

Empty sella Syndrome as a Cause of Hypopituitarism

Case study

Yousif Baha'addin Ahmed (PhD)¹

Abstract

Background: Empty sella syndrome is the radiological appearance of an enlarged or deformed sella turcica which is partially or completely filled with cerebrospinal fluid. In 20-50% of patients there may be endocrinologic dysfunction, pan hypopituitarism is present in 25% of cases and isolated one in 10%.

Objective: To present a patient in with features of hypopituitarism caused by empty sella syndrome, with related literature review.

Patients and Methods: A Case of 16 years adolescent male was presented with short stature, the height of the patient was less than 3% percentile for his age and sex, weakness, and lack of secondary sexual characters and delayed puberty due to empty sella syndrome, the patient has deficiency of all important anterior pituitary hormones in addition to corresponding target hormones, total hypopituitarism is the diagnosis and it is the cause of short stature.

Results: He has delayed puberty, the laboratory findings of the patient clearly indicate pan hypopituitarism and the MRI confirmed empty sella syndrome.

Conclusion: Empty sella syndrome can cause panhypopituitarism, and adequate pituitary imaging is required for cases with isolated pituitary hormone deficiency or panhypopituitarism.

Key words: Empty sella, Hypopituitarism, Delayed puberty.

Corresponding Author: You_xati@yahoo.com

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¹college of medicine - Hawler Medical university -Erbil -Iraq.

Introduction

Empty sella is the radiological appearance of an enlarged or deformed sella turcica which is partially or completely filled with cerebrospinal fluid. It is usually accidentally discovered on radiological study for conditions other than pituitary disease [1]. figure(1) and (2).

The incidental finding of this condition has always been seen as simple anatomical variant without great functional significance, especially in adults [2]. It can be primary when individual did not receive radiation and

not underwent surgery [3]. Secondary empty sella syndrome is used for cases caused either by pituitary adenomas undergoing spontaneous necrosis (ischemia or hemorrhage), or by infective, autoimmune, or traumatic conditions, other causes of secondary empty sella include radiotherapy, drugs, and surgery [4].

Compression of the pituitary gland may affect its function resulting in hypopituitarism [2]. Empty sella syndrome has been found in 5.5-23% of autopsies, it is

found that 20-50% of patients with empty sella syndrome have endocrinologic dysfunction, total hypopituitarism in 25% of cases and partial one in 10% [5].

While the aetiology of empty sella is not completely clear, it is currently being viewed as related to the mechanical transmission of the CSF pressure through an incomplete sellar diaphragm [6]. One of the accepted explanations is congenitally deficient sellar diaphragm being acted upon by a sustained or intermittently increased intracranial

pressure, thus promoting herniation of arachnoid membrane into sella turcica[7].

The gonadotropic and somatotropic axes are the most frequently affected [8]. Management of the condition depends on presence of the symptoms. An asymptomatic patient needs no treatment while hormonal supplementation is needed for patients with hormonal deficiency symptoms, The indications for surgery are visual disturbances and cerebrospinal rhinorrhea [9].

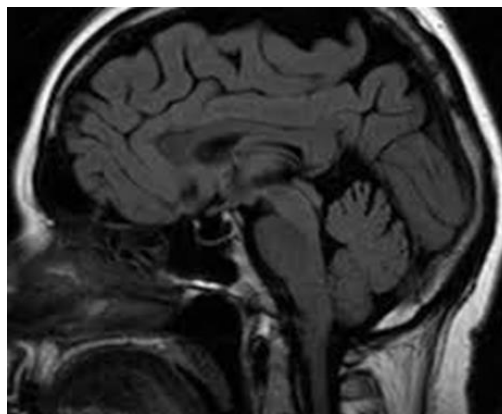


Figure (1): Pituitary MRI of a patient with empty sella syndrome.

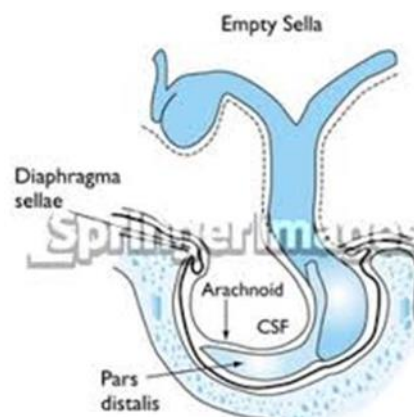


Figure (2): Sella Turcica, Bone function site.

Case presentation

A sixteen and a half years old boy presented to private endocrine consultation in Erbil city at 25th August, 2017, complaining of short stature, delayed puberty and poor weight gain, The parents denied presence of any chronic diseases or previous accidents , he has no history of trauma with no previous hospital admissions, the boy had no history of diarrhea, cough or fever.

Initial physical examination revealed a pale, thin and hypoactive adolescent with no

evidence of any hair on his face, his weight was 30 Kg. which is less than 3rd percentile for his age and sex, measurement of his height revealed 146 Cm which is well below 3rd percentile at this age in boys [10].

