Bulletin of the National Research Centre

CASE REPORT Open Access

A rare case of pituicytoma-related hypercortisolism in a patient with Cushing syndrome—case report

Magdelene Amoateng[1*], Eiman Ibrahim[1], Yasir Ahmad[2], Anoush Calikyan[3] and Ilja Hulinsky[4]

Abstract

Background: Distinguishing between a pituitary adenoma and a pituicytoma can be challenging. Hormonal changes in pituicytomas are uncommon, and the tumor's mass effect has triggered most symptoms. There were only two reported cases of pituicytoma with elevated hormonal levels in patients who presented with Cushing syndrome as of 2017.

Case presentation: This report describes a rare case of a patient who had Cushing syndrome from a pituitary-related hypercortisolism, most likely a pituicytoma with neuroendocrine features with the benefit of early detection and surgical resection.

Conclusions: Pituicytomas have malignant tendencies while pituitary adenomas are mostly benign, hence the need to distinguish them accurately for prompt treatment and improved prognosis.

Keywords: Pituicytoma, Pituitary adenoma, Cushing syndrome, Transsphenoidal resection, Case report

Background

Pituicytoma is a low-grade glioma arising from the pituicytes in the neurohypophysis located in the posterior lobe of the pituitary or the pituitary stalk (Chang et al. 2018; Schmalisch et al. 2010). It occurs in adulthood and has a higher male predilection. It can be clinically misdiagnosed as a pituitary adenoma (Chang et al. 2018; Guo et al. 2016). Hormonal changes in pituicytoma are unusual and triggered mainly by the tumor's mass effect. However, there have been recent reports of elevated hormonal levels in such patients initially considered to have a functioning pituitary adenoma (Marco Del Pont et al. 2019). Literature review as of 2019 reported only 6 cases of pituicytoma with elevated hormonal levels in patients who presented with Cushing's disease (Li et al. 2019). The distinction, however, can be challenging. It is unclear whether the pituicytoma has endocrine secretory influence or causes a normal pituitary gland to reach hypersecretory status (Chang et al. 2018; Feng et al. 2018) and hence the focus of this discussion. There is limited literature on pituicytoma-associated hyperactivity of surrounding pituitary glandular tissue, resulting in the clinically abnormal hormone secretion in Cushing syndrome. Pituicytomas have high malignant tendencies, while most pituitary adenomas are benign. Hence the need to increase awareness for prompt management with improved prognosis.

Herein we present a 33-year-old female with multiple possible etiologies of hypertension, diabetes, hypercortisolism, and obesity who was found to have a pituitary mass consistent with a pituicytoma with a good response to treatment.

*Correspondence: damoateng7@gmail.com

[1] Department of Internal Medicine- St. Vincent's Medical Center, Quinnipiac University Frank H. Netter MD School of Medicine, 2800 Main Street, Bridgeport, CT 06606, USA
Full list of author information is available at the end of the article

Case presentation

The patient is a 33-year-old female with a medical history significant for uncontrolled hypertension, diabetes, and hyperlipidemia (diet-controlled and not on medication for preceeding conditions), osteoporosis complicated by a foot fracture after low-intensity trauma, depression, an eating disorder, cocaine abuse, and leukoplakia and papillary hyperplasia of the tongue. Her primary care physician referred her to an endocrinologist for an unrelenting weight gain of 70 pounds in one year. She gained 30 out of the 70 pounds in the month prior to the presentation. She had associated swelling and fat pads in the back of her neck, muscle weakness, joint pain, body aches, hirsutism, and easy bruising. She reported irregular menstrual periods: about 4 to 5 times per year and had never been pregnant (Table 1). She denied the use of exogenous steroids. There was a family history significant for thyroid disease in her mother, coronary artery disease in her father, and a pituitary tumor in her sister. Her examination was pertinent for cushingoid features.

