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*Thomas A. Fahy, MB, BCh, BAO, *Registrar in Psychiatry, Maudsley Hospital, London*; Paul H. Robinson, MRCP, MRCPsych, *Senior Lecturer, Department of Psychological Medicine, King's College Hospital, London*; Gerard F. M. Russell, MD, FRCP, FRCP(Ed), FRCPsych, DPM, *Professor of Psychiatry, Institute of Psychiatry, London*; Brian Sheinman, MRCP, *Medical Advisor, Medical Foundation for the Care of Victims of Torture, National Temperance Hospital, London*

*Correspondence: *Maudsley Hospital, London SE5 8AZ*

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Post-Partum Psychosis in Adult GM_2 Gangliosidosis

A Case Report

Adult hexosaminidase A deficiency is a form of GM_2 gangliosidosis with autosomal recessive inheritance. Only 35 cases (mostly among Ashkenazic Jews) have been reported worldwide. Symptoms include, in a third of the cases, psychosis. A 27-year-old sufferer with no prior psychiatric history, developed a post-partum psychosis, with affective and hebephrenic components, 3 days following her first delivery. She responded to lithium within 10 days of initiating treatment; the full episode lasted 1 month. We conclude that lithium is the preferred treatment for psychosis in such adult patients, especially in light of possible long-term neurological deterioration caused by phenothiazines. Ashkenazic Jews with atypical neurological syndromes presenting with psychosis should be tested for hexosaminadase A deficiency.

Adult GM_2 gangliosidosis is a rare autosomal recessive disorder caused by a deficiency of hexosaminidase A, leading to the accumulation of GM_2 gangliosides in neurons (O'Brien, 1983). Total absence of this enzyme occurs in Tay–Sachs disease, which leads to death in late infancy. A juvenile form of GM_2 gangliosidosis, usually leading to death in the second decade, has also been diagnosed.

Hexosaminidase A deficiency was first discovered in adults by Navon *et al* (1973). Thirty cases have subsequently been reported in the literature (reviewed by Navon *et al*, 1986). The disorder is apparently inherited as an autosomal recessive gene, with the highest frequency found among Ashkenazic Jews (disease prevalence of 1:67 000 in the USA, Greenberg & Kaback, 1982; and 1:14 000 in Israel, Navon & Adam, 1985). Age of onset varies from the first to the third decades of life. Clinical manifestations also vary, even within the same family, and include pyramidal, lower-motor-neuron (especially of proximal lower limbs), and cerebellar signs, often suggesting amyotrophic lateral sclerosis, spinocerebellar, and spinal muscular atrophy-like syndromes (Argov & Navon, 1984; Navon *et al*, 1986). Sensory impairment is not found.

In 9 of 33 reported cases, psychosis developed in the course of the disease (Navon *et al*, 1986). The clinical picture usually reported is one of hebephrenic schizophrenia, with long-term decline, sometimes leading to an incapacitating dementia.

We report a case of post-partum psychosis in a woman who had earlier been diagnosed as suffering from adult GM_2 gangliosidosis.

Case report

The patient was a 27-year-old woman of Ashkenazic Jewish origin with no prior psychiatric history. She had been a sub-average student in high school, although she managed to pass the qualifying exams. Her social contacts were limited. She had difficulty physically keeping up with her friends on class outings. Since childhood she had had a speech impediment with a stutter. After high school, before the age of 20, she noticed progressive difficulty in climbing stairs. No diagnosis was made. She held a succession of jobs.

At age 25 the patient married. At this time, she underwent a routine screening test for Tay–Sachs disease. Severe deficiency of hexosaminidase A was found in the patient's serum, leucocytes, and cultured skin fibroblasts. At age 27, the patient gave birth to a boy, following her first pregnancy. The ante-partum course had been full term and uneventful, with no evidence of gestational diabetes, pre-eclampsia, or aberrations of mood or cognition. Parturition was by spontaneous vaginal delivery. The first 3 days post-partum were also uneventful, as the new mother began nursing the infant.

On day 4, the patient's mother noticed that the patient began to relate in an unfeeling, mechanical way towards the infant, and to attend to her own care in a disorganised manner. On day 5, the patient developed sleep disturbances and began to speak incomprehensibly, and to hold conversations with her deceased father. Her behaviour became increasingly bizarre, and she claimed that her child came from God. Her clinical picture continued to deteriorate, and she was admitted to our hospital on day 6.

On examination, she appeared her age and was in no physical distress, but communication with her was virtually impossible. Her vital signs were normal. Physical examination revealed an enlarged post-gravid uterus and a healing episiotomy site; otherwise, results of examination were within normal limits. Neurologically, her cranial nerves were normal except for nystagmus on leftward gaze. Motor examination revealed weakness in her proximal lower extremities, with mild atrophy. Sensory examination gave normal results. Her gait was mildly waddling. Tendon reflexes were present in her arms, but absent bilaterally in the patellas and ankles. Babinski's sign was not elicited. Primitive reflexes, such as rooting and the palmo-mental reflex, were present.