There was no pubic neither axillary hair, he has clear male genitalia but was underdeveloped with lack of secondary sexual characters which indicate delayed puberty Figure (3).



Figure (3): There is no axillary hair while he is more than 16 years old.

On that base investigations started and are illustrated in the following tables.

Table (1): Basic important investigations of the patient including biochemical and some serological tests.

Test	Result	Reference value	Notes
Hb	11.1 G/dl	11.5-16.5	Anemia
MCV	73.8 fl	75-100	Microcytic
Creatinin	0.5 mg/L	13.6-48.51	Normal
Calcium	10 mg/L	8.5-10	Normal
ALT	20.7 U/L	7-45	Normal
AST	45 U/L	10-45	Normal
S. ferritin	44 Ng/ml	24-2336	Normal
TTG. Ig.G	7.8 U/ml	<12	Normal
Anti-Gliadin-A	2.3 U/ml	<20	Normal
Anti-Gliadin-G	2.5 U/ml	<12	Normal

Table (2): Investigations of the patient showing serum levels of anterior pituitary hormones ,testosterone ,IGH-1 ,and thyroid hormones.

Tests	Results	Reference values	Notes
IGF-1	87 ng/ml	130-600	Low
Basal GH	0.059 ng/ml	0.077-10.8	Low
GH after stimulation	0.080 ng/ml	0.077-10.8	Low
S. ACTH	10.39 pg/ml	7.2-63.3	Low normal
S. cortisol, 8 am	32.87 nmol/L	171-536	Low
F T4	6.36 pmol/L	12.-22	Low
FT3	4.9 pmol/L	3.1-6.8	Normal
TSH	3.4 mIU/L	0.27-4.2	Normal
S. prolactin	22 ng/ml	3-25	Normal
FSH	0.4 mIU/ml	1.7-12	Low
LH	< 0.1 mIU/ml	1.1-7	Low
S. Testosterone	0.025 ng/ml	2.3-10.3	Low

Pituitary MRI revealed empty sella syndrome.

(Normal gray and white matter, no focal lesion noted. Normal ventricular system and CSF cistern. Normal brain stem density with clear CP angles, Empty sella is noted, no midline structure deviation . Normal posterior fossa structures)

Discussion

Primary empty sella syndrome is an incidental radiological finding and usually asymptomatic, It is more common in middle-aged obese females [11]. the patient is an adolescent male presented with features of delayed puberty according to classical criteria showed that prepubertal boys at the age of 14 years or more are defined as having delayed pubertal onset [12]. It has been mentioned that high incidence of pituitary dysfunction documented in patients with primary empty sella syndrome, as panhypopituitarism, secondary

hypogonadism, hyperprolactinemia and isolated ACTH deficiency[13] .

Presentations of combined pituitary hormone deficiency in adolescents include absent/partial puberty and short stature[14]. Hypopituitarism can be easily diagnosed by measuring basal pituitary and target hormone levels [15]. A recent study reported that a basal FSH level below 1.2. IU/liter in boys presenting with delayed puberty confirmed the diagnosis of hypogonadotropic hypogonadism with 100% specificity [16].

This patient has deficiency of all important pituitary and target hormones as listed in table (2) panhypopituitarism is the diagnosis of this case and it is the cause for presentation as short stature, in the view of absence of history of head trauma, surgery and radiation injuries combined with absence of evidence of infection, primary empty sella syndrome is the most likely cause of hypopituitarism in this boy as documented by MRI, In a study by Vikrant Ghatnatti et al which included 24 cases of primary empty

sella syndrome, 12 of them had significant hormonal abnormalities including GH deficiency [17]. In another study which is done by Wael fuad and included 20 cases of symptomatic empty sella syndrome, 35% had endocrine abnormalities[18]. The unusual points in this case of empty sella syndrome is the age of presentation of panhypopituitarism rather than isolated single hormone deficiency, other cases of symptomatic empty sella syndrome have been reported to present with headache at age of 27 years old without significant hormonal deficiencies [19], In an other, short stature was the presenting feature at age of 4 years [20].

Conclusion

Although often asymptomatic, empty sella syndrome could be a cause of hypopituitarism and mandating adequate endocrine assessment.

