore mortality rates to normal, and preserve pituitary function.
- Surgical resection of GH-secreting adenomas is the initial treatment for most patients.

CONCLUSION

Acromegaly is usually clinically not diagnosed for 10 years or more. Early identification and prompt treatment enhance the prognosis of the patient. IGF-1 levels and Radiological imaging is the main stay in the diagnosis of pituitary adenoma. Surgical resection is the main stay of treatment.

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