

Munchausen Syndrome – Presenting as Immunodeficiency A Case Report and Review of Literature

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Munchausen syndrome is one of the most intriguing of factitious disorders. Doctors working in teaching institutions and medical emergency centers are more likely to come across such patients and these patients can present with so many diverse complaints that doctors and physicians working in all fields of medicine should be aware of this entity.¹ Most clinicians will encounter at least one patient with a factitious disorder during their practice years of which Munchausen syndrome is the most extreme type. In this report, we present a case of Munchausen syndrome referred as a possible case of immune deficiency.

Case Report

A 22-year-old single female (university student) was referred to our hospital as a possible case of immune deficiency or neutrophil disorder. On admission, she had painful swelling over the right breast for five days and gave a history of similar recurrent swellings over the abdominal wall during the past few months which needed repeated surgical drainage in other hospitals on about 20 occasions. She denied a history of fever, upper or lower respiratory tract symptoms, trauma or such lesions on other parts of the body except the abdominal wall.

Her problems started at the age of 17 when she was seen for amenorrhea. She had only two cycles and then failed to menstruate. She was prescribed oral contraceptives and after a few months she developed symptoms suggestive of deep vein thrombosis in one leg. It was proven on venogram and she was started on anticoagulants; while being on warfarin, she developed recurrent deep vein thrombosis (DVT). She was again hospitalized and the dose of warfarin was gradually increased to 105 mg but still the prothrombin time and INR did not increase significantly. Even her medications were given under supervision so she was labeled as a case of warfarin resistance possibly due to poor absorption.

Soon after this, she again presented with complaints of bilateral painful swellings in the groin area associated with weakness of the lower limbs. Physical examination was consistent with bilateral femoral nerve palsy and investigations (ultrasound, computed tomographic [CT] scan) were suggestive of hematoma. Anticoagulation was stopped and surgical evacuation was carried out and she recovered well except for residual partial left femoral nerve palsy. She was also investigated for amenorrhea and it was suggested to be hypothalamic in origin but no treatment was prescribed. The patient's father is a teacher and described by her as a supportive and friendly man. Her mother had breast cancer. She has six sisters and no brothers. She is the third in line. The patient was intelligent but seemed to have only modest knowledge of the medical field. None of her near relatives are related to the medical field.

Physical examination during the current admission revealed an intelligent young female, afebrile, with normal pulse and blood pressure. There were multiple scars over the abdominal wall and there was a tender, hot and indurated area (4 x 5 cm) over the lateral aspect of the right breast. There were two small tender lymph nodes palpable in the right axilla. The rest of the examination was normal. A provisional diagnosis of cellulitis and recurrent abscesses was made. Possible immunodeficiency (?leukocyte disorder) was also considered and results of relevant investigations were as follows: CBC showed WBC 8900 (80% polys, 18% lymphs, 2% monocytes), Hb 11.1, platelets 403. Urea and electrolytes, liver function test, MSU and chest x-ray were all normal. Immune function studies: quantitative immunoglobulin levels were normal. C3, C4 and CH50 were normal. There was normal phagocytic and opsonic activity. HIV was negative.

She was treated with cloxacillin but later, surgical drainage had to be done as features of an abscess fully developed. Cultures from the abscess grew *Citrobacter* and *Pseudomonas aeruginosa* and cloxacillin was changed to gentamicin and ceftazidime. After about four days of hospitalization, the patient developed a similar lesion in the opposite breast and this had to be drained surgically as well. Cultures grew *Enterobacter cloacae* and *Enterobacter agglomerans* (sensitive only to ciprofloxacin).

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At this stage, suspicion was raised regarding the possible factitious nature of her problem because of an inability to explain the cause of her abscesses and growth of multiple organisms from the lesions.

A psychiatric consultation was made after counseling the patient without giving her a hint about suspected factitious disorder.

In the psychiatrist's view, the patient was adjudged to be very defensive and extremely rationalizing her answers. She showed a superficial affect about her problem and seemed to be under a great amount of stress because of conflicts. There were no clear features of depression. It was suggested that the patient may benefit from long term psychotherapy but with minimal chances of improvement and poor prognosis. A factitious disorder was the most likely diagnosis.

Finally, one day when the patient was not in bed, the nurses found a syringe full of fecal material along with needles. When the patient came back, one of the other patients told her about this. She became very angry and hostile and left the hospital against medical advice immediately and was lost to follow-up. A diagnosis of factitious illness (Munchausen syndrome) was confirmed.

Discussion and Review of Literature

Factitious disorders are characterized by physical or psychological symptoms that are intentionally produced or feigned in order to assume the sick role. Factitious disorders should be distinguished from malingering, also in which the symptoms are produced intentionally but the individual has an obvious goal to achieve, such as to avoid standing trial or essential services and sometimes to receive compensation. In contrast, in factitious disorder, the motivation is a psychological need to assume the sick role and there are no external incentives for the behavior.

