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MANIA ASSOCIATED WITH LAURENCE MOON BIEDL SYNDROME

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Laurence Moon Biedl Bardet Syndrome, inherited in an autosomal recessive fashion, is a rare disease with a multiplicity of clinical symptoms including mental retardation, retinitis pigmentosa, obesity, short stature and hypogonadism. It was first described by Laurence and Moon in 1866 and later Bardet and Biedl in the 1920's reported additonal features of polydactyly and familial occurrence (Nyhan and Sakati, 1976). The syndrome shows great variation in expression and the pentad of clinical features is present in less than half the cases (Hamilton et al., 1984).

Psychotic behaviour has been very rarely reported to be associated with this syndrome. Three cases of schizophreniform psychosis (Weiss et al., 1981 ; Klein and Ammann, 1961) and a psychotic state similar to that usually seen in oligophrenia (Chernyakov, 1974) have been reported in literature *. We describe a case with mania.

Case report

SK, a 13 year old boy was brougbt with a two week history of abnormal behaviour. Her illness was acute in onset and without precipitation stress. His symptoms were characterized by excessive and irrelevant talk, sleep disturbance, tendency to run away from home, crying spells and threats of assault. He also complained of progressive deterioration of vision.

He was born to consanguinous parents (1st cousins) and the pregnancy and delivery were unevenful. There was no family history of mental illness and the family members did not have any of the features of Laurence-moon-Biedl Bardet Syndrome. His scholastic performance was poor and after repeated failures he discontinued his studies after reaching the fourth grade.

On examination he was short statured, obese and unkempt. His attention, concentration, orientation and memory appeared to be normal. He talked excessively and expressed grandiose ideas. He was restless, irritable, had pressure of speech, flight of ideas, elated mood and lacked insight. Physical examination revealed remnants of extra digits on both hands, small external genitalia and no hair over the pubis. His vision was 6/24 in both eyes and fundus examination revealed evidence of retinitis pigmentosa. His Intelligence Quotient (IQ) on formal testing was 61.

With antipsychotic medication his symptoms abated within two weeks. He was discharged on Trifluoperazine 15 mg/day and Chlorpromazine 50 mg/day. The antipsychotics were gradually reduced. Six months later he did not have any psychotic features and was off all antipsychotic medication.

Discussion

Psychosis has been rarely reported with Laurence Moon Biedl Bardet Syndrome. The case described presented with magic symptomatology, was brief in duration, and completely remitted suggesting an affective illness. This is in contrast to the other cases reviewed which had a schizophreniform presentation. Weiss et al. (1981) in their report raised questions about the relationship between the syndrome and the associated psychosis. They considered the possibilities of a direct relationship, a chromosomal connection and the possibility of chance association. The rarity of the association between the syndrome and the psychosis and the heterogeneity of the psychosis described would support the contention that the two conditions are not related. We would favor argument that the occurrence of Laurence Moon Biedl Bardet Syndrome and psychoses is coincidental.

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Corrigendum

The name of the book appeared in the section of "Book Review" of this journal in Vol 33, No.3 shouldbe read as :-
SOCIAL STATUS AND MENTAL HEALTH : A SOCIAL PSYCHIATRIC FIELD STUDY OF CALCUTTA by Ajita Chakraborty.