On mental-status examination, the patient was uncooperative and uncommunicative. Her actions had a restless quality and were neither directed nor co-ordinated. Her affect was labile, with alternating laughing and crying. Her speech was garbled and pressured, with sentence and word fragments, and signs of blocking. Her attention was occasionally drawn to seemingly empty corners of the room, but other evidence of hallucinations or delusions could not be elicited. She knew her name (sometimes using her maiden name), but was otherwise disoriented.

Test of electrolyte, calcium, phosphorus levels, and liver-function, and a complete blood count all gave results within normal limits. A Venereal Disease Research Laboratory test (VDRL) was negative. T_4 levels, by radioimmunoassay, and T_3 resin uptake were normal. A computerised-tomography scan of the head and an electroencephalogram revealed no abnormalities.

During the first 10 days of her hospital stay, the patient's bizarre behaviour continued. On day 10 of her stay, treatment with lithium carbonate was started, reaching by day 14 a dose of 1200 mg daily, with a serum lithium level of 0.82 mmol/litre. By day 16 she was beginning to utter complete sentences, recognised her treating physician by name, and mentioned the name of her newborn infant. On day 21 she was brought the child for the first time, and responded appropriately. On day 23, she was becoming more co-operative. On day 26 certain delusions of relationship to spiritual leaders remained, although she was oriented and conversational. By day 28 (day 31 of the illness, day 34 post-partum), these last residua resolved, and she appeared to have returned to her baseline functioning.

The patient was discharged from the hospital and maintained on lithium for 4 months, after which the treatment was discontinued. She returned to premorbid functioning, with no further evidence of psychiatric symptoms, and no progression of neurological symptoms. One year after her hospital stay she remained well.

Discussion

We have reported here a case of post-partum psychosis following first pregnancy in a woman with adult GM_2 gangliosidosis. While the literature on this rare disorder is growing, little attention has been given to the psychosis that accompanies the disease in about 30% of known cases. This is the first report of post-partum psychosis in conjunction with the disorder. The likelihood that this patient's post-partum psychosis was related to her underlying enzyme deficiency is enhanced by the relative rarity of post-partum psychosis in general; according to a recent study by Kendell *et al* (1987), the risk that a woman will be admitted to hospital within 90 days of childbirth for psychiatric causes other than depression is only 1.0 per 1000 births.

The DSM–III diagnosis for our patient would be schizophreniform disorder (American Psychiatric Association, 1980). However, as in other studies of post-partum psychosis (Brockington *et al*, 1981; Munoz, 1985), we did note affective features, such as emotional lability, in our patient.

Post-partum psychosis is usually treated with neuroleptic medication (Kane, 1985; Munoz, 1985). With our patient, we withheld this treatment because of evidence that phenothiazines promote further accumulation of gangliosides in the neurons (Lullman-Rauch, 1979). Indeed, Navon *et al* (1986) have suggested that the dementia and neurological

deterioration that have been seen to follow a period of psychotic illness in adult GM_2 gangliosidosis may be secondary to treatment with phenothiazines, which may accelerate the course of the disease.

Accordingly, proper diagnosis of this disease in the presence of psychosis is essential in order to prevent inappropriate treatment with neuroleptic medication. Any patient, especially of Ashkenazic Jewish origin presenting with psychosis and the neurological signs enumerated above, needs to be evaluated for hexosaminidase A deficiency so that neuroleptic medication may be avoided.

Our patient responded well to lithium carbonate. Whether this treatment is effective only in psychosis in the post-partum period, which as noted above has an affective component, or whether it can be expanded to other cases of psychosis in the presence of hexosaminidase deficiency, will be determined by further clinical experience.

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*P. Lichtenberg, MD, *Department of Psychiatry, Jerusalem Mental Health Center*; R. Navon, PhD, *Department of Human Genetics, Tel-Aviv University and the Chaim Sheba Medical Center*; E. Wertman, MD, *Director of Neurology*; H. Dasberg, MD, *Professor of Psychiatry, Medical Director*; B. Lerer, MD, *Director of Research, Department of Psychiatry, Jerusalem Mental Health Center*

*Correspondence: *Department of Psychiatry, Jerusalem Mental Health Center, P.O.B. 140, Jerusalem, Israel*

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Schizophrenia and Multiple Sclerosis

Distribution in Italy

The present study extended an earlier report of USA states with high levels of schizophrenia also having high levels of multiple sclerosis (MS). A high correlation ($r = 0.81$) between schizophrenia and MS rates in the districts of Italy was found.