Henry Ford Hospital Medical Journal

Volume 39
Number 1 *Festschrift: In Honor of Raymond C. Mellinger*

Article 2

3-1991

The Phenomenology of Cushing's Syndrome: One Patient's Account

Andrew Armstrong

J. David Fachnie

The Phenomenology of Cushing's Syndrome: One Patient's Account

Andrew Armstrong, MSW*

Introduction, Case Report, and Comment by J. David Fachnie, MD†

Cushing's syndrome caused by ectopic secretion of adrenocorticotropic hormone (ACTH) is often a serious disease and a diagnostic dilemma. In the reported patient, the source of ACTH proved to be a benign pulmonary carcinoid tumor. The patient describes his trying experiences through the six months from initial diagnosis to definitive therapy. (Henry Ford Hosp Med J 1991;39:8-9)

The importance of neuropsychiatric manifestations of Cushing's syndrome was recognized by Harvey Cushing (1) who stated in 1912 that in pituitary hyperfunction "certain temperamental changes are often apparent, with wakefulness, lack of concentration, indecisiveness, irritability, distrust and so on." Other reports of neuropsychiatric changes attest to this high prevalence—a well-known medical text noted that of 450 patients with Cushing's syndrome, 46% experienced such changes (2). Depression is the most common personality disorder (3). Recent observers, applying standardized test instruments, found in addition varying degrees of diffuse bilateral cerebral dysfunction, especially affecting nonverbal, visual-ideational, and visual memory (4).

This paper combines a medical case report with a personal report by the patient of his experience having Cushing's syndrome. The patient is a social worker who received treatment at Henry Ford Hospital. As his endocrinologist, I was deeply moved when he described the extent to which the disease had crippled his psyche. I urged him to write about the experience so that others, especially the afflicted or their physicians, might obtain a greater appreciation for the phenomenology of Cushing's syndrome.

Case Report

A 45-year-old social worker, married and the father of two children, was well until April 1989. At that time he began to notice generalized fatigue, weakness of lower extremities, swelling that was generalized but especially marked over the face and abdomen, insomnia, crying spells, irritability, lack of sexual interest, easy bruising, and slow healing of minor wounds. His alcoholism was in remission. His father had died of lung cancer, but no endocrine tumors were reported in the family history. Physical examination revealed obesity (weight 102 kg [224 lb], height 183 cm [75.2 in]), normal blood pressure (110/74 mm Hg), moon facies, buffalo hump, and melanotic hyperpigmentation.

His local physician suspected Cushing's syndrome and studies revealed markedly elevated 24-hour urinary cortisol (12,051 nmol [4,368 μg]). On the second day of low-dose dexamethasone administration (0.5 mg every 6 hours) urinary cortisol was 7,201 nmol (2,610 μg), and on the second day of high-dose dexamethasone (2.0 mg every 6 hours) the value was 3,178 nmol (1,152 μg). The patient was referred to us for further diagnostic studies and treatment.

The clinical and biochemical data suggested the presence of a severe disease, likely caused by ectopic secretion of adrenocorticotropic hormone (ACTH). The dramatic elevation of urinary cortisol was confirmed (10,788 nmol [3,910 μg]), but serum ACTH concentration was only moderately elevated (65 and 30 pmol/L [295 and 135 pg/mL]; reference range 8 to 29 pmol/L [36 to 130 pg/mL]). Computed tomography (CT) revealed no pituitary or adrenal lesions, but a 1 cm nodule was seen in the lower lobe of the left lung. The nodule had a benign appearance with punctate central calcifications, and the Pulmonary Disease and Radiology consultants suggested that it did not require immediate surgical treatment. Accordingly, we sought additional evidence that the disease did not originate in the pituitary gland. The initial petrosal sinus study failed because of severe venous spasm. Subsequent successful petrosal sinus catheterization, with sample collection before and after administration of corticotropin-releasing factor (CRF), produced data consistent with ectopic secretion of ACTH. There was no central/peripheral ACTH gradient and no response to CRF administration. Ketoconazole was administered to reduce cortisol production in preparation for lung surgery.

Surgical treatment was delayed by the patient's significant upper respiratory infection, but on October 17, 1989, the left lower lobe was removed with a carcinoid tumor that stained for ACTH. The following day, the patient's facial plethora was markedly diminished, serum cortisol was 14 nmol/L (0.5 μg/dL), and ACTH less than 1 pmol/L (< 5 pg/mL). Replacement hydrocortisone treatment was required.

Patient Account

I am writing about my disease after it has been treated, I hope successfully. The effects of the disease were gradual, cumulative, and seemed to develop in stages.

In the spring of 1989, I first experienced heart palpitations, fluid retention, occasional sexual impotence, and a "tennis elbow." My moods became unpredictable, and I had periods where I couldn't sleep. These changes were unexplained, but were mild and responded to simple psychologic remedies such as relaxation and the support of an Alcoholics Anonymous

Submitted for publication: March 6, 1990.
Accepted for publication: September 28, 1990.

*Mr. Armstrong is a social worker in Flint, MI.
†Division of Endocrinology and Metabolism, Henry Ford Hospital.
Address correspondence to Mr. Armstrong, 2307 Radcliffe Ave, Flint, MI 48903, or to Dr. Fachnie, Division of Endocrinology and Metabolism, Henry Ford Hospital, 2799 W Grand Blvd, Detroit, MI 48202.

group. Changes occurred in my social relationships, especially with family; conflicts between us intensified. In the midst of these changes, I developed an increased interest in the spiritual dimensions of life; prayer and meditation became more and more important to me. Finally, an episode of severe heart palpitations after a large evening meal brought me to a physician. Among other things, he noticed my fluid retention and asked me to restrict dietary salt. Laboratory tests confirmed the diagnosis: Cushing's syndrome.

