

Remission of Acromegaly after transsphenoidal surgery: A case report

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Abstract

Acromegaly is a rare, chronic disorder that mostly results from growth hormone (GH)-secreting pituitary adenoma. It is a rare neuroendocrine condition that can lead to significant morbidity. Transsphenoidal surgery is the first-line treatment of this adenoma. We report a case of recovery from acromegaly after post transsphenoidal surgery.

Keywords: Acromegaly; Growth Hormone; Insulin-Like Growth Factor 1; Pituitary, Remission.

1. Introduction

Acromegaly is a rare disease resulting from an uncontrollable hypersecretion of growth hormone most often by a pituitary adenoma. Treatment can be surgical, medical or by radiotherapy. Transsphenoidal surgery is still the first-line treatment for acromegaly with rapid normalization of IGF-1. We report a case of recovery from acromegaly after post transsphenoidal surgery.

2. Observation

A 43-year-old woman, without any particular medical history, presents for 5 years with a dysmorphic syndrome made of an enlargement of the extremities with a sign of the ring and a change of shoe size (from 6 to 9 us), and 3 years later, a hoarseness of the voice, snoring, hypersudation. She did not present with decreased visual acuity or headaches.

Clinical examination notes a conscious patient, blood pressure 123/67 mmHg, heart rate 82 beats/min and respiratory rate 18 breaths/min.

There is also moderate obesity with a BMI of 34.5 kg/m²; facial dysmorphic syndrome, pudgy hands and feet, and thickened skin on the hands and feet.

The hormonal assessment showed: IGF-1: 898.3 ng/ml (N:75-249); Prolactin: 81.69 ng/ml; TSH: 1.40 µUI/L; FT4: 12 pmol/L; FSH: 4.76mUI/mL; LH: 1.88mUI/mL; Estradiol: 65.27 pg/mL; Testosterone: 0.6 ng/mL; Cortisol: 11.90 µg/dL; and, HbA1c: 5.5%.

Pituitary MRI showed a pituitary macroadenoma measuring 12.7x9.2x8.8 mm, T1 isosignal, T2 hyposignal, without contrast, with two cystic areas measuring 2 mm and 1.6 mm respectively, with a bulging aspect of the roof of the pituitary gland and the anterior wall of the sella turcica, and respect for the optic chiasm (Figure 1). The visual field is normal. Abdominal, cervical and transthoracic ultrasound were without abnormality.

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The diagnosis of acromegaly by pituitary macroadenoma was retained.

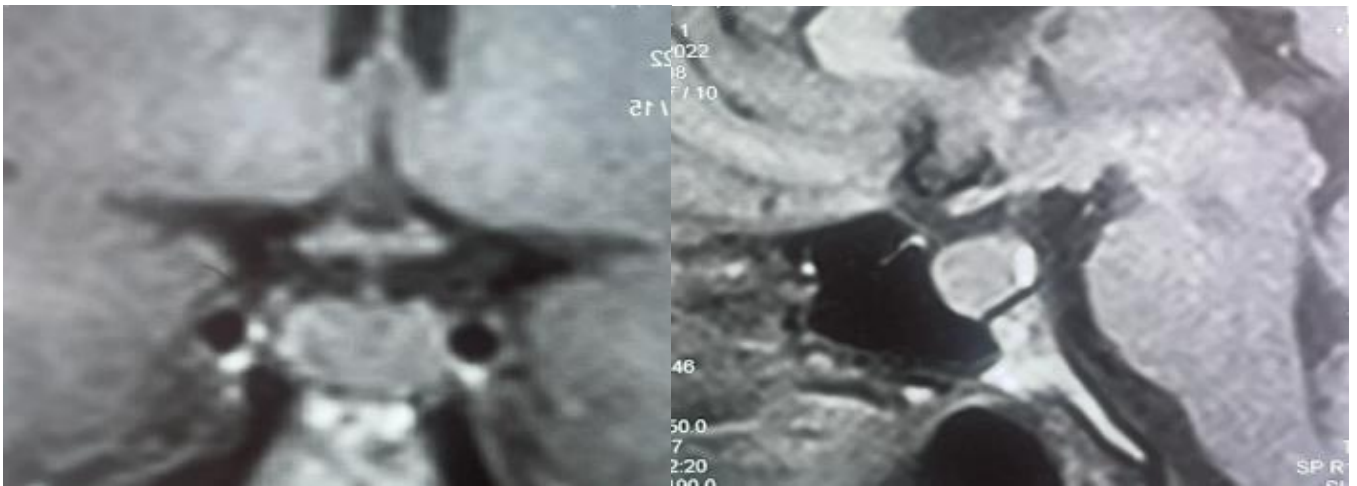


Figure 1 Preoperative sagittal and coronal hypothalamic-pituitary MRI showing a sellar tumor process measuring 12.7x9.2x8.8 mm

The patient was operated by transsphenoidal approach and the anatomic-pathological and immunohistochemical study revealed a pituitary adenoma expressing anti GH and anti PRL antibody.

However, the patient presented postoperatively with diabetes insipidus and was put on Minirin.

The patient was then seen at 3 and 6 months after her surgery.

The follow-up clinical examination noted a moderately obese patient but with regression of the facial dysmorphic syndrome with a return to the initial shoe size.

The hormonal workup showed normalization of IGF-1 level at 144.5 ng/mL (N:75-249); Prolactin level at 6.83 ng/mL.

The control pituitary MRI was without abnormalities or signs of recurrence (Figure 2).

Suppressed GH test during an oral glucose tolerance test shows a GH Nadir at 0.07 ng/mL (N < 0.4 ng/mL). The patient is in remission after 6 months of transsphenoidal surgery of her pituitary adenoma.

