The Curious Case of a Violently Ill Woman

John H. Enterman*,1 and Dyllis van Dijk2

1Parnassia, Department Klinisch Centrum Volwassenen Gesloten, Kiwistraat 5, 2552 DH Den Haag, The Netherlands; 2Parnassia, Department Anxiety Disorders, Kiwistraat 5, 2552 DH Den Haag, The Netherlands.

*To whom correspondence should be addressed; tel: +31 703916094, fax: +31 703916133, e-mail: j.enterman@parnassia.nl

Key words: atopic eczema/beta-thalassemia/petit mal epilepsy/treatmentresistant/interaction of comorbidity

Introduction

Schizophrenia, as our readers know all too well, is a syndrome characterized mainly by the presence of psychotic symptoms that cannot be explained by an underlying disorder. It was described by Kraepelin in 1893 under the name of “Dementia Praecox,” and later, in 1908, Bleuler gave it the word “Schizophrenie.” Its etiology is uncertain,1 almost by definition, and its presentation so heterogeneous that some authors propose the use of the term Aberrant Salience syndrome.2 In some patients, one may intuit psychological mechanisms in the emergence of this disorder. In other patients, however, one senses, without being able to pinpoint exactly how, more biological underpinnings. We feel this case illustrates this second form.

Background

Violence in persons with schizophrenia has a higher incidence than in the general population, although, as the excellent review of Walsh et al concludes,3 this should not be overestimated. Causes are not generally known despite numerous hypotheses,4 but an intriguing investigation on amygdalofrontal disconnectivity has recently been published in this Bulletin.5 Unfortunately, no investigation to either prove or reject the disconnectivity hypothesis in this patient has been performed. A likely association between substance abuse and violence is of no importance here because our patient has only sporadically tried any substances. There is a longstanding association between petit mal epilepsy and violence,4,6 sometimes even necessitating surgery.7 The likelihood of our patient benefiting from surgery, however, seems small as the petit mal epilepsy was not found prior to her electroconvulsive therapy (ECT), and the period of ECT was the period in which the violent attacks were at a minimum. Medication that may have an effect on violence was given, including olanzapine, clozapine,8 and valproate.9 Medication alone did not seem to affect the violent outbursts. The most effective antiviolent treatment for our patient appeared to be the combination of clozapine and ECT.

Not only does there seem to be a genetic link between beta-thalassemia and schizophrenia,10 there also seems to be a heightened propensity to psychopathology for patients with beta-thalassemia.11 Indeed, some authors suppose a link between beta-thalassemia and violence,12 although a recent study could not find a significant association.13 Other intriguing leads are found in the association of metabolism of polyunsaturated fatty acids with both atopic eczema and schizophrenia.14 There may be a role for antipsychotic medication both in the treatment of the associated pruritus and perhaps also of the eczema itself.15 Unfortunately, we have not seen much influence, either positive or negative, from the medication.

Case Report

Our patient is a tall, 30-year-old woman of Caribbean descent. She has beta-thalassemia and severe atopic eczema on arms and legs and episodically on the torso as well. She is an only child. Mother is a physiotherapist, and father is a realtor. Our patients’ parents separated when she was about 1 or 2 years because father did not wish to accept his parental responsibilities: he would go out on the town when she was only a few months old, leaving her alone in the house. Nothing is known about other relatives. Already as a young girl, our patient had behavioral problems: she could become unreasonably angry and physically assault fellow schoolchildren, once even with scissors. At the age of 12, she had gotten into such a fight with her mother that the latter felt compelled to call in the police. She has a normal intelligence and completed 3 years of secondary education. She worked at odd jobs between psychiatric admissions and would have subsided into a homeless existence but for the continuing support of her mother. She developed prominent religious feelings.
and devoted much of her meager income and plentiful time to a denomination with views not shared by the greater part of society. Her first admission, of 10 days to a psychiatric hospital in Surinam (Dutch Guyana), with auditory and visual hallucinations occurred at the age of 16. She had developed a paranoid hostility toward the world at large and demonstrated a belief in God that verged on the delusional. This was repeated in her first admission in the Netherlands 4 years later. She expressed a wish to live according to biblical standards and would live exclusively on milk and honey. In other ways, she was strangely uninhibited, and would occasionally walk the ward corridors naked, or, when using her free pass privileges, she would expose her body to young boys, using explicit language. These behaviors would distress her when the psychosis had subsided and she reverted to her normal shy and modest self. During her psychotic episodes, her atopic eczema would not only worsen as a result of a morbid fear of germs and the abundant washing this entailed, she would inexplicably refuse treatment. She would have no awareness of illness and would justify her hostility as a reaction to perceived discrimination, deduced from simple looks of ward staff or from refusals of impossible requests. During admissions, she would unexpectedly use extreme violence toward ward personnel, perceived as unprovoked. Thus she once assaulted a nurse with a knife, resulting in a suspended sentence for attempted manslaughter; about a year later, she kicked a nurse so severely as to render her incapable for work during several months. She also tried to hit a fellow patient on the head with an earthenware plate, but this was prevented by timely intervention of the staff. When we confronted her with her actions, she would profess to have no knowledge of these events, yet 1 or 2 moments later, she would affirm her right to violence on the grounds of having been provoked by a chance word or glance. In conversation, however, she could be at times quite charming and display sensitivity to not only her own situation but also to the feelings of others. Yet, she remained aloof and withdrawn and did not form closer contacts with fellow patients or ward staff. Both conventional and second-generation antipsychotics were insufficient to alleviate her symptoms, even though they were consistently taken (supervised by ward staff) in sufficiently high doses over a sufficient length of time. Thus, we started her on clozapine. This resulted in a sharp decline in her granulocyte count and was discontinued, especially after a retreat of 4 weeks later met with the same result. After the violence toward others occurred on our ward, we felt it was of psychotic origin and thus attempted a maximal antipsychotic treatment. We used ECT in conjunction with olanzapine 20 mg/day. When this did not quell the violence, we conferred with a specialist in internal medicine and very carefully and slowly started with clozapine up to a normal blood level. Accepting lower than usual limits for white blood cell counts allowed us to continue with the clozapine. Under clozapine and ECT the violence diminished and disappeared, her hostility also lessened markedly. Her eczema persisted, and dermatological consultations resulted only in temporary lessening of symptoms since she could not be persuaded to a lengthier treatment adherence. Her aloofness remained, even toward her mother. Psychological interventions, including cognitive behavioral therapy, were repeatedly instigated but proved impractical because it was impossible for her to reflect on her own actions or to agree on a focus for treatment long enough to warrant expending her own energy. On the ward, we perceived episodes of unresponsiveness to simple requests such as turning down the volume of her radio: she appeared not to have heard the question. Interpreting these episodes as absences, we requested an electro-encephalogram (EEG). This confirmed the existence of petit mal episodes. On valproate (blood levels above 0.50 mg/l), these episodes disappeared, but her violent outbursts did not. These last diminished under ECT. As in our first Curious Case, the energy level required to elicit a seizure at ECT was not changed by the addition of an anti-epileptic. In spite of all our efforts, the patient did not attain to anything near her premorbid levels of functioning but remained withdrawn, aloof, and with multiple negative symptoms. She was able to be around others in the absence of her violent spells and free of her petit mal episodes. She was not content with her continued hospitalization, yet unable to take any steps requiring initiative. Ultimately, she has been transferred to a long-stay ward and lives on the grounds of our institute, where she has gained considerable weight.

