Scholars Journal of Medical Case Reports

OPEN ACCESS

Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559(Online)
Journal homepage:https://saspublishers.com

Case Report of Acromegalo-Gigantism in Mixed Adenoma

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DOI: 10.36347/sjmcr.2021.v09i02.007 | **Received:** 01.02.2021| **Accepted:** 08.02.2021| **Published:** 12.02.2021

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Abstract **Case Report**

Before the end of the 19th century, many reports dealing with acromegaly and gigantism appeared. The two syndromes were clinically linked to each other by Brissaud and Miege in 1895; Hutchinson described the pathological similarity in 1900. Thirty-two years later, Cushing reaffirmed that pituitary adenomas were routinely found in patients exhibiting acromegaly or gigantism. He expressed the theory that the relative rarity of gigantism resulted from the recognized lack of adenoma formation in early life. Following Cushing's writings in 1927, scattered case reports dealing with the subject of pituitary gigantism appeared. Recent reports have presented the first detailed studies on human growth hormone (HGH) levels and their response to various manipulations in two giants, but it is still true that much of what is written in textbooks regarding Pituitary gigantism is derived by inference from what is known about the more common acromegaly syndrome in adults.

Keywords: Acromegalo-Gigantism, Pituitary adenoma, human growth hormone (HGH), acromegaly syndrome.

INTRODUCTION

Acromegalo-gigantism is a rare condition characterized by extreme size and physical stature occurring in children or adolescents before the epiphyseal growth plaques close (gigantism), followed by acromegaloiddysmorphic syndrome, defining acromegalo-gigantism, due to hypersecretion of growth hormone (GH) most often secondary to a pituitary adenoma which may be pure somatotropic (60% of cases) or sometimes mixed [7]. The main symptom of the disease is accelerated abnormal growth affecting the musculoskeletal system associated with other co-morbidities. Resection, most often transsphenoidally, is the fastest way to reduce GH and IGF-1 levels. However, complete remission of the disease is usually not achieved with surgery alone, and pharmacological treatment becomes necessary, of which somatostatin analogues are the most common. If no response is obtained with somatostatin analogues, a dopamine agonist or a GH receptor antagonist may be added. In cases that do not respond to surgery and pharmacological treatment, radiotherapy is used; however, the risk of hypopituitarism should be taken into account [5]. We report the case of a patient followed for acromegalo-gigantism on mixed adenoma, who has had a surgical resection and then placed on a somatostatin analogue and a dopaminergic agonist.

CASE REPORT

24-year-old patient, who has presented gigantism for 10 years (rapid height gain, change in size) with progressive pituitary tumor syndrome (headache, decreased visual acuity).Follow-up for depressive syndrome on anti-depressant and for diabetes mellitus for 6 months for which he consulted.The requested IGF1 returned to 677.5ng / l (normal: 120-238) or 2.8 times normal with a cerebral MRI showing a sellar and suprasellar tissue lesion process measuring 4 * 3.5 * 3.1cmexerting a mass effect on neighboring structures.Clinical examination revealed a large height (2.02m), acromegaloiddysmorphic syndrome with macroglossia, prominence and enlargement of the nose, bulging fingers and toes, nodular goiter and bilateral gynecomastia.

