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Case report: Patient presenting with Cushing's disease

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Abstract

Backgound: Cushing's syndrome is a rare disease that is caused by the overproduction of cortisol by the adrenal glands. This can be caused by a tumor of the adrenal glands, the lungs or the pituitary gland. When a pituitary tumor produces too much ACTH (adrenocorticotropic hormone), it causes the overproduction of cortisol by the adrenal glands. When the pituitary is the source of the over production, it is called Cushing's disease.

Case Description: A 32-year-old female who developed symptoms of Cushing's about one and a half years prior to her visit at a large teaching hospital in the Mid-Atlantic. Her symptoms included amenorrhea, facial hair and acne, and back pain. She had previously been diagnosed with polycystic ovarian syndrome.

Conclusion: Cushing's disease is a rare disease, which can be successfully treated by experienced pituitary specialists.

Key Words: Adrenocorticotropic hormone, Cortisol, Cushing's

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INTRODUCTION

The Pituitary Program is an integrated program consisting of (2) Neurosurgeons and (3) Neuro-endocrinologists. They work closely with the Neuro-ophthalmologist, Neuropathologist, Otolaryngologists, Neuro-radiologists, and the Gamma Knife surgeon. Patients often come from long distances seeking treatment for their pituitary tumors. They are seen by both the Neurosurgeon and the Neuro-endocrinologist together in a joint clinic. If they need to see the Otolaryngologist or the Neuro-ophthalmologist, we can usually arrange that during the same visit.

Cushing's syndrome is a rare disease that is caused by the over production of cortisol by the adrenal glands.[2] This can be caused by a tumor of the adrenal glands, the lungs, or the pituitary gland. When the tumor produces too much ACTH, it causes over production of cortisol by the adrenal glands. If the source is the pituitary, it is called Cushing's disease.[2] Cushing's disease occurs more often in women than men and more often occurs between the ages of 20 and 40. Common features of Cushing's disease are weight gain, hypertension, diabetes, poor short-term memory, irritability, excess hair growth (women), red-ruddy face, extra fat around the neck, round face, fatigue, poor concentration, and menstrual irregularity in addition to muscle weakness. Some of the less common features include insomnia, recurrent infection, thin skin and stretch marks, easy bruising, depression, weak bones, acne, balding (women), hip and shoulder weakness, violaceous striae, hypokalemia, unexplained osteoporosis, diabetes mellitus, and swelling of feet/legs.[3] Not all patients have all of the signs and symptoms of Cushing's disease.

The diagnosis for Cushing's disease is made by laboratory testing, which demonstrates the consistent overproduction of cortisol. The tests most commonly used are midnight salivary cortisol test, a 1 mg dexamethasone suppression test, or a 24-h urine-free cortisol level.[1,2] All of these tests are approximately 92% accurate (2015, personal communication by Dr. Mary Lee Vance). A pituitary protocol magnetic resonce imaging (MRI) is also done to see if there is a visible tumor, if the blood ACTH level is detectable or elevated. Approximately 50% of patients with Cushing's disease do not have tumors which are visible on MRI.[1,3] If no obvious tumor is seen, patients may have the inferior petrosal sinus sampling test. If the pituitary is the source of the excess cortisol production, the patient should undergo surgery by an experienced pituitary neurosurgeon.

Cushing's disease is characterized by a delay in diagnosis because of overlaps with other more common medical problems such as polycystic ovarian syndrome, diabetes, obesity and high blood pressure. The average time from onset of symptoms to diagnosis is 3–5 years.[1]

CASE REPORT

The patient is a 32-year-old female who developed symptoms of Cushing's disease about one and a half years prior to her visit at the Pituitary Center in a large hospital in the Mid-Atlantic. After she stopped nursing her baby, she had no resumption of menses. She was told by her gynecologist that she had Polycystic ovary syndrome (after five successful pregnancies achieved without assistance). She then developed facial hair and acne about 6 months later. She subsequently developed back pain and was found to have a compression fracture of the lumbar spine. She saw a local physician who diagnosed her with Cushing's syndrome.

Local testing showed the following results:
Laboratory testing showed urine-free cortisol values of 228, 235, 265, and 246 (normal is less than 50).

Inferior petrosal sinus sampling was performed, which showed that the pituitary was the source of her Cushing's. Her prior MRI, done locally was interpreted as negative per the MRI report.