Considerations

In our opinion, her long history of violent episodes gives her case report a decidedly “biologic feel,” all the more so because of her severe atopic eczema, her beta-thalassemia, and her petit mal epilepsy by themselves also constitute severe afflictions in their own rights. As stated above, there are many intriguing theoretical links between her psychosis and her violent outbursts, her atopic eczema, her beta-thalassemia and her petit mal epilepsy. We have not been able to influence her beta-thalassemia, hardly her eczema, and her psychosis and have gone to extreme lengths to curb her violence. Her petit mal attacks, which had only been diagnosed long after the start of her treatments, and thus did not seem linked to her violence, have responded well to the addition of valproate to her medication regimen. Yet her aloofness, at times slipping into downright hostility, her unawareness of illness, her absence of introspection, together with her perceptibly normal intelligence, her charming smile, and her despair at her prolonged hospitalization combine to produce the feeling of our having failed in treatment.
Questions

Have we overlooked essential elements in diagnosis or treatment? In retrospect, our patient would probably have warranted a brain scan and more extensive EEG evaluation. However, would there be findings that would justify psychosurgical interventions? Would there be more to the combination of atopic eczema and schizophrenia than is justified by the paucity of publications? Should we look for an association between schizophrenia and petit mal19,20? Or rather look for a clinical link between beta-thalassemia and schizophrenia11 and How does the occurrence of petit mal in this patient reflect on the ECT?

Commentary of Our Previous Curious Case

We received questions from A.S. Bassett, MD, from Toronto University, as to whether we had considered16: (1) Parkinson disease (very early onset), (2) 22q11.2 deletion syndrome, and (3) hypocalcemia or other metabolic condition or Parkinson disease, though she had not seen mention of any particular features in this regard. Also, (4) was there any helpful family history for this patient?

To answer the Commentary questions

(1) Parkinson disease. No, we had not considered this. Even with hindsight, however, there are no symptoms other than the mutism and immobility that would point us toward this diagnosis. There was no rigidity, pill rolling, propulsion phenomena, or other signs of Parkinson disease, and there was a very significant fluctuation of severity, which also decreases the likelihood.

(2) 22q11.2 deletion syndrome. There were no features pointing in the direction of this syndrome, with the self-evident exception of psychiatric disorders, and perhaps dental problems and medical history, both of which are not well known to us. However, there were no dysmorphic facial features, no developmental and learning disabilities, and no hypernasal speech. On the basis of these absences, we judge the chances of this relatively rare syndrome being present in our subject to be small indeed.

(3) Other metabolic conditions were judged to be absent on account of normal hematological and blood chemistry lab results.

(4) The family history was not known beyond first- and second-degree relatives and offered no new insights.

We wish to thank Ms Bassett for her time and comments, which have proven immediately helpful in our regarding other patients in our hospital, although they unfortunately were not able to further our understanding of the patient we had presented.

Ms Bassett kindly supplied us with relevant literature.21,22

Acknowledgments

The authors have declared that there are no conflicts of interest in relation to the subject of this study.

References