Factitious disorder is divided into three subtypes: with predominantly psychological signs and symptoms – with predominantly physical signs and symptoms (Munchausen syndrome) – with combined psychological and physical signs and symptoms. Munchausen syndrome refers to a special type of factitious illness meeting DSM-IV criteria for chronic factitious illness with physical signs and symptoms. This term was first used by Asher in 1951.³ The characteristic presenting features⁴ of this syndrome are listed in Table 1 and there is a consensus that these features are essential in recognizing Munchausen syndrome.

A high index of suspicion is necessary whenever a patient shows any of the above features. Our patient had two (1 and 3) of three essential features while it was difficult to be sure about the second feature (peregrination) at this stage. Some of the minor features (supporting features) were also present. Even then, true diagnosis was missed for quite some time and immunodeficiency was

suspected. Neither the patient nor any of her near relatives have any medical training but she seems to have some medical knowledge because of her previous hospital admissions.

Many patients with Munchausen syndrome show features of borderline personality disorder and patients often have histories of difficult childhood relationships with parents. A history of milder degrees of abnormal illness behavior prior to the development of this syndrome may exist. Subsequent illness behavior reinforced by professionals over the years then leads to an escalation of abnormal illness behavior.⁵

Often, a true organic lesion from the past has left some genuine physical signs upon which the patient elaborates to convey a convincing story.⁶

Many patients report important childhood relationships with physicians or other parent figures who became selected objects against whom love and anger could be acted out.⁷

The three organ system modes of presentation originally reported by Asher include abdominal, hemorrhagic and neurologic. The other common modes of presentation have been reviewed by Carney and Brown, Ford, Reich and colleagues, and Nanji and Associates and includes almost every organ system of the body.^{8,11}

Buddemeyer and associates¹² reported a case of a 30-year-old white male who presented to the emergency room with complaints of pain and swelling of the calf, chest pain, intermittent hemoptysis and shortness of breath. While doing a lung scan to rule out pulmonary embolism, pre-existing radioactivity was found mainly in the lungs but also in other organs. The patient, who gave a rather dramatic story of exposure to nuclear materials in the previous weeks, had presented to several area hospitals where ventilation-perfusion scans were performed; thus,

TABLE 1. *Diagnostic features of Munchausen syndrome.*

<i>Essential features</i>	
Pathologic lying (pseudologia fantastica)	
Peregrination (traveling or wandering)	
Recurrent, feigned or simulated illness	
<i>Supporting features</i>	
Borderline and/or antisocial personality traits	
Deprivation in childhood	
Equanimity for diagnostic procedures	
Equanimity for treatments or operations	
Evidence of self-induced physical signs	
Knowledge of or experience in a medical field	
Most likely to be male	
Multiple hospitalizations	
Multiple scars (usually abdominal)	
Police record	
Unusual or dramatic presentation	

many tests in a short period resulted in the unusual findings.

An interesting case was reported by Papperney and colleagues¹³ involving a 15-1/2-year-old female who required 23 hospitalizations, 13 major surgical procedures, and numerous visits to physicians before the diagnosis was made. Perhaps the most unusual is the case of a Yemeni female reported by Karnik and colleagues.¹⁴ She presented with subcutaneous emphysema of the face and many other areas and was later found to be injecting air with hypodermic needles.

Information is very limited on the prevalence of Munchausen syndrome and it is thought to be a rare disorder. It may have been overreported because the patients often change their names and identities and present to different physicians at different hospitals. Most of the case reports in literature come from North America and Western Europe, so it seems to be more common in highly educated societies. We did not come across any case report of Munchausen syndrome from Saudi Arabia so we believe this to be the first report of this disorder. This should alert the physicians in the area and the possibility of more cases.

Ethical and legal issues regarding the measures which may have to be used for establishing the diagnosis of a factitious disorder are complex and have been discussed by many authors. Some people recommend that methods like searching the room are justified if the danger is likely due to a self-induced condition and the patient's life is at risk.¹¹

While the prognosis for Munchausen syndrome remains poor, treatment is extremely difficult as most of the patients disappear very early. Only one case has been reported as being successfully treated¹⁵ and this patient was willing to stay in the hospital for three years.

Psychiatric consultation should be requested and if confrontation is necessary, the primary physician should do this in a nonpunitive manner.¹⁶

There are many different methods described for treatment of such patients, perhaps the best treatment for Munchausen syndrome is early recognition, as dangerous diagnostic or therapeutic interventions can be avoided and attempts to help the patient can begin without delay.⁴

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