

Case Report: Acromegaly

By 6th Year Medical Students, Faculty of Medicine, Thammasat University
Department of Neurosurgery, Thammasat University Hospital

1. Bhumirabhee Srirakoon 5929640166
2. Pijak Phothaporn 6029640072
3. Nawit Yoosuksomboon 6029640080
4. Nanthida Arora 6029640098
5. Nicharee Teeraumphon 6029640122

Patient's Profile: A 40-year old Thai female from Saraburi who is an accountant.

Chief Complaint: Facial Structure Changes 5 months PTA

Present Illness:

10 years PTA: Patient wore size 4 shoes and weighed 61 kg.

5 years PTA: Patient experienced bilateral tingling numbness of fingers and hands after working on the keyboard. Her symptoms were not relieved with rest and medication. She was diagnosed with carpal tunnel syndrome and received a carpal tunnel release surgery on her left hand.

4 years PTA: Patient noticed that she could no longer fit in her shoes and switched to size 6 shoes. She was not able to wear her wedding ring. She reported increased appetite and weight gain which she thought was due to her DM. Patient was diagnosed with DM. Patient also received a carpal tunnel release surgery on her right hand.

3 years PTA: Patient no longer has periods.

5 months PTA: Patient noticed that her face has become 'larger' with increased protrusion of jaws and deepening of voice. Therefore, she sought medical attention. Patient's shoe size increases to a size 7 with an increase in weight to 71 kgs (increased from 61 kg 10 years prior). She reports enlargement of her tongue with snoring every night. Patient does not feel tired/sleepy during the day and does not have to wake up at night to grasp air. She denies blurry vision, double vision, narrowing of vision, headache, nausea/vomiting, diarrhea/constipation, chest pain, dyspnea, palpitations, milky discharge from nipple, breast pain, joint pain. Patient voids 2 times at night and 4 times during the day. Patient decided to visit Saraburi hospital where lab investigations were done and the patient was referred to Thammasat University Hospital.

On Admission: Patient's signs and symptoms were the same as 5 months prior.

Past History

- Underlying diseases: DM (diagnosed 4 years ago), DLP (diagnosed 2 years ago)

- Current Medications:
 - Metformin (500mg) 2 tabs po bid pc
 - Pioglitazone (15mg) 1 tab po OD pc
 - Insulin Aspart 30% + Aspart Protamine (100U/ml)
 - Vitamin D2 Cap 20,000 IU
- No known food or drug allergies
- No history of herbal drug use, NSAIDs or anticoagulants use
- Surgical history:
 - Carpal tunnel release surgery left side 5 years ago and right side 4 years ago
- No history of trauma

Social History:

- Alcohol consumption: social beer drinker
- No smoking history

Family History:

- No family history malignancies
- Mother had diabetes.

Gynecologic History:

- Menarche 15 years old
- Menstruation history: interval 30 days, duration 3 days, regular menstruation, used 2 pads/day (not full), mild dysmenorrhea on day 1 with pain score of 3/10
- History of OCPs use from age 15 to 38 years (for 23 years)
- Menopause 38 years old, age of maternal menopause not remembered
- Patient has 1 child, no complication during delivery, no postpartum hemorrhage

Physical Examination:

V/S	BP 132/77 mmHg PR 90 bpm RR 18 /min BT 36.8 °C
Anthropometrics	BW 71 kg Ht 154 cm BMI 29.9 kg/m ²

GA	Thai female, alert, oriented to time, place and person no pallor or jaundice, no dyspnea
HEENT	No pale conjunctiva, anicteric sclera, no parotid enlargement, no palpable cervical or supraclavicular LN, increased frontal bossing, enlarged nose base, thickening of lips, increased prognathism, widened space between teeth, macroglossia
CVS	Full and equal pulses all extremities, no heave or thrill, normal S1&S2, no murmur
RS	Clear and equal breath sounds both lungs, no adventitious breath sounds, no retractions
Abd	No distension, no surgical scars, normoactive bowel sounds, soft, not tender, no guarding, no hepatosplenomegaly, no flapping tremor, no suprapubic or CVA tenderness
Extremities	No edema, no petechiae or purpura, enlargement of hands and feet