Her dexamethasone suppression test indicated early morning cortisol of 17.5 mcg/dL and a serum dexamethasone concentration of 222 ng/dL. Her total 24-h urine cortisol was 336 mcg/24 h. Adrenocorticotropic hormone level was 94 pg/mL, and a chromogranin level was just above the upper limit of normal. All other pituitary hormone levels were within normal limits. Her chest X-ray was negative for any cardiopulmonary pathology. An abdominal computer tomographic scan was negative for an adrenal mass but positive for a 3.2 cm incidental liver lesion, which increased from 1.6 cm on a scan five years prior. She had a repeat dexamethasone suppression test, resulting in a cortisol level of 9.9 mcg/dL and 0.5 mcg/dL of free cortisol. The total early morning cortisol was 11.3 mcg/dL at a dexamethasone level of 461 ng/dL.

She also had a pituitary protocol contrast magnetic resonance imaging (MRI) that showed a pituitary stalk in the expected location but appeared slightly thickened, with no mass effect, normal cavernous sinus, and a non-enhancing 0.46×0.33 cm focus of low signal in the left central portion of the gland on the post-contrast sequences (Figs. 1, 2 and 3). She had a neurosurgical evaluation with transsphenoidal resection of the tumor and biopsy with an inconclusive pathology report (dense collagenous tissue with crush artifact, no evidence of adenoma, and no immunohistochemistry staining for hormones was done). She was started on Mifepristone (a cortisol receptor blocker) at 300 mg daily at incremental dosing and spironolactone at 50 mg twice daily at incremental dosing to 100 mg twice daily with close monitoring of her potassium and blood pressure. The patient had a resolution of symptoms following her medical and surgical management.

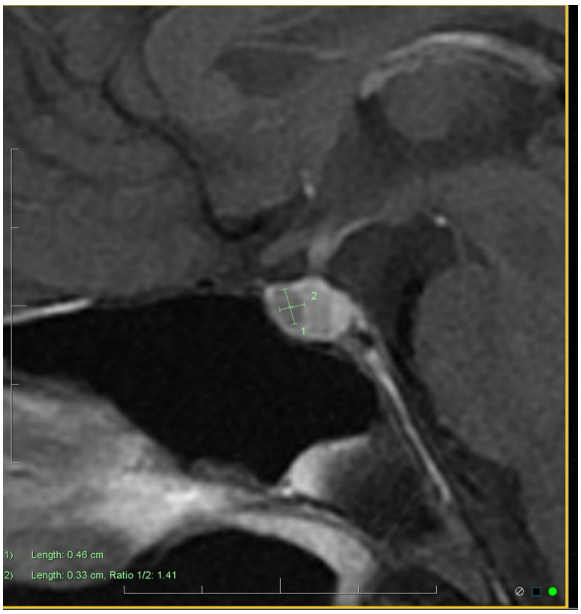

Fig. 1 MRI T1-weighted sagittal view of non-enhancing 0.46×0.33 cm focus of low signal in the left central portion of the pituitary gland on the post-contrast

Table 1 Patient characteristics

Age and sex	33yrs and female
Ethnicity/race	White
Medical history	Osteporosis at 17yrs, hypertension, diabetes, hyperlipidemia, depression, foot fracture after low-intensity trauma, an eating disorder, cocaine abuse, and papillary hyperplasia of the tongue
Medication history	Trazodone 150 mg nightly, aripiprazole 20 mg daily, fluoxetine 40 mg daily
Past surgical history	Tongue biopsy (tongue leukoplakia/papillary hyperplasia), foot and rib fracture (conservative management)
Sign and symptoms (preceding presentation)	Weight gain, fatigue, joint pain, body aches, hump in upper back and neck within 1 month of presentation, hirsutism, and easy bruising, irregular menstrual periods