The patient presented to the Pituitary Clinic with this information. Upon examination, she denied changes in mood, depression, sleep disturbance, or symptoms of sleep apnea. She had not gained weight, which is typical of Cushing's but she did note redistribution of body fat. She noted enlargement of her abdomen. She continued to have some back pain. She had developed fairly profound muscle weakness and was unable to lift her 3-year-old child. She was prescribed mifepristone 300 mg BID 2 months prior to her visit at the Pituitary Center. This drug blocks the effects of cortisol. She noted her face was less round but there was no improvement in the acne, hirsutism, or other symptoms. The Neurosurgeon and Neuro-endocrinologist ordered repeat imaging using both thin cut pituitary and dynamic imaging.[4] The imaging clearly showed a 7 × 5 mm micro-adenoma on the left side of her pituitary that reached the medial wall of the left cavernous sinus but did not show evidence of invasion on the scan.

The patient was scheduled for surgery and underwent transsphenoidal resection of her pituitary micro-adenoma. After surgery, the patient was carefully monitored, measuring cortisol levels every 6 h in addition to checking urine specific gravity levels (diabetes insipidus) and sodium levels (diabetes insipidus and syndrome of inappropriate diuretic hormone).[6] Diabetes insipidus following pituitary surgery is usually transient.[5,8] She was also carefully monitored for a cerebrospinal fluid (CSF) leak. On postoperative day #1, cortisol was 2.8. There was no sign of diabetes insipidus. On postoperative day #2, morning laboratory results showed that cortisol was 1.8. She had two cortisol levels of 1.8 and on postoperative day #3, she was replaced with hydrocortisone. She did not develop any signs of diabetes insipidus or SIADH and was discharged on a dose of 40 mg of hydrocortisone each morning upon waking and 20 mg each evening between 5:00 and 6:00 pm. She was instructed to continue this dose for 3 weeks and then reduce her hydrocortisone dose to 20 mg each morning and 10 mg each evening. She was to continue this dose until 2 days before her follow up visit at which time she was instructed to hold the medication.[6] Her cortisol level would be checked at her postoperative visit to see if her pituitary was "waking up" and stimulating normal cortisol production. Before discharge, the patient was also given information regarding signs and symptoms of meningitis, Syndrome of Inappropriate Antidiuretic Hormone (SIADH), and diabetes insipidus, which can occur postoperatively. She was given instructions to lift no more than 10 pounds for 2 weeks and then not more than 25 pounds for an additional 4 weeks, no blowing her nose for 6 weeks, no submerging her head for 8 weeks, and no straining for 6 weeks. She was instructed to use saline nasal spray every hour she was awake for 2 weeks and then every 3–4 h for another 4 weeks and Neil Med Sinus Rinse three times per day for 2 weeks and then two times a day for 3 months.[7]

On postoperative day 5, the patient's father called to report the patient was having an increased headache, nausea, and two episodes of emesis. Laboratory reports were obtained and it was determined that the patient's symptoms were most likely related to cortisol withdrawal.

The patient returned to the Pituitary Clinic 8 weeks later for her follow-up appointment. She had lost 8.6 pounds. Her facial plethora had resolved and her face was thinner. Her bilateral supraclavicular fat pads were

smaller but not completely resolved. She still had some facial acne. She denied fatigue or myalgias. Patient stated she was weaker than she had been in the past but was better than before surgery. She reported her skin felt like it was burning, particularly over her legs. Patient asked about contraception and the Neuro-endocrinologist recommended that she not become pregnant for at least 6 months after her surgery in order for her to recover from the catabolic effects of Cushing's.

The patient's laboratory tests drawn at her visit showed the following results:

- Cortisol normal at 7.2 after holding hydrocortisone for 2 days and her ACTH was normal at 15. This showed that her own pituitary gland was "waking up" and she could stop taking the hydrocortisone. Her thyroid tests were normal, FT4 at 1.31 and the thyroid-stimulating hormone (TSH) 1.14. Her testosterone level was normal at 34.

Patient was successfully treated with surgery and achieved remission from her Cushing's disease. She will be monitored for recurrence the rest of her life.

DISCUSSION

Cushing's is a challenging disease to diagnose. The diagnosis is often delayed because Cushing's is frequently masked by its overlap with more common medical problems such as diabetes, high blood pressure, obesity, and polycystic ovary syndrome. Cushing's may be more common than previously thought. In this case, a 32-year-old female was ill for at least one and a half years before diagnosis. We have seen other people who were not diagnosed with Cushing's for many years. Some patients exhibit very few symptoms clinically but have testing, which confirms Cushing's. Other patients have many of the clinical symptoms of Cushing's and are very ill by the time they are diagnosed. Because of the damage hypercortisolism does to the body including muscles, joints, and bones, recovery is often painful and challenging.

CONCLUSION

Cushing's is difficult to diagnose and increases morbidity and mortality in patients who are untreated.[1]

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