Physical Exam



Neurological Exam:

- Mental status: good consciousness, oriented to time, place and person
- Sensory: normal sensation to pain, temperature and touch
- Reflex: DTR 2+ (biceps, triceps, brachioradialis, patella and achilles)
- Motor: normal muscle tone, muscle power grade V/V all extremities, no pronator drift
- Babinski: plantar flexion
- Cerebellar sign: no dysdiadochokinesia, normal finger-to-nose test
- Cranial nerves:
 - CN I: normal sense of smell (tested using soap and coffee beans)
 - CN II: Pupil 3mm RTLBE, RAPD negative, VA sc 20/100 BE, c PH 20/20 BE, no visual field defects, A:V ratio 2:3, no papilledema, no hemorrhage/exudate
 - CN III, IV, VI: full EOM
 - CN V: normal facial sensation, normal strength of masseter and temporalis muscle
 - CN VII: no facial palsy
 - CN VIII: no lateralization of Weber's, Rinne's air > bone
 - CN IX, X: no uvular deviation, normal gag reflex
 - CN XI: normal strength of trapezius and sternocleidomastoid muscle power
 - CN XII: no tongue deviation, no tongue fasciculation

Pertinent Findings:

1. Increasing in appetite and weight
2. Early menopause
3. Enlargement of hands and feet
4. Increased frontal bossing
5. Enlarged nose base
6. Thickening of lips
7. Increased prognathism
8. Widened space between teeth
9. Macroglossia
10. No parotid enlargement
11. Obesity
12. No visual field defects

13. No headache
14. No nausea or vomiting
15. No galactorrhea or nipple discharge
16. History of carpal tunnel syndrome s/p carpal tunnel release surgery
17. U/D: DM, DLP

Problem List:

1. Acromegalic Change
2. U/D: DM, DLP, Obesity

Differential Diagnosis:

1. Pituitary tumor

Supporting:

- Acromegaly features (acral enlargement, coarse facial features, teeth separation)
- Compatible age group with onset of changes
- Most common cause of acromegaly is pituitary adenoma
- History of carpal tunnel syndrome
- Early menopause

Against:

- No mass effect (such as headache, visual changes)
- No other hormonal abnormalities (such as galactorrhea)

2. Non-Pituitary GH-Secreting Tumor

Supporting:

- No mass effect
- Acromegaly features (acral enlargement, coarse facial features, teeth separation)

Against:

- No other organ specific symptoms (lungs, pancreas and adrenals)

3. Hypothyroidism

Supporting:

- Excessive weight gain and obesity

- Early menopause
- No mass effect (visual changes, headache)

Against:

- No other hypothyroid symptoms (cold intolerance, decreased appetite, fatigue, constipation)
- Usually does not present with morphological changes of bone and skin thickening
- No history of hyperthyroid phase
- No enlarged thyroid

Investigation:

- Anterior pituitary hormones

Prolactin	190 ng/mL
Estradiol	<5 pg/mL
TSH	1.01 uIU/mL
LH	0.384 mIU/mL
Free T4	1.09 ng/mL
FSH	2.17 mIU/mL
Cortisol	15.03 mcg/dl

