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Case Report

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Acromegaly with empty sella, a case report

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Abstract

Acromegaly is characterized by progressive somatic deformity and systemic symptoms associated with excessive growth hormone (GH) secretion. The mean age at diagnosis of acromegaly ranges from 40 to 47 years, with a prevalence of 28–137 per million and an incidence of 2–11 new patients per year. Men and women are equally affected. Due to its slow progression and insidious nature, diagnosis is often delayed.[1]

In approximately 95% of cases it is due to GH-secreting pituitary adenoma, and most tumors are visible on magnetic resonance imaging (MRI) of the Sella. The GH-producing adenomas of the pituitary gland are usually larger than 1 cm and can be easily identified on MRI, micro adenomas are rare. In 5% of cases, acromegaly results from ectopic secretion of growth hormone-releasing hormone (GHRH). Therefore, if an adenoma cannot be detected on pituitary MRI, a contrast-enhanced computed tomography (CT) scan of the chest, abdomen, and pelvis (CT CAP) should be performed to look for the source of ectopic GH / GHRH. In rare cases MRI can be entirely normal or may show an empty Sella (ES).[1] In this case report, we present a patient with acromegaly and ES on pituitary MRI.

Keywords: acromegaly, empty sella

Case report

A 62-year-old Saudi postmenopausal female with a 10 -year history of diabetes mellitus was referred to our endocrine clinic as a case of micro adenoma to rule out acromegaly. She reported no history of nausea, vomiting, headache, blurred vision, coarsening of her facial, change in ring nor shoes size, constipation or joint pain. She denied any previous history of a pituitary procedure or head and neck trauma and radiation.

On examination, she is 153.5 cm tall, weighted 60 kg with a BMI of 25.4. Her blood pressure was 140/ 78, pulse of 117 with normal respiratory rate & temperature. The patient had a big nose, prognathism & her hand were spade-like. Neck examination revealed a palpable 1.5 cm left thyroid nodule. Systemic cardio-respiratory and abdominal examinations otherwise normal. Her visual fields, visual acuity and fundoscopy were unremarkable.

Biochemical testing showed an elevated GH & IGF-1 levels. A 75 oral glucose tolerance test showed no suppression of GH values; nadir GH level of 7.66 ng/mL (Table 1). Prolactin level post -PEG precipitation was normal as well as the reminder of the pituitary panel investigations (Table 2).

Table 1 75 g oral glucose tolerance test

Reference range (min)	Glucose (mmol/L)	GH(ug/l)
0	0	2.36
30	15.8	2.74
60	20.3	3.79
90	22.2	5.94
120	18.9	7.66

Table 2 Summery of biochemical results

Variable	Initial result	Follow up results after initiation of octreotide	Reference range
Growth hormone (GH)	5.94	3.51	0 – 5 ug/L
Insulin-like growth factor (IGF)	599.2	473	54.6 - 185.7 ng/mL
Prolactine	29	8	5.18- 26.53 ug/L
Luteinising hormone (LH)	24.15	23.9	Post-menopausal: 10.39 - 64.57 mIU/ml
Follicle-stimulation hormone (FSH)	56.47	66.01	Post-menopausal: 26.7 - 133.4 mIU/ml
Estradiol	82	0	Post-menopausal: <103 pmol/L
Thyroid-stimulation hormone (TSH)	1.13	0.96	0.35 - 4.94 mIU/L
Adrenocorticotropic hormone (ACTH)	22.51	18.27	0.35 - 4.94 mIU/L
Cortisol	173	212.2	101.2 - 535.7 nmol/L

A pituitary MRI revealed a partially empty Sella, with no evidence of pituitary adenoma (Figure 1). In order to exclude an ectopic source of the GH hypersecretion, further imaging (CT CAP, octreotide scan) tests were performed, all were normal. Whole-body 68 Gallium DOTATATE PET/CT revealed normal uptake in the pituitary and body (Figure 1). Thyroid US showed, two nodules the left lobe, measuring 2 cm (TR3 & TR4) with benign cytology. She refused to do colonoscopy. As clinical context of acromegaly with an empty Sella,

patient has been commenced on 20 mg monthly octreotide acetate. After receiving 4 doses of octreotide, her IGF-1 & GH has reduced (Table 2).

