Indian J. Psychiat. (1981), **23**(3), 263—265

JUVENILE GENERAL PARALYSIS OF INSANE : A CASE REPORT

J. K. TRIVEDI[1], M.D. (Psychiat.)
A. M. KAR[2], M.D. (Med.)
SUNIL SRIVASTAVA[3], M.B.B.S.

Juvenile parenchymatous syphilis, or juvenile general paralysis of insane, first described by Clouston (1877) is among one of the rarer manifestations of congenital syphilis, incidence being about ½—1 percent (Stewart, 1933). The infection is transmitted to the fetus via the placenta and the incubation period is about 5-20 years. The intrauterine Spirochaetal infection of the nervous system may lead to actual developmental arrest. Gross disappearance of Purkinje cells with gliosis of the cerebellar cortex is rather characteristic of juvenile general paresis. It affects male slightly more than females. In clinical features or pathology, it does not differ in essential respects from the acquired form but the mental symptoms are less florid and positive symptoms are infrequent. The usual presentation is gradually progressive dementia usually first manifesting at the end of first decade, with which there may be apathy, forgetfulness, restlessness, temper tantrums, irritability, impulsive behaviour and antisocial acts. Euphoria, expansiveness, grandiosity, delusions, hallucinations and delirious pictures are less common than that in acquired variety. Delusions if present have a childish quality. Optic atrophy and pupillary abnormalities are frequent but pupils are often widely dilated and fixed rather than typical Argyll Robertson pupil. Most Indian studies relating to congenital syphilis are confined to children up to five years of age (Benakappa *et al.*, 1978 ; Mehta and Gharpure, 1979), therefore there is dearth of Indian studies regarding this illness.

Case Report :

N. S. 15 years old Hindu male was third live child in parity order. It was a full term normal delivery. Soon after birth he developed umblical sepsis, in which he had one convulsion during high fever. Sepsis was cured after a few days treatment. Milestones were slightly delayed. Delay was more marked in the area of speech. He could speak one or two words first time at the age of three years. Scholastic performance of the child was below average. He was weak in studies. Once he failed in class 6th, then in class 7th, in spite of his best efforts. For his poor performance he was frequently punished, therefore he started running away from classes. Finally he was taken off from school when he was in class 8th.

At the age of 14 years he sustained head injury when he fell from bicycle. Bleeding was profuse but consciousness was retained. He was seen immediately by a doctor. There was no evidence of fracture. He was given primary care and was well within a week. Three months later it was noticed that he had become forgetful. He used to forget way back to home, assigned works and instructions. It was followed by progressive motor weakness and unstability while walking. His comprehension deteriorated. He could not follow instructions unless explained repeatedly and given in

1. Lecturer, Department of Psychiatry
2. Reader Department of Neurology
3. Senior Resident, Department of Psychiatry
K. G.'s Medical College, Lucknow.

steps. Even in daily routine patient needed assistance at every step like in bathing, dressing, brushing teeth and eating. The speech changed from normal clearly comprehensible speech to hesitant, interrupted, explosive, lalling, slurred speech, mostly of monosyllables. There was disturbance of sleep too. Total sleep time was of three to four hours. He got sleep late, woke up early and it was interrupted. This behaviour was punctuated by episodes of confusion, disorientation, restlessness and perplexity lasting for few hours. Along with these changes, patient developed abnormal movements of tongue, which he used to protrude out suddenly repetitively; mastication like movements of mouth, purposeless jerky sudden repetitive movements of upper limbs and coarse tremors of fingers. All these movements used to disappear during sleep. Gradually the patient became withdrawn and apathetic, remained silent mostly, did not show interest in any work and lost his appetite. At that stage he was treated by a doctor who put him on vitamins by which some improvement in appetite occurred but otherwise the symptoms remained unaffected. Course of the illness was gradually progressive.

