HALLGRÉN’S SYNDROME: TWO CASE REPORTS
S.CHAUDHURY, V.S.GURUNADH, G.P.SINGH, G.S.SUNDARI

Two cases of Hallgren’s syndrome presenting with retinitis pigmentosa, sensory-neural hearing loss along with schizophrenia in one patient and major depression in the other, are reported along with a brief description of this rare syndrome.

Key words: Hallgren’s syndrome, schizophrenia, depression.

INTRODUCTION

Pigmentary degenerations of the retina has been associated with congenital deafness ever since Von Graefe reported their simultaneous occurrence in 1858, an observation confirmed by Liebreich in 1861 who found retinal degeneration in 5.8% of congenitally deaf persons in Berlin (Duke-Elder, 1964). Detailed analyses of these cases were carried out by Lindenov (1945), as well as by Bell (1922) who found that 10.4% of 919 cases of retinitis pigmentosa were deaf or deaf-mute. Hallgren (1959) investigated 177 patients belonging to 102 families and established the syndrome in a genetico-statistical study. The incidence of this syndrome in Sweden was approximately 3:1,00,000 people.

The clinical appearance of the primary retinal pigmentary dystrophy is characteristic of that seen in patients with retinitis pigmentosa and is associated with night blindness. All patients suffer from a congenital neurosensory deafness. More than 90% of the clinically deaf have a gait disturbance of a vestibulo-cerebellar type, possibly due to a labyrinthine disorder, since other cerebellar signs are not generally found. There is a positive correlation between the degree of hearing loss and vestibulo-cerebellar ataxia. About 25% of patients have mental retardation and 25% have a psychosis. Hallgren’s series contained no fewer than 26 (14.7%) patients with schizophreniform psychoses. Nystagmus occurs in 10% of patients and cataract usually develops before the age of forty (Duke-Elder & Dobree, 1967; Hughes, 1977). Skeletal anomalies (short stature, genu valgum, kyphosis, club foot are occasional findings. Mortality among patients is not increased whereas fertility is greatly reduced (Gaeraets, 1969). Two cases of Hallgren’s syndrome are reported because of its rarity.

CASE REPORTS

Case 1: A 33 year old married male hailing from rural Uttar Pradesh presented with night blindness of one year duration and abnormal behavior for fifteen days. There was no past history of psychiatric illness. He had performed poorly in studies and discontinued after the third standard. His sister and a cousin also suffered a similar illness. His wife had undergone surgery for carcinoma cervix eight months ago, but had developed secondaries and he was very worried about her health.

Physical examination showed an obese individual. Ocular fundus examination revealed early changes of pigment degeneration and Lister’s perimetry showed concentric constriction of about 25-30° to 3/330 white. He was diagnosed to have retinitis pigmentosa. ENT examination with pure tone audiometry revealed congenital bilateral sensory-neural deafness (45-50 dB).

On psychiatric interview, the patient complained that for the past two months he had been hearing people always discussing about him and believed that some unknown enemies wanted to kill him and his family members. Mental status examination revealed poor personal care, lack of contact with reality and vague and wooly talk. He was anxious and depressed with third person auditory hallucinations, delusions of reference and persecution in a clear sensorium with impaired insight. Sleep was reduced and appetite was normal. Relevant investigations including skull radiograph and CT scan head were within normal limits. With a diagnosis of schizophrenia, he was treated with antipsychotics, five ECTs and psychotherapy and recovered fully.

Case 2: A 32 year old married male hailing from rural Tamil Nadu presented with night visual deficit and headache of two years duration. His cousin had a similar illness along with mental subnormality. There was no past history of psychiatric illness. He had a poor academic performance and left school after the seventh standard.

General physical and systemic examination were within normal limits. Ocular fundus examination revealed early changes of pigment degeneration. Lister’s perimetry showed concentric constriction of about 25-30° to 3/330 white. He was diagnosed to
have retinitis pigmentosa. ENT examination with pure tone audiometry revealed congenital bilateral sensory-neural hearing loss (40-50 dB).

The patient was noted to remain aloof and depressed in the ward which led to psychiatric referral. He complained of headache, forgetfulness, poor appetite, initial insomnia and early morning awakening. Mental status examination revealed poor appearance, marked psychomotor retardation, monotonous speech, extreme preoccupation with somatic symptoms, anxious and depressed mood with fleeting suicidal ideation, ideas of helplessness and hopelessness and ideas of reference.

All routine investigations including CT scan were within normal limits. Radiograph of the paranasal sinuses showed mucosal thickening of the maxillary antra. A diagnosis of major depression was made and he was treated with antidepressants, anxiolytics, 7 ECTs and psychotherapy, following which his psychiatric symptoms ameliorated.

DISCUSSION

These patients had gross nyctalopia, concentric field constriction, normal central vision, equatorial pigment degeneration, positive family history, concomitant sensory-neural deafness and psychosis on the basis of which the diagnosis of Hallgren's syndrome was made. There is general agreement that congenital deafness is psychologically damaging and may be an important causative factor in paranoid psychoses (Slater & Roth, 1969; Mahapatra, 1974). It has therefore been suggested that the psychotic behavior and mental retardation seen in 25% of patients with Hallgren's syndrome may be a nonspecific response in some patients to a double sensory handicap (Hughes, 1977). In addition, Remvig (1969) who followed up Lindenov's cases opines that in some patients, the psychosis had been precipitated by environmental stress (like our Case 1), but adds that it is also reasonable to presume an inherited predisposition to psychosis. Similar findings were reported by Pandey et al (1982).

The ocular and aural abnormalities in Hallgren's syndrome are inherited traits and no treatment is available for them. However, psychotropic drugs often have a good ameliorative effect on the psychotic episode, as was seen in our cases (Remvig, 1969; Pandey et al, 1982).

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