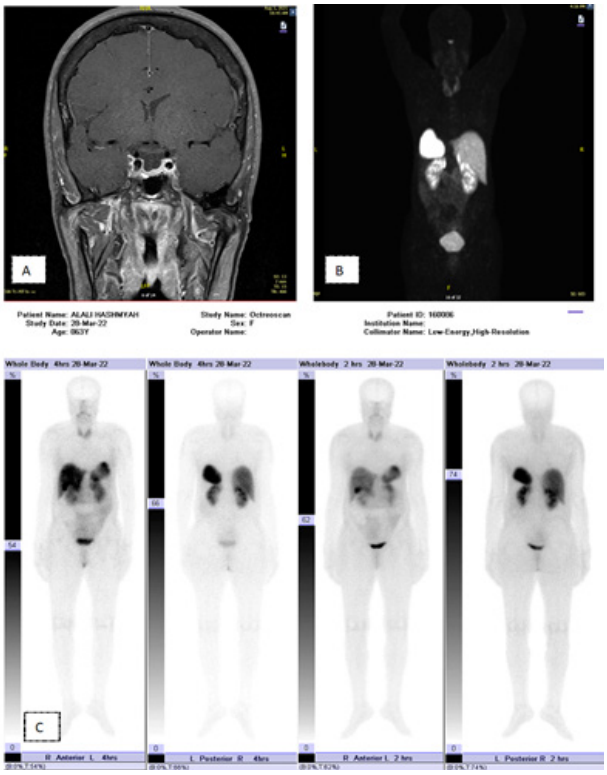

Figure 1 A: MRI shows empty Sella, B: DOTATATE PET/CT showing receptor activity in the pituitary gland due to physiological somatostatin receptor expression & C: Octreotide scan shows physiological radiotracer distribution with no abnormal activity to suggest neuroendocrine tumor

Discussion

Acromegaly is a systemic disorder that is characterized by GH hypersecretion and secondary increase in IGF-1 levels. It is caused by a benign pituitary adenoma in most cases. In 0.5%, it is due to an ectopic source, including ectopic GH, hypothalamic GHRH-secreting tumor or ectopic peripheral GHRH hypersecretion (pancreatic or bronchial carcinoid tumor).[2,3]

Empty Sella (ES) is an anatomical and radiological condition, describes a Sella turcia that is filled (partially or completely) with cerebrospinal fluid mainly due herniation of subarachnoid space within the Sella, which is often associated with some degree of flattening of the pituitary gland.[2] It can be classified as primary (no prior pituitary disease) or secondary (due to surgery, radiation, medical treatment, pituitary apoplexy or autoimmune hypophysitis).[4]

Pituitary function is generally preserved in primary ES, while 30% of patients have partial-to-complete hypopituitarism. GH deficiency is the most prevalent hormone deficiency, affecting 35 to 61% of adult patients with ES.[4] Rarely, pituitary hyper function occurs in ES, and acromegaly combined with ES is the least common[5] and the first cases were described in the 1980s. Molitch[6] found two patients with an ES who presented with active acromegaly.[6] Bjerre[7] evaluated 23 patients with acromegaly untreated for 2-13 years, 11 patient revealed ES on CT scans.[7] In another study including 36 acromegalic patients, 3 cases had ES were described.[8] Gallardo et al. reported only three patients with acromegaly in their retrospective study involving 76 patients with ES.[9]

Acromegaly with ES was thought to be due to spontaneous remission of the GH secreting adenoma after pituitary apoplexy, but though, it is uncommon for an acromegaly-causative pituitary adenoma to evade visualization on pituitary MRI, much less for an ES alone to be present. In cases where only an ES is visualized, ectopic acromegaly should be considered.[5]

Multiple physio pathological theories have been suggested to explain the association of acromegaly and ES. The first and most common hypothesis would be an undetectable micro adenoma in the pituitary tissue lining the Sella as the source of excess GH production or silent infarction of a pituitary mass or adenoma with the development of a partial or total ES.[10] Secondly, Bjerr[7] suggested that the growth pattern of GH-secreting pituitary tumors and accompanying seller floor remodeling may induce the morphological ES aspect in patients with acromegaly. This theory was actually inspired by the fact that sellar enlargement could be the result of a paracrine role of local GH on bones. Possibly, the local sellar bone changes (paracrine GH action) may be distinct from the general bone changes (systemic/endocrine GH and IGF-1 action) observed in patients with acromegaly. The systemic changes are characterized by increased bone turnover (bone formation and bone resorption, usually resulting in increased appositional bone growth and cortical thickening) and mediated by endocrine and locally produced IGF. Finally, ES may be caused in acromegalic patients by the intra-sellar herniation of the supra-sellar subarachnoid spaces and by the sellar enlargement due to infra-sellar extension of GH-secreting adenomas.[2]

There are no guidelines for the treatment of patients with acromegaly non-visualized sellar tumors pituitary imaging, and no evidence of ectopic GH production. Therefore, in his case, repeated testing and imaging were necessary to rule out a nonpituitary etiology and to better predict a central pathology amenable to neurosurgical resection. Medical management with somatostatin analogues or transsphenoidal pituitary exploration are the reasonable treatment options for patients with acromegaly and an ES.[5,11,12]

Our patient has an ES without a history of a prior pituitary procedure or head and neck radiation. Ectopic source of GH or GHRH secretion is unlikely as there was no extra-pituitary uptake in the imaging. Therefore, we postulate she most likely has a prior pituitary micro adenoma that underwent asymptomatic infarction followed by development of a partial ES.

Acknowledgments

None.

Conflicts of interest

None.

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