Munchausen Sickle Cell Painful Crisis*

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ABSTRACT

A 22-year-old female with factitious sickle cell anemia and recurrent painful crises is described. Because she had sickle cell trait and iron deficiency anemia, she could successfully feign the symptoms of homozygous sickle cell anemia. The identification of this syndrome in patients with genetic disorders is presented.

Introduction

The term "Munchausen syndrome" was coined by Asher² for Baron Hieronymus Karl Friedrich von Munchausen, a storyteller of alleged heroic deeds in the German cavalry. Patients with this syndrome deceive physicians by consciously simulating signs and symptoms of physical illness for no obvious gain. A hallmark of Munchausen syndrome is a tendency for patients to give a fabricated but plausible history and seek out care by wandering from hospital to hospital. Lindenbaum⁷ described two patients, with hemoglobin Munchausen, who fabricated the signs and symptoms of sickle cell anemia on numerous occasions but were found to have normal hemoglobin on repeated electrophoresis. Fishbain et al⁵ described Munchausen syndrome in a 35-year-old white female presenting with chronic pain and alerted clinicians to the possibility of the occurrence of this diagnosis within the chronic pain population. The purpose of this paper is to describe a more subtle presentation of this syndrome. Our patient, who presented herself with recurrent painful crises, had sickle cell trait and severe iron deficiency anemia that required, at one time, blood transfusion. The combination of these events made it difficult to unmask the features of Munchausen syndrome in this patient and required time and effort to pinpoint the diagnosis.

Case Report

A 22-year-old single American black female was first seen in our sickle cell center to rule out sickle cell disease because she presented to the Emergency Room with abdominal pain and was found to have anemia. There was no history of nausea, vomiting, hematemesis, diarrhea, tarry bowel movements or hematochezia. She did give a history of heavy menstrual periods each lasting five days and requiring three pads daily. Family history was significant in that her nephew and cousin had sickle cell disease and that the latter died at age 18 years of complications of sickle cell anemia. She also mentioned that her father had abdominal pain caused by an "intestinal disease," the nature of which she did not know. Detailed past history showed no suggestion of painful crisis or other complications of sickle cell disease. There was no his-

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tory of blood transfusion. She used to be an avid basketball and softball player in her teens and suffered a few twisting-type injuries in the past.

Physical exam showed mild tenderness in the left upper quadrant but otherwise was noncontributory. Stools were guaiac negative. Lab data showed hemoglobin = 8.0 g per dl, hematocrit = 27.5 percent, mean corpuscular volume = 69 fl, mean corpuscular hemoglobin concentration = 29 g per dl, reticulocyte count = 10.6 percent, white blood count 6500 per µl with normal differential, and platelet count = 390,000 per µl. Examination of the peripheral smear showed microcytosis and hypochromia, and no sickle cells were observed. Serum iron was 10 µg percent, total iron binding capacity was 390 µg percent, and serum ferritin was 2.3 ng per ml. Sequential multiple analyses of serum (SMA-12) were all within normal limits. Hemoglobin electrophoresis on cellulose acetate (pH 8.6) and on citrate agar (pH 6.2) showed Hb AS pattern. Her Hb A2 was 2.6 percent and Hb F was <2.0 percent of total hemoglobin.

Based on these findings, the diagnosis of sickle cell trait and iron deficiency anemia was made, and she was started on ferrous sulfate replacement therapy. She did not tolerate this treatment which caused epigastric discomfort with nausea and vomiting. Trials with other iron preparations were equally unsuccessful, and her Hb dropped to 7.0 g per dl. Because of this, she was given IV iron dextran to which she responded with an increase in her Hb to 12.1 g per dl and Hct to 36.5 percent.

While the investigation was continuing and her iron deficiency anemia being treated, she continued to complain of recurrent attacks of abdominal pain and started to go to the Emergency Room about once a month where she was treated with narcotic analgesics. Her abdominal pain was investigated thoroughly with barium enema, upper GI series, ultrasonography of her abdomen and pelvis, liver/spleen scan, intravenous pyelogram, chest x-rays, and obstruction series. All these studies were reported within normal limits and there was no evidence of cholelithiasis, peptic ulcer disease, renal abnormalities, or functional asplenia. Her bones showed no evidence of changes usually seen in patients with sickle cell disease. The diagnosis of acute intermittent porphyria was ruled out by finding normal levels of porphobilinogen in her urine. Because of these negative findings it was thought that she had a variant of irritable bowel syndrome, and a counsellor was assigned to her for follow-up as an outpatient with poor compliance.

