

Pictorial CME

An Interesting Case of Acromegaly

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ABSTRACT

Acromegaly is caused by pituitary tumours that secrete GH (growth hormone) or very rarely by extrapituitary disorders. Regardless of the etiology, the disease is characterized by elevated levels of GH and IGF-1 (insulin like growth factor) with resultant signs and symptoms of hypersomatotropism. Prompt diagnosis and treatment can prevent the metabolic and other local complications due to this macroadenoma.

Key words : Acromegaly, Growth hormone, Macroadenoma, Treatment.



Fig. 1 : Patient having peculiar acromegalic facies (fleshy nose, prognathism, peg like incisors, macroglossia)



Fig. 3 : X- ray foot showing increased heel pad thickness

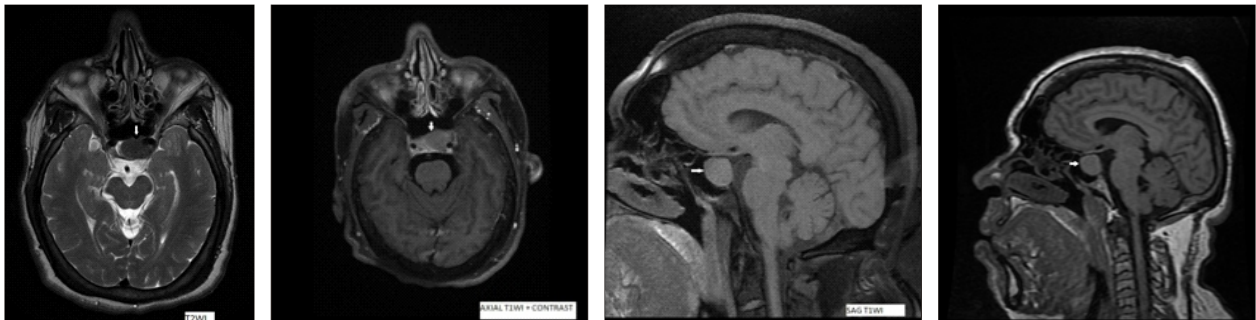


Fig. 2 : MRI Brain with contrast showing the pituitary macroadenoma in various views.(arrows)

Case Report :

A 64 years old postmenopausal woman was admitted with complaints of bilateral knee pain

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Received on 23rd June 2019

Accepted on 27th June 2019



Fig. 4 : X-ray skull shows sellar widening

since one month and swelling over both feet since 15 days. There was no history of swelling over joints, fever, oliguria, headache or visual symptoms. On examination she was found to be hypertensive with BP of 160/100 mmHg. There was pitting pedal edema. On careful observation, her facies were peculiar with a frontal bossing, coarse facial features, large fleshy lips, prognathism, widely spaced peg like incisors, large hands and feet and macroglossia. She was obese with a BMI of 27.8 kg/m². There were no skin tags or acanthosis, no goiter. She had a deep voice. A clinical suspicion of Acromegaly was kept and she was investigated further. There was no family history of such a phenotype resemblance in any of her relatives. She had no history of paraesthesias in upper limbs, no h/o galactorrhea and no h/s/o sleep apnea. She was hard of hearing since 7 years and was using hearing aids.

On laboratory investigations, her fasting plasma glucose was 165 mg% Post meal was normal and HbA1C was 6.6%. She was thus a newly diagnosed Diabetic with deranged kidney function also. Urea 55 mg% and sr. creatinine 3.92 mg%. She had Stage 5 CKD (chronic kidney disease) and was managed conservatively. She also had dyslipidemia (total cholesterol 242 mg% and LDL cholesterol 181 mg%). Initially we sent her serum IGF-1 (Somatomedin C) level as a screening test which turned out to be very elevated : 542 ng/ml (range 51 to 187 ng/ml). We had planned an oral glucose tolerance test to look for failure of GH level to be suppressed to less than 1 microgram/L but patient refused for financial reasons. However, an MRI Brain with contrast and pituitary cuts was done which confirmed our suspicion and revealed a pituitary macro-adenoma (a well defined rounded homogeneous altered signal intensity lesion in sellar and suprasellar region with minimal displacement of pituitary stalk measuring 1.4 x 2.5 x 1.7 cm in AP, Transverse and CC dimensions). Other hormonal tests were ordered. Her sr. prolactin was raised 70.15 ng/ml (1.2 to 19.2); T3 0.38 ng/ml, T4 7.42 microgram/dl and TSH 1.23 micro IU/ml (all

normal) FSH, LH and sr. cortisol were not done. Her Skull X-Ray showed Sellar Widening. X-Ray of the foot showed increased heelpad thickness (normal is 21mm). She was not cooperative for Perimetry. She also refused to undergo a polysomnography and a screening colonoscopy. Patient was reluctant for surgery and hence was started on Tablet Cabergoline 0.25 mg biweekly with plan to escalate dose monthly. She was also started on antihypertensive, diuretic, antidiabetic and statin medications.

Discussion :

In 1886, Pierre Marie published the first clinical description of disordered somatic growth and proportion and proposed the name “Acromegaly”¹. More than 95% of patients with acromegaly harbor a GH secreting pituitary adenoma. Manifestations of acromegaly are caused by either central pressure effects of the pituitary mass or peripheral actions of excess GH and IGF-1. Headache is seen in 50 to 60% of patients. Effects of hypersomatotropism on acral and soft tissue growth and on metabolic function occur insidiously over several years. The slow onset and elusive symptomatology often result in delayed diagnosis with a mean delay of almost 9 years². Patients may seek care for dental, orthopedic, rheumatologic or cardiac disorders. Hyperhidrosis is prominent in 70%³. Serial review of old photographs often accentuates the progress of subtle physical changes. Dyslipidemia, insulin resistance, hypertension, decreased libido are common metabolic complications. Stalk compression by tumor causes hyperprolactinemia and sometimes galactorrhea. Tumor mass compression of surrounding normal pituitary tissue may cause hypopituitarism. Acromegaly maybe a part of MEN1 (Multiple endocrine neoplasia 1 which includes hyperparathyroidism and pancreatic islet cell tumours). Familial Acromegaly due to germline inactivating mutations of AIP gene has been described especially in a subset of younger patients^{4,5}. A high index of suspicion is required to pick up this condition and systematic biochemical followed by imaging evidence will help to institute early therapy and prevention of complications.

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