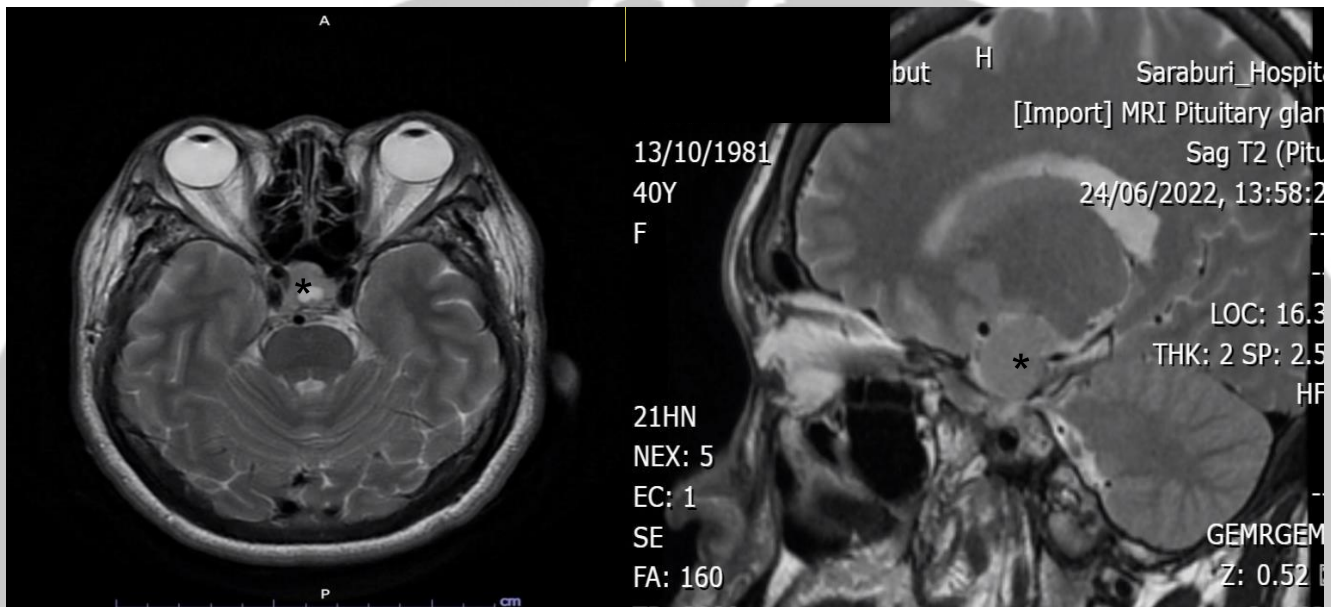
- IGF-1: 751 ng/mL
- 75g oral glucose suppression test

Time (mins)	0	30	60	120	180
Blood Sugar (mg/dl)	144	281	341	264	181
Growth Hormone (ng/ml)	26.1	24.9	30.1	35.6	29.7

- MRI Brain with Gd

IMP: Rt side sellar pituitary macroadenoma (Asterisk) incomplete surrounding right ICA size

1*1x1*1 cm with Gd enhancing with abutting to Rt optic nerve



Management

- Set OR for endoscopic transsphenoidal surgery for tumor removal

Acromegaly

Epidemiology

Acromegaly is a rare medical condition, although it can happen at any age many studies found higher rates of diagnosis in middle-aged adults. Sex differences are insignificant. In reference to a systematic review and meta-analysis of global epidemiology of acromegaly by European

Journal of Endocrinology, The mean age of diagnosis ranged between 40 to 47 years of age. The pooled prevalence of acromegaly was 5.9 (95% CI: 4.4–7.9) per 100 000 persons, while the incidence rate (IR) was 0.38 (95% CI: 0.32–0.44) cases per 100 000 person-years.

Etiology

- Excess Growth Hormone Secretion
 - Pituitary Tumor
 - Growth hormone cell adenoma
 - Mammotroph adenoma
 - Somatotroph hyperplasia
 - Growth hormone cell carcinoma
 - Ectopic Pituitary Tumor
 - Sphenoid or parapharyngeal sinus location
 - Extrapituitary Tumor
 - Pancreas, Lung, Ovary, Breast location
- Excess Growth Hormone-Releasing Hormone Secretion
 - Eutopic
 - Hypothalamic hamartoma
 - Ectopic
 - Small cell lung cancer
 - Adrenal adenoma
 - Pheochromocytoma
- Excessive growth factor secretion
 - Acromegaloidism