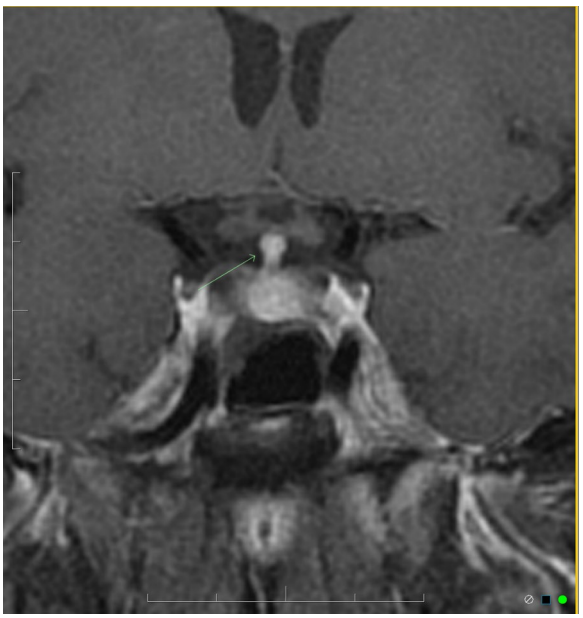

Fig. 2 MRI T1-weighted coronal view same non-enhancing 0.46 × 0.33 cm pituitary lesion

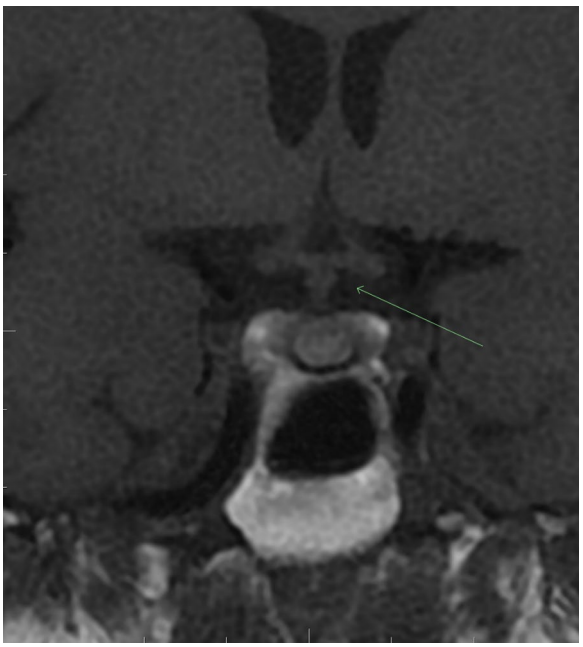

Fig. 3 MRI T1-weighted coronal view same non-enhancing 0.46 × 0.33 cm pituitary lesion and an arrow indicating the thickening of the pituitary stalk

Conclusions

It is essential to know the distinction between pituitary adenoma and pituicytoma. The former is a tumor from the cells of the anterior endocrine lobe of the gland with three main pathways of adenohypophyseal cell differentiation for hormone production: corticotrophs, somatotrophs, mammosoamtotrophs, thryotrophs, and gonadotrophs (Cheng et al. 2021b). The World Health Organization requires immunohistochemical staining for hormones produced by these cells in diagnosis (Feng et al. 2018; Gezer et al. 2019). According to the American Cancer Society, about 10,000 pituitary tumors are diagnosed annually in the USA, most of which are benign (Cheng et al. 2021b). A staggering one out of four autopsies will reveal a pituitary tumor in deceased patients who would not have known about the condition (Marco Del Pont et al. 2019; Cheng et al. 2021b).

Pituicytomas present as a neoplasm of the sellar and suprasellar regions (World health organization grade 1), which arise from the neurohypophysis or infundibulum of the pituitary and its stalk, as in this case (Marco Del Pont et al. 2019; Li et al. 2019; Gezer et al. 2019; Rumeh et al. 2020). It is a rare tumor that can mimic pituitary adenoma and can be associated with Cushing's disease (Li et al. 2019; Rumeh et al. 2020). Less than 100 cases have been reported thus far, as found through a PubMed literature review. While pituicytomas can cause a range of symptoms, often due to a mass effect, this case highlighted pituicytoma causing Cushing syndrome likely from inducing a hypersecretory effect in the surrounding pituitary gland. This is supported by the absence of evidence of adenoma in pathology. Only few other such cases have been reported in the literature (Schmalisch et al. 2010; Guo et al. 2016; Marco Del Pont et al. 2019; Li et al. 2019; Feng et al. 2018; Gezer et al. 2019; Rumeh et al. 2020; Barresi et al. 2017; Cambiaso et al. 2015; Chakraborti et al. 2013; Yang et al. 2016; Cheng et al. 2021a). In other cases reported in the literature, all adult patients achieved remission of their Cushing disease with surgical dissection of the tumor, as was done for this case.