References

- [1] JK Agarwal, RK Sahay, SK Bhadada, Vijay Sekhar Reddy, NK Agarwal. Empty Sella Syndrome. Journal of indian academy of clinical medicine, 2001.v.3 (2) Pp:198-202.
- [2] Maurizio Poggi, Salvatore Monti, Chiara Lauri, Chiara Pascucci, Valeria Bisogni and Vincenzo Toscano. Primary empty sella and GH deficiency: prevalence and clinical implications. Ann Ist Super Sanità 2012.Vol. 48, No. 1: 91-96.
- [3]Kiran Kumar¹, Mohammed Khalid¹, Alyaa Fadhil², Pankaj Lamba³, Shaikh Altaf. Empty Sella Syndrome. ULF medical journal; 2014.3(S1):S67-S71.
- [4] Laura De Marinis, Stefania Bonadonna, Antonio Bianchi, Giulio Maira, and Andrea Giustina.. Extensive clinical experience, Primary Empty Sella. J Clin Endocrinol Metab, 2006. 90(9):5471–5477.
- [5] Hisaco komada, Masak Yamamoto, sak Okubo, Kanton Nagai, Keuii IIDA et al. A case of panhypopituitarism with empty sella syndrome: case report and review of the literature .. Endocrine journal, 2009.56 (4).585-569.
- [6]Shahid Ahmed, Nadir Ali, Shahzad Ashraf, Taimoor Ahmed. Sever Anterior hypopituitarism , rare presentation of primary empty sella syndrome. A Journal of Army Medical & Dental Corps. 2009.59 (2).
- [7]Shashank Shekhar¹, Chandrdeep Sharma¹, Mukesh Surya², Neeti Aggarwal. Isolated gonadotropin deficiency with primary empty sella: Causal or casual association?. Open Journal of Obstetrics and Gynecology, 2012. , 2, 276-278. <http://dx.doi.org/10.4236/ojog.2012.23057>. Accessed at 1.10.2017.
- [8]Paula Andujar-Plata¹, Eva Fernandez-Rodriguez¹ and Felipe F. Casanueva. Andujar-Plata et al ; An Update on Subclinical Hypopituitarism, J Steroids Horm Sci, . 2016.7:2. 2-5.
- [9] Pokhraj P. Suthar, Gaurav R. Parmar, Chinmay Trivedi, Hemen I. Vithlani. Case Report, Utility of MRI in diagnosis of empty Sella syndrome in a young female with amenorrhoea and bilateral nipple discharge. Int J Res Med Sci ; . 2017.5(8):3734-3736.
- [10]<http://www.chartsgraphsdiagrams.com/HealthCharts/growth-2-20-boys.html>. Growth

- Chart for Boys, 2 to 20 Years. Accessed on 1.12.2017.
- [11] Kiran Kumar^{1*}, Mohammed Khalid¹, Alyaa Fadhil², Pankaj Lamba³, Shaikh Altaf Basha¹. Empty Sella Syndrome: A Case Report and Literature Review. *Neuroscience & Medicine*, 6, 42-45. <http://dx.doi.org/10.4236/nm.2015.61007>. Accessed at 2.10.2017 .
- [12] Jacob Gerner Lawaetz, Casper P. Hagen, Mikkel Grunnet Mieritz, Martin Blomberg Jensen, Jørgen Holm Petersen, and Anders Juul. Evaluation of 451 Danish Boys With Delayed Puberty: Diagnostic Use of a New Puberty Nomogram and Effects of Oral Testosterone Therapy. *J Clin Endocrinol Metab*, 2015. 100(4):1376–1385.
- [13] P. Aruna • B. Sowjanya • P. Amaresh Reddy. M. Krishnamma • J. N. Naidu. Partial Empty Sella Syndrome: A Case Report and Review. *Ind J Clin Biochem* . 2014.29(2):253–256
- [14] Andrew A Dwyer^{1,2}, Franziska Phan-Hug^{1,3}, Michael Hauschild^{1,3}, Eglantine Elowe-Gruau^{1,3} and Nelly Pitteloud Transition in endocrinology, Hypogonadism in adolescence. *European Journal of Endocrinology* . (2015) 173, R15–R24.
- [15] Seong Yeon Kim. Diagnosis and Treatment of Hypopituitarism. *Endocrinol Metab*: 2015. 30(4): 443–455.
- [16] Jennifer Harrington and Mark R. Palmert .2012. Distinguishing Constitutional Delay of Growth and Puberty from Isolated Hypogonadotropic Hypogonadism: Critical Appraisal of Available Diagnostic Tests. *J Clin Endocrinol Metab*, September 2012, 97(9):3056–3067.
- [17] Vikrant Ghatnatti, Dipti Sarma, Uma Saikia. Empty sella syndrome – beyond being an incidental finding. *Indian Journal of Endocrinology and Metabolism* , 2017.16 .S.2. <http://www.ijem.in> on., IP: 109.127.68.37]. Accessed at 20.9.2017.
- [18] Wael Fouad . Review of empty sella syndrome and its surgical management. *Alexandria Journal of Medicine* , 2011.47, 139–147.
- [19] Sanwar Hossain¹, Mahbuba Ashrafi Mumu, Moyenuddin , Anwer Khan , Mahbuba Ashrafi Mumu, Case report: Primary Empty Sella Syndrome: A case Report. *AKMMC J* 2010; 1(1): 23-25.
- [20] Debasmita Rath, Ranjan Kumar Sahoo,¹ Jasashree Choudhury, Dillip Kumar Dash, and Anuspadana Mohapatra. Empty sella syndrome in a male child with failure to thrive. *Pediatr Neurosci*. 2015; 10(1): 45–47.