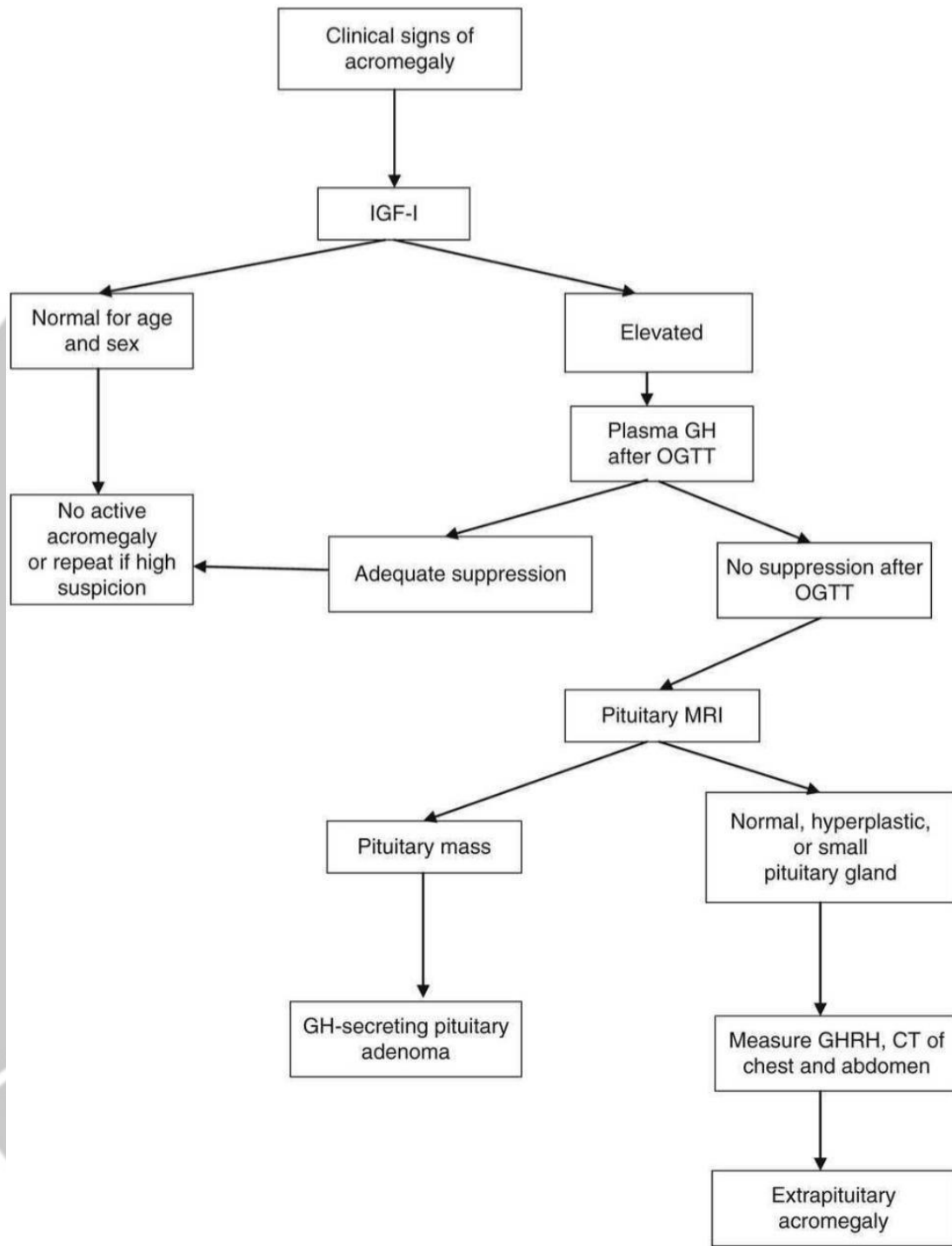
Pathophysiology

Acromegaly is caused by an elevation of growth hormone - most commonly secreted by pituitary adenomas. Excess in growth hormones results in increased glycogen breakdown and IGF-1 by the hepatocytes. IGF-1 increases cell growth and proliferation, bone mass and bone length.