The patient presented with rapid weight gain concerning hypercortisolism from a paraneoplastic phenomenon. Her weight gain could have been attributed to depression, Cushing's syndrome, or an eating disorder. However, her high levels of the repeated dexamethasone suppression test argued against an unlikely physiological process such as obesity, psychiatric disorders, or bulimia as the etiology of her symptoms. Of note, these conditions can cause an elevated early morning cortisol level which would be suppressible with a higher dexamethasone test. Her free cortisol level of 0.5mcg/dL following a repeat dexamethasone suppression test ruled out an abnormality with cortisol binding.

Findings from her pituitary protocol contrast MRI were consistent with pituicytoma due to the thickening of the pituitary stalk and the particular lesion location in the

infundibulum. One study describes how to use the presence of flow voids on MRI to distinguish between pituicytomas and pituitary adenomas (Law-ye et al. 2018).

One case series noted that the expanded endoscopic transsphenoidal and transplanum was preferred to the craniotomy approach and was associated with higher rates of total gross removal and lower rates of neurological complication, as was observed in our case (Feng et al. 2014). Gross surgical removal should be the intention of management.

In conclusion, pituicytomas are scarce tumors with more malignant tendencies compared to pitutary adenoma and can be associated with cushing syndrome (Feng et al. 2018). Radiological studies are often unable to provide sufficient evidence for the diagnosis of pituicytomas, as they often masquerade as pituitary adenomas. Thus, diagnosing pituicytomas remains a challenge. Surgical resection continues to be the best treatment of pituicytomas to date (Cheng et al. 2021b). Continued evidence and research into this area are needed to diagnose and treat pituicytomas more accurately.

Figures

A non-enhancing 5 mm signal abnormality within the left central portion of the pituitary gland, non-specific, most concerning for a pituitary microadenoma based on the provided clinical history. There is associated mild thickening of the pituitary stalk.

Abbreviation

MRI: Magnetic resonance imaging.

Acknowledgements

We want to thank Joshua M. Sapire MD, FCAP, and Judith Aquino for their help and arrangements.

Author contributions

MA conceived and designed the study. MA, EI, YA and AC collected and assembled the data. MA, EI, YA, AC and IH wrote the article and gave the final approval of the article. All authors have read and approved the manuscript.

Funding

There are no funding sources to report.

Availability of data and materials

All data discussed in this study are included in this article. Further inquiries can be directed to the corresponding author.

Declarations

Ethics approval and consent to participate

All personally identifiable information has been withheld, and complete patient anonymity was guaranteed. Ethical approval is not required for this case report following local guidelines.

Consent for publication

Written informed consent was obtained from the patient to publish this case report and accompanying images.

Competing interests

The authors declare that they have no competing interests.

Author details

[1]Department of Internal Medicine- St. Vincent's Medical Center, Quinnipiac University Frank H. Netter MD School of Medicine, 2800 Main Street, Bridgeport, CT 06606, USA. [2]Department of Radiology- St. Vincent's Medical Center, Quinnipiac University Frank H. Netter MD School of Medicine, Bridgeport, CT, USA. [3]Frank H. Netter School of Medicine at, Quinnipiac University, North Haven, CT, USA. [4]Department of Endocrinology- St. Vincent's Medical Center, Quinnipiac University Frank H. Netter MD School of Medicine, Bridgeport, CT, USA.

Received: 3 June 2022 Accepted: 6 July 2022
Published online: 18 July 2022

References

Barresi V, Lionti S, Messina E, Esposito F, Angileri FF, Cannavò S (2017) A 53-year-old woman with Cushing's disease and a pituitary tumor. Neuropathology 37(1):86–90. https://doi.org/10.1111/neup.12319

Cambiaso P, Amodio D, Procaccini E, Longo D, Galassi S, Camassei FD, Cappa M (2015) Pituicytoma and Cushing's disease in a 7-year-old girl: a mere coincidence? Pediatrics 136(6):e1632–e1636. https://doi.org/10.1542/peds.2015-0638