Fig-1: Patient gigantism

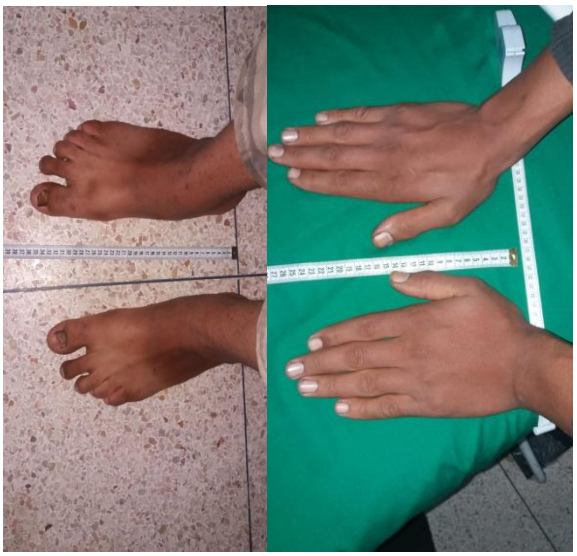

Fig-2: Bulging fingers and toes

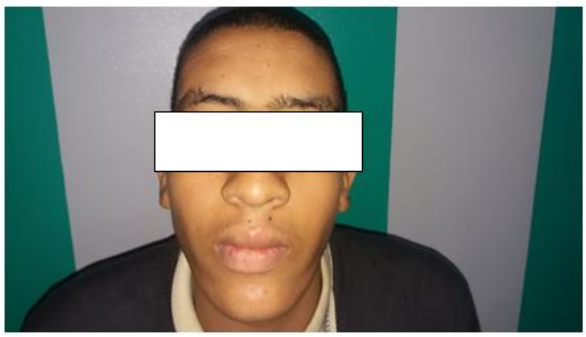

Fig-3: Acrofacial dysmorphia

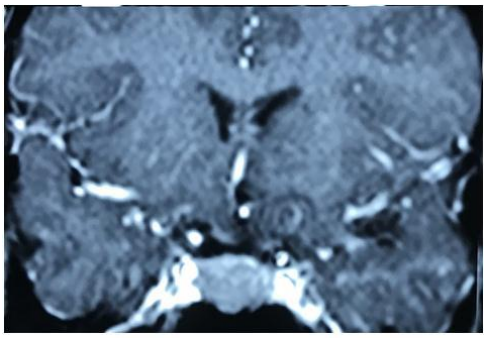

Fig-4: Macroadenoma on MRI

A further work-up revealed a thyrotropic insufficiency, a slight increase in prolactinemia, the electrocardiogram and the Goldman visual field were normal, the cervical ultrasound objectified a multinodular goiter whose fine needle aspiration of the thyroid nodules was benign, the abdominal ultrasound objectified a homogeneous hepatomegaly. The pituitary MRI revealed a sellar and supra sellarlesional process of 2.8 * 4.5 * 3.2cm responsible for an enlargement of the sellaturcica with collapse of the sellar floor at the bottom, at the top a filling of the optochiasmatic cistern including the optic chiasma. The diagnosis of acromegalo-gigantism in a mixed pituitary macroadenoma was suggested in view of gigantism and acromegaloiddysmorphic syndrome, confirmed by IGF1 and bilateral gynecomastia. A transsphenoidal surgery was performed with anatomopathology and immunohistochemistry a morphological appearance compatible with a pituitary adenoma expressing the anti GH antibody and focal (5%) the anti prolactin antibody with 4% Ki67. Postoperative pituitary MRI revealed a tumor residue, the patient was put on somatostatin analogue and dopaminergic agonist (cabergoline), a surgical revision is planned.

DISCUSSION

Acromegalo-gigantism is an extremely rare disease caused by increased secretion of growth hormone. Gigantism differs from acromegaly in the time of its onset, that is, before and after epiphyseal fusion respectively. The extra large size is very rare these days due to early care. The most consistent biochemical abnormality seen in patients with acromegalo-gigantism is elevated IGF-1. Some patients with hypogonadism responsible for delayed fusion of the epiphyses may continue to grow up to their twenties or even their thirties. The Gold Standard for diagnosing excess GH is based on the inability to suppress serum GH to an appropriate level (typically <1 ng / ml) after an oral glucose tolerance test (OGTT). Hyperprolactinemia is also frequently observed, usually due to co-secretion . In fact, adenomas secreting GH and prolactin are the most frequent mixed adenomas, either they contain both cell types, or they are developed from a stem cell, mammosomatotropic: the same cells, monomorphic, more mature, express both GH and prolactin [8].

Most GH pituitary adenomas are macro adenomas (approximately 80% of cases) and 30 to 60% are invasive. A higher frequency in men is reported in the literature. A late diagnosis of acromegalo-gigantism can occur due to poor understanding of the extent of symptoms, delayed consultations, and limited knowledge of the disease by general practitioners. It is the case with our patient who consulted several times for co morbidities linked to his acromegalo-gigantism before being referred to us. The therapeutic goals are to relieve symptoms, reduce tumor volume, prevent recurrence and improve long-term morbidity and mortality. The criteria for cure or good control are now much more stringent: it is required that the concentration of GH, be reduced to less than 1µg / l or 3mIU / l and IGF- 1 is standardized. To achieve these goals, a multi-step treatment strategy is often required. In acromegalo-gigantism, surgery is the procedure of first choice and can be curative. When surgical treatment is indicated as a first-line treatment, it makes it possible to obtain an immediate result if the resection is complete. The most commonly used way in pituitary adenoma surgery is the transsphenoidal one. This technique is minimally invasive and causes limited side effects and postoperative complications .

Several factors influence the effectiveness of surgical treatment:

- Supra-sellar extension, degree of intracavernous invasion and patient age.
- The size of the tumor: the larger the size of the adenoma, the less effective the surgical treatment, on the other hand, well limited non invasive adenomas have a very good prognosis.
- Preoperative GH and IGF1 levels: the lower they are, the better the postoperative remission. - Finally, the other essential factor to take into account is the experience of the neurosurgeon.

Because pituitary surgery is a very specialized surgery, remission rates are better in teams that are used to frequently operating pituitary adenomas. Biochemical control is obtained in approximately 70% of patients with intrasellarmicroadenomas, although this rate is lower with macroadenomas (approximately 40%). In view of this poor biochemical control by surgery, patients with acromegalo-gigantism generally require additional pharmacological management with somatostatin analogues. Dopaminergic agonists are useful in cases of associated hyperprolactinaemia or as adjunct therapy to somatostatin analogues in cases with lack of biochemical control and IGF-1 levels up to 1.5 times greater than normal range. Because according to a meta-analysis, cabergoline normalizes IGF-1 in 34% of patients, rather those whose IGF-1 is moderately increased [5,8].

If there is no response to somatostatin analogues, combination with a GH receptor antagonist is recommended.Appropriate monitoring with testing should be done due to the risk of tumor growth even with adequate biochemical response and improvement in symptoms.The use of radiotherapy as a third-line treatment should be considered, but the risk of hypopituitarism should be considered.To avoid the continued growth of flat bones in patients with tumor residue, management with somatostatin analogues is required, although the best option for residual tumors is combination therapy between somatostatin analogues andGH receptor antagonists [5].

CONCLUSION

This article highlights the difficulty of managing acromegalogigantism, in particular the unsatisfactory results of transsphenoidal surgery. It is important to note that the diagnosis is often late and transsphenoidal surgery is the first treatment, and due to the poor biochemical control, pharmacological treatment with somatostatin analogues is necessary. The association with dopaminergic agonist is systematic especially in the case of associated perprolactinemia. In case of residual tumor and lack of control by somatostatin analogues, combination with GH antagonists is recommended.Raiotherapy remains the last intention.

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