She continued to go to the Emergency Room about once per month requesting treatment with meperidine and was admitted once to the Hematology Service with the diagnosis of sickle cell painful crisis. During that admission, it became apparent that she was telling the Emergency Room physicians and the housestaff that she had sickle cell anemia and was regularly followed in the sickle cell center. At that point she was told that she did not have sickle cell anemia but had sickle cell trait plus iron deficiency anemia and irritable bowel syndrome; that her iron status had to be monitored; and that she must comply with her appointments with her counselor. She left the hospital the next day and was never seen again in our facility.

Apparently, she continued going to other emergency rooms and hospitals in the area, saying she had sickle cell anemia and used to be enrolled in our sickle cell program. Indeed, some physicians called our facility to verify her diagnosis. In one instance it was learned that she presented with diffuse bone pain and abdominal pain with a hemoglobin level of 7.9 g per dl. She was treated with oxygen therapy, intravenous fluid, and blood transfusion. It also came to our attention that she was enrolled in another sickle cell program in the area where she was frequently treated in the Emergency Room for pain and occasionally admitted with the diagnosis of sickle cell painful crisis. Because of the combination of sickle cell trait, iron deficiency anemia, and blood transfusion (perhaps repeatedly), it is our suspicion that it was difficult for others to discern the accurate diagnosis. Interestingly, a consent form signed by her requesting release of her records to other facilities was never sent to us.

**Discussion**

There are three types of factitious disorders: factitious disorder with physical symptoms; factitious disorder with psychological symptoms; and atypical factitious disorder. Munchausen syndrome is a subtype of factitious disorders with physical symptoms and is, itself, subdivided into two types: prototypical and nonprototypical. Patients with the former type are usually unemployed males who drift from hospital to hospital. Patients with the nonprototypical Munchausen syndrome could be either males or females who often simulate illness and who often stay in one place for months or years. Our patient seems to fit into the nonprototypical type of Munchausen syndrome.

The choice of the symptom complex associated with the painful sickle cell crisis by our patient appears to be appropriate and effective simulation of a factitious disorder. The fact that her family history was significant for sickle cell disease and the fact that her cousin died of complications of sickle cell anemia gave her the opportunity to realize the association of severe pain with few or no objective find-
ings in patients with authentic painful crisis on physical exam. Moreover, her father had chronic intestinal disorder with abdominal pain. It is thus possible that she, subconsciously, internalized these physical aspects seen in her relatives and transformed them into an effective mechanism to simulate a factitious disorder. In addition, because she herself had sickle cell trait and iron deficiency anemia, it was easy to feign credibility of her story and give a plausible history. It is interesting to note that her iron deficiency anemia recurred soon after adequate intravenous therapy with iron and, later, after blood transfusion. We did suspect, but could not prove, that she might have, somehow, practiced self-administered phlebotomies.

Asher proposed five possible motives for this syndrome and emphasized that generally a motive in a certain patient may never be clearly ascertained: (1) to be the center of interest and attention; (2) to satisfy a grudge against physicians and hospitals by deceiving them; (3) to get narcotic drugs; (4) to escape police; and (5) to get free room and board. The motives in our patient most likely include the desire to be the center of attention and to get free room and board. The desire to get narcotic drugs (meperidine) may have been a third motive. It is believed that Munchausen syndrome is more common in countries with free medical systems. Our patient had medical assistance, and this may have encouraged her to abuse the system in the described manner. Having the diagnosis of sickle cell anemia would guarantee her continued support by the welfare and medical assistance systems. This may explain why pathological lying (pseudologica fantastica), another characteristic ascribed to Munchausen syndrome, was present in our patient.

Munchausen syndrome, in general, is a difficult diagnosis to make and even more difficult to treat. In "Hemoglobin Munchausen," however, using modern molecular biology techniques, analysis of genomic DNA, isolated from peripheral blood leukocytes, by the Southern blotting technique or by the polymerase chain reaction will differentiate hemoglobin A from sickle cell trait (AS) and from sickle cell anemia (SS), irrespective of a history of recent blood transfusions. The same methodology can be used to diagnose other inherited disorders. Patients who fabricate the signs and symptoms of an inherited disease could be checked with this modern technology and confronted with the results. This approach may limit the extent of the abuse of the health system by these patients.

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References