Clinical presentation and Diagnosis

Symptoms of Acromegaly are vary and tend to develop very slowly over time. Early symptoms include swollen hands and feet, tiredness, difficulty sleeping, changes in facial features (larger bone, larger nose, widening of gap between teeth), and numbness or weakness of hands (carpal tunnel syndrome). Other possible symptoms include hand and feet enlargement, coarser skin, oilier skin, deepening of voice, blurred vision, and joint pain.

The physicians may suspect Acromegaly from history, symptoms and clinical signs. Measuring IGF-1 level and growth hormone levels before and after a 75g oral glucose tolerance test (OGTT), and imaging can help diagnose Acromegaly. The diagnosis algorithm for acromegaly is shown below.



Endocrinol Nutr. 2013;60:457.e1–457.e15

Investigation

- IGF-1
- Growth hormone suppression test
- Pituitary MRI with contrast

Treatment

The definite treatment and one that is advised for most patients is to undergo transsphenoidal tumor. However, nonsurgical (medical, radiotherapy) management may be considered in patients who are unfit for surgery or in cases of incomplete resection.

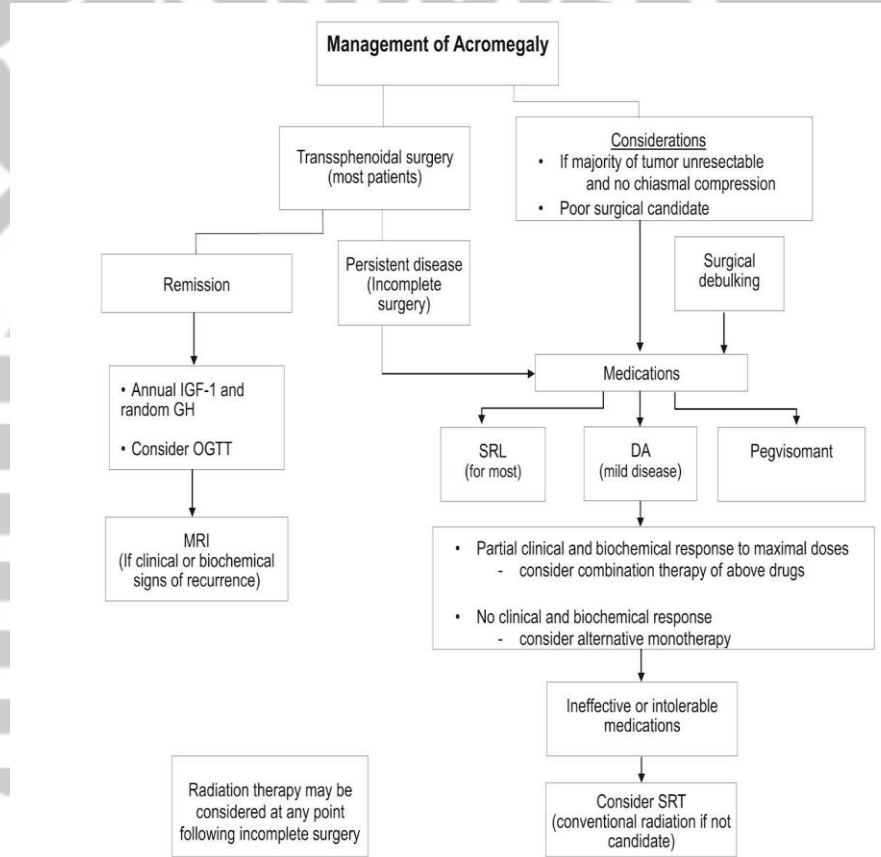


Figure 1. Treatment considerations in the approach to a patient with acromegaly. This approach refers to management of a patient with a pituitary adenoma. DA, dopamine agonist; OGTT, oral glucose tolerance test.

Post-operative care

- Allow oral fluid but do not force oral fluid
- Advice to lie flat, can sleep sideways
- Avoid blowing your nose, using straws, coughing and sneezing
- Nasal care:
 - Mometasone furoate 1 puff both nostrils bid
 - Oxymetazoline hydrochloride 1 drop prn for nasal bleeding

References

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