CASE REPORT

WEST SYNDROME

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INTRODUCTION: West syndrome is a rare epileptic disorder in infants, children and adults, characterized by triad of infantile spasm, pathognomic EEG pattern and developmental regression. Hence we are reporting a case of 1 year old male who had typical clinical, radiological and EEG pattern.

A 7 and half month old baby, first born to non-consanguineous marriage presented with global developmental delay, flexor spasm, and significant perinatal insult in the form of moderate birth asphyxia with neonatal sepsis required 30 days of hospital stay at birth.

On Examination: Infant had microcephaly, visual impairment, spastic quadriparesis (LL>UL), MRI showed diffuse cerebral atrophy, ex-vacuo dilatation of ventricles. EEG done was showing modified hypsarrythmia, hence diagnosed as a case of west syndrome and baby was started on injection ACTH at 30 units then gradually increased to 60 units depending upon the response of baby. With treatment there was significant decrease in number of spasm per day from 60 to less than 10. Infant was also started on levetiracetam. After infant was spasm free physiotherapy was also advised.

DISCUSSION: West syndrome is characterized by triad of infantile spasm, pathognomic EEG pattern and developmental regression. Spasm can be flexor, extensor and mixed. Spasm usually occurs during drowsiness, feeding, shortly after infant awakes from sleep.⁽¹⁾ A rapid flexor spasm occurs in the form of involving neck, trunk and limbs followed by tonic contraction sustained for 2-10 seconds. When spasms are secondary to identifiable cause the infant is usually abnormal neurologically. Microcephaly is common in this group. Prognosis depends on cause, the interval between clinical spasm and hypsarrythmia, rapidity of start of treatment.⁽²⁾ The prognosis is better in the idiopathic or cryptogenic cases without known etiologic factor, and those having normal development status before the onset of the spasm, and normal neuroimaging prior to therapy Earlier studies, observed that in the subgroup of cryptogenic patients, a delay in initiation of treatment may be associated with worse outcome.^(3,4) In three series, with larger number of patients and longer follow up time, a short time between onset and treatment correlated with a more favorable outcome than a long lag in institution of ACTH.^(5,6,7,8) A study concluded that treatment lag is possibly a prognostic factor.⁽⁹⁾

Fig. 1: Showing intermittent spasms

CASE REPORT

Fig. 2: Showing Microcephaly

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CASE REPORT

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