Patient came from a nuclear family where he lived with parents and one younger brother. His two elder brothers had died in infancy for which no cause could be established. Third sib was a still birth. Younger brother was of seven years and was mentally and physically healthy. There was history of schizophrenic psychosis in patient's real uncle. Father had history of premarital heterosexual contact at the age of 25 years. It was followed by development of generalised syphilitic rash which subsided after about four months by itself.

Before the illness, N. S. was of dominantly schizoid temperament.

In physical examination there was generalised wasting and weakness along with flat foot and genu valgus. Gait was clumpsy, unstable with slight limp on right side. Pupils were widely dilated on both sides. Reaction to light and accommodation was very weak and response to accommodation was also poor. Fundus showed primary optic atrophy on both sides. Patient had generalised hypotonia. Power was grade four and incoordination was present. Superficial reflexes were normal, deep reflexes were brisk. There were abnormal movements in the form of tremors, dyskinesia and choreiform movements of tongue, lips and limbs. Trombone sign was positive.

In mental status, N. S. was an undernourished, weak child of asthenic built. He was dressed appropriately but was not cooperative. Psychomotor activity was increased. He was restless hyperactive and showed abnormal movements in the form of dyskinesia, tremors and choreiform movements. Communication was difficult as speech was hesitant, explosive, lalling and slurred with echolalia and perseveration at times. He was fully conscious and well oriented. Attention was not easily rousable and was ill sustained. Affect showed sudden changes. He did not show any evidence of delusions or perceptual anomaly. Intelligence was subnormal. Because of subnormal intelligence and motor weakness other localizing signs especially agnosia, aphasia and aproxia could not be tested.

His haemogram and routine investigations did not show any abnormality. X-ray skull was negative. In serology WR was strongly positive (three plus), V.D.R.L. was positive. CSF colour was clear, protein was 60 mg%, sugar was 54 mg%, cells were less than five per high power field. CSF WR was also strongly positive (three plus). I. Q. assessment was tried on Coloured Progressive Matrices and Sanguin Form Board but patient did not cooperate. On Vineland Social Maturity Scale, developmental age was 3½ years, giving a D. Q.

of 23. Serology of patient's father was positive for WR and VDRL. His mother's WR, VDRL were negative.

Patient was put on procain penicillin—8 lacs units twice daily parenterally but on second day he developed high fever with chills, muscular pains, malaise and trachycardia. Penicillin was stopped and tetracycline 200 mg. four times daily orally along with vitamins was started. After five weeks tetracycline was stopped and xantinol nicotinate was started. He was followed up regularly. At four month follow up he showed improvement in speech, motor power, self care, and vegetative functions.

COMMENTS

The diagnosis of Juvenile Neurosyphilis was established by characteristic clinical feature, supported strongly by laboratory investigations and Herxheimer reaction to penicillin. The interest of reporting this case was first that it is extremely rare disease and secondly these cases are likely to be missed and misdiagnosed unless one is specially aware of its presentation. These patients, because of mental changes, are more likely to consult a psychiatrist. Early recognition and detection is important as response to treatment is disappointing in patients who come for treatment after development of mental symptoms. Therefore it is important that CSF should be examined in all congenitally syphilitic children at an early age in order to prevent later complications.

REFERENCES

BENAKAPPA, D. G., SURESH, P., MANIKYA, R., CHANDRASHEKHAR, S. K. AND SHIVNANDA (1978). A clinical study of congenital syphilis. Indian Pediatrics, XV No. 11, 943.

CLOUSTON, T. S. (1877). A case of general paralysis at the age of sixteen. J. Ment. Sci., 23, 419.

JEANS, P. C. AND COOKE, J. V. (1930). Prepubescent syphilis. Clin. Pediatrics, XVII, New York.

MEHTA, K. P. AND GHARPURE, S. V. (1979). Renal involvement in congenital syphilis—a review and study of 60 cases. Indian Pediatrics, Vol. XVI, No. 7, 611.

STEWART, R. M. (1933). Juvenile types of general paralysis. J. Ment. Sci., 79, 602.