Thereafter I began to appreciate that some of my unusual experiences were due to the disease. Accordingly, when I presented to the doctors at Henry Ford Hospital, I described the symptoms which I attributed to this strange disease: weight gain, bloating, excessive perspiration, rapid fatigue, and loss of sexual interest. The physicians observed a hump at the back of my neck, something neither my wife nor I had noticed. Their questions probed into my experience and I recognized that my usual state of psychic equilibrium was rapidly deteriorating. I was becoming more irritable, more hypomanic. I could not stay asleep at night and awakened at 2 AM. Surprisingly, I could be quite productive at this hour. I read journals, did paperwork, reflected and meditated. I became more emotionally sensitive and turned to a book of daily readings titled *God Calling*. I searched for meaning in my suffering.

About the end of July began the stage of my illness that I can best describe as a descent into hell. This began with the decision that the nodule in my lung was calcified and the doctor would have to look elsewhere for the cause of Cushing's syndrome. This decision was difficult to accept. We had traveled to the hospital many times before with the expectation that we would be given a conclusive decision about the condition and its treatment. I cannot list all the trials and delays in diagnosis which I underwent. On five separate occasions we had expected to be told the cause of Cushing's syndrome and given a definite plan for treatment, but then we were told that more tests were necessary. After we were told that the cause might not be in the lung, there began a more serious effort to study the pituitary gland.

The first catheterization study of the pituitary failed because a vein closed. There were delays in getting the second study done, but it was successful. Although initial results looked as though the pituitary might be the cause, subsequently it appeared that the pituitary was not at fault. Yet another CT of the pituitary, chest, and adrenals took more time. Finally, the decision was reached that the facts indicated ectopic Cushing's syndrome, and because no other cause was found, the nodule in the lung should be removed after all. Surgery scheduled for September was once again delayed when I developed a lung infection.

During this time, the fatigue, weight gain, melanosis, and the sense of overall loss of function progressed. I felt as if my body was simply running down. A major concern to me was that if we had to wait until late October, I might be too sick to come through surgery. I was receiving ketoconazole, a treatment to decrease my blood cortisol, but there were difficulties in adjusting the medication. I experienced extreme fatigue when my blood cortisol actually dropped below normal and I had to be given prednisone. For a brief period the cortisol levels rose again above normal and I experienced a return of the excessive energy levels. However, I was so physically depleted I was unable to translate this energy into action.

As a result of the progressive sickness, the frustrating delays, and the fluctuating hormone levels, I experienced extreme mood swings. During one period of depression I had thoughts of suicide. I felt helpless. As I became sicker and there was no definite date for surgery, I worried that I might not recover. During the period of deepest depression, I resigned myself to the fact that if I were to die from this disease, I needed to trust in the will of my God. With that decision, I experienced a sense of peace.

Finally we moved into actual treatment. I believe that the decisions that I made, to use the psychological techniques of cognitive-behavioral approaches and relaxation training, along with my awareness of the cause of the mood swings, helped me to focus my energies on recovery. The emotional support of numerous get-well cards, the encouragement of my employer who assured me that I would receive full pay during my illness, the awareness that many people were praying for me, and the fact that my family was able to sustain itself during this time of great stress were contributing factors to my recovery. My belief that this suffering would have meaning for my life and that I would be able to use this illness to assist me in my calling enabled me to focus my energies on recovering from the surgery.

After surgery I experienced a rapid leveling of moods. I found that I could identify when my blood cortisol was at an appropriate level. I saw the state of my family, wife, and children return to normal. Marital communication improved.

I am disappointed that recovery from the disease is going to take so long. I continue to feel residual pain from the surgery, but there are many pluses: the return of more normal thinking, greater emotional stability, the ability to resume working again in the near future, renewed marital closeness, and a sense of hope. I am grateful to be better.

Comment

In this phenomenologic account, Andrew Armstrong, MSW, unwraps the black box of patient experience in the Cushing's syndrome. The account allows the reader to experience vicariously the tortures of the disease. Physicians were tantalized by the severity of the disease and the clear diagnosis—ectopic ACTH syndrome. Yet they found it difficult to decide about surgical treatment because the evident cause, a pulmonary nodule, looked innocuous. Mr. Armstrong was beset by the demons of hypercortisolism: physical and psychic deformity. He suffered further from a seemingly interminable diagnostic witch hunt. Fortunately, he was cured when his pulmonary carcinoid tumor was removed. We are indebted to him for sharing his perceptions with us.

References

1. Cushing H. The pituitary body and its disorders. Philadelphia: JB Lippincott, 1912:270.

2. Soffer LJ, Dorfman RI, Gabrilove JL. The human adrenal gland. New York: Lea & Febiger, 1961:456.

3. Cohen SI. Cushing's syndrome: A psychiatric study of 29 patients. Br J Psychiatry 1980;136:120-4.

4. Whelan TB, Schteingart DE, Starkman MN, Smith A. Neuropsychological deficits in Cushing's syndrome. J Nerv Ment Dis 1980;168:753-7.