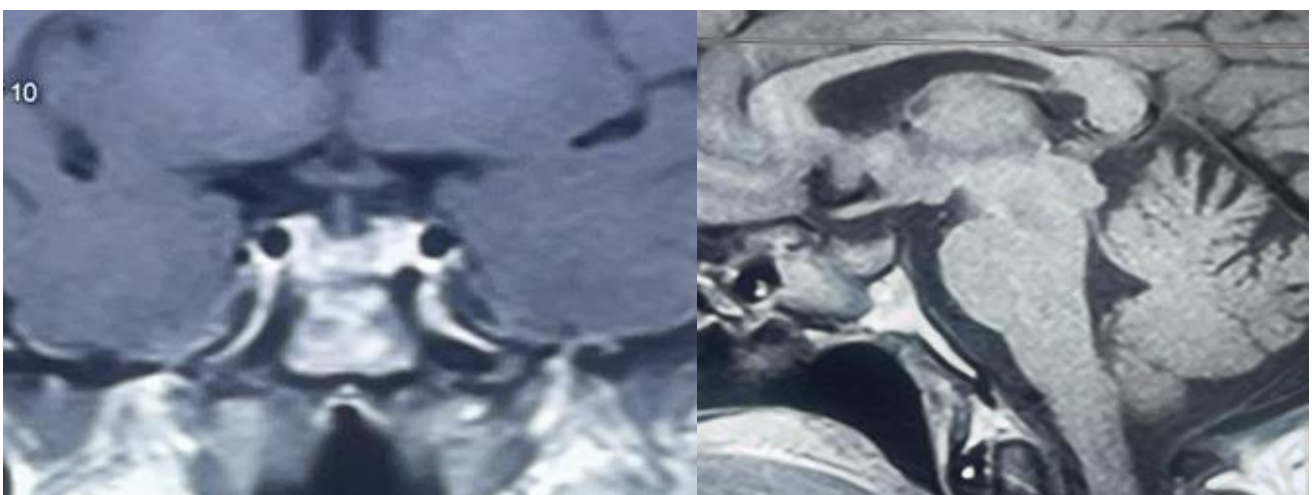


Figure 2 Post-operative sagittal and coronal hypothalamic-pituitary MRI showing no abnormalities or signs of recurrence

3. Discussion

Acromegaly is typically caused by a growth hormone-secreting pituitary adenoma, driving excess secretion of insulin-like growth factor 1. In more than 95% of cases, acromegaly is secondary to GH hypersecretion by a benign monoclonal pituitary adenoma which develops from somatotroph cells. [1] Its prevalence is estimated at 40-130 cases per million inhabitants. [2] Acromegaly is characterized by slowly progressive acquired somatic disfigurement (mainly involving the face and extremities) and systemic manifestations. Acromegaly may result in a variety of cardiovascular, respiratory, endocrine, metabolic, musculoskeletal, and neoplastic comorbidities. Early diagnosis and adequate treatment are essential to mitigate excess mortality associated with acromegaly. [3] Acromegaly is generally suspected on clinical signs and symptoms: the dysmorphic syndrome. The extremities (hands and feet) are broadened, the fingers are widened, thickened and stubby, and the soft tissues are thickened. The patient may have had to enlarge his or her ring in recent years, or to change shoe size. The facial aspect is characteristic, and patients with established acromegaly are generally alike in this respect: the nose is widened and thickened, the cheekbones are prominent, the forehead bulges, the lips are thick and the facial lines are marked.

The diagnosis of acromegaly is confirmed on biochemical grounds, including elevated serum insulin-like growth factor 1 and lack of growth hormone suppression after glucose administration. Pituitary magnetic resonance imaging is advised in patients with acromegaly to identify an underlying pituitary adenoma. Transsphenoidal pituitary surgery is generally first-line therapy for patients with acromegaly. Tumor excision, usually by the transsphenoidal route, is the most rapid way of reducing GH and IGF-1 concentrations in patients with acromegaly. Nevertheless, levels normalize in only 40–70% of cases [4], depending on the size of the tumor (microadenomas are more amenable to cure), the preoperative GH concentration, and the surgeon's experience. Endoscopic techniques are now used in the majority of expert centers [5]. Surgical outcome is carefully assessed at 3 months.

Mixed GH- and prolactin (PRL)-secreting adenomas are frequent (25%). Some adenomas contain both cell types, while other develop from a mammosomatotropic stem cell and consist of more mature monomorphic cells that express both GH and PRL. [6] Remission was defined as a normal IGF-I and either a suppressed GH less than 0.4 ng/ml during an oral glucose tolerance test or a random GH less than 1.0 ng/ml. IGF-I levels measured 6 weeks postoperatively can be used in most patients to assess remission, although patients with mildly elevated IGF-I may yet normalize by 3–6 months. [7]

Preoperative insulin-like growth factor 1 index ≥ 2.5 and Knosp classification grade 3-4 are important prognostic factors that determined remission outcome after treatment. Patients who have both of these poor prognostic factors should be aggressively treated with surgery, medication, and probably radiation to optimally control the disease while patients with neither of these factors had high remission rates (87.5%) following surgery like our patient. [8]

4. Conclusion

Acromegaly is a rare, chronic disorder that mostly results from growth hormone (GH)-secreting pituitary adenoma. Transsphenoidal surgery is the first-line treatment of this adenoma. Remission of acromegaly can be achieved by surgery alone, especially when the diagnosis is early and the tumor is not invasive.

Compliance with ethical standards

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Conflict of interest statement

I declare no conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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