Chakraborti S, Mahadevan A, Govindan A, Sridhar K, Mohan NVS, Satish IR, Rudrappa S, Mangshetty S, Shankar SK (2013) Pituicytoma: report of three cases with review of literature. Pathol Res Pract 209(1):52–58. https://doi.org/10.1016/j.prp.2012.10.006

Chang T-W, Jung S-M, Lai H-Y, Chuang C-C, Hsu P-W, Chang C-N, Wei K-C (2018) The impact of pituicytoma on hormone secretary function. J Cancer Res Pract 5(2):47–52. https://doi.org/10.1016/j.jcrpr.2017.12.002

Cheng ML, Zhao J, Lin Y, Qiu HS (2021a) Pituicytoma: Four cases with unusual imaging features and a literature review. Neuro Endocrinol Lett 42(7):425–432 (**Epub 2021/12/01 PubMed PMID: 34847317**)

Cheng J-H, Nie D, Li B, Gui S-B, Li C-Z, Zhang Y-Z, Cavallo LM, Zhao P (2021b) Clinical features, radiological profiles, pathological features and surgical outcomes of pituicytomas: a report of 11 cases and a pooled analysis of individual patient data. Mil Med Res 8(1):39. https://doi.org/10.1186/s40779-021-00332-5

Feng M, Carmichael JD, Bonert V, Bannykh S, Mamelak AN (2014) Surgical management of pituicytomas: case series and comprehensive literature review. Pituitary 17(5):399–413. https://doi.org/10.1007/s11102-013-0515-z

Feng Z, Mao Z, Wang Z, Liao B, Zhu Y, Wang H (2020) Non-adenomatous pituitary tumours mimicking functioning pituitary adenomas. Br J Neurosurg 34(5):487–491. https://doi.org/10.1080/02688697.2018.1464121

Gezer E, Selek A, Cetinarslan B, Canturk Z, Tarkun I, Ceylan S (2019) The coexistence of infundibular pituicytoma and Cushing's disease due to pituitary adenoma: A case report. Endocr Regul 53(4):263–267. https://doi.org/10.2478/enr-2019-0026

Guo X, Fu H, Kong X, Gao L, Wang W, Ma W, Yao Y, Wang R, Xing B (2016) Pituicytoma coexisting with corticotroph hyperplasia: literature review with one case report. Medicine (baltimore) 95(10):e3062. https://doi.org/10.1097/md.0000000000003062

Law-ye B, Cholet C, Leclercq D (2018) First depiction of flow voids to differentiate pituicytomas from giant adenomas. World Neurosurg 109:304–306. https://doi.org/10.1016/j.wneu.2017.10.036

Li X, Liu Y, Miao Y, Wang J, Wang L, Wang E-H (2019) A rare case of pituicytoma presenting with severe Cushing disease: A case report and review of literature. Medicine 98(44):e17772-e. https://doi.org/10.1097/MD.0000000000017772

Marco Del Pont F, Villalonga JF, Ries-Centeno T, Arakaki N, Katz D, Cervio A (2020) Pituicytoma associated with acromegaly and cushing disease. World Neurosurg. 136:78–82. https://doi.org/10.1016/j.wneu.2019.12.085

Rumeh ASAL, Bafaqeeh M, Khairan SJA, Al SW (2020) Pituicytoma associated with Cushing's disease: a case report and literature review. J Surg Case Rep 2020(6):rjaa104-rjaa. https://doi.org/10.1093/jscr/rjaa104

Schmalisch K, Schittenhelm J, Ebner FH, Beuschlein F, Honegger J (2012) Beschorner R () Pituicytoma in a patient with Cushing's disease: case

report and review of the literature. Pituitary 15(Suppl 1):S10–S16. https://doi.org/10.1007/s11102-010-0262-3
Society AC. About Pituitary Tumors. Available at :https://www.cancer.org/content/dam/CRC/PDF/Public/8788.00.pdf
Yang X, Liu X, Li W, Chen D (2016) Pituicytoma: a report of three cases and literature review. Oncol Lett 12(5):3417–3422. https://doi.org/10.3892/ol.2016.5119

Publisher's Note