SUBACUTE SCLEROSING PANENCEPHALITIS

(A Report on Two Cases)

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ABSTRACT

We report two cases of subacute sclerosing panencephalitis (SSPE) which is a rare, late neurological sequel, of measles infection acquired in childhood. The diagnosis was reached by adopting criteria defined by Dyken. Past history of measles was present in both the cases and they showed high titres of measles specific antibody in CSF and serum by quantitative ELISA.

MJAFI 1994; 50 : 298-300

KEY WORDS : SSPE; Measles.

Introduction

Subacute sclerosing panencephalitis is a distinct degenerative neurological disorder [1], which is preceded by measles infection in the distant past, often early in life. Very occasionally measles immunisation may be the preceding event [2,3]. The reported incidence of SSPE is 8 per million after measles infection and 0.7 per million doses of measles vaccine [1,2]. Dyken (1985) laid down definite criteria to diagnose SSPE (a) classical presentation and clinical findings that include intellectual deterioration and myoclonus, (b) EEG findings that show characteristically periodic slow bizarre complexes, (c) elevated CSF immunoglobulin levels, (d) elevated CSF measles specific antibody and (e) classical histopathological findings. Presence of any two criteria is considered enough for a diagnosis of SSPE, but serial (a) and (d) are considered the most important markers [1]. There are many reports of SSPE from India (5-7), but detailed immunological work up has been done in only one (6). Quantitative ELISA has not been used for immunological confirmation of SSPE in any earlier reports.

CASE REPORT

The salient clinical and immunobiochemical data of the two patients are summarised in Table 1

Discussion

SSPE is now considered a slow viral disease that is caused by measles virus that is defective in its genes involving envelope proteins like M, F and H (1), which are needed for budding of virus. As no extracellular expression of viral antigens occurs at cell surface immune destruction of infected cells can not occur. The defective virus is selected out in brain. Being unable to bud out, it spreads slowly within the network of CNS cells in the presence of normal immune response. The virus in SSPE has been shown to be more neurotropic than the regular wild virus [8].

SSPE follows one to ten years after measles. In the two cases reported, the interval has been seven years to as less as six months. In the first case, measles and SSPE have followed despite measles immunisation pointing to vaccination failure. The onset of SSPE is insidious with slow progression. Our first patient had a rapid progression in just two months. Scholastic deterioration was present in both the patients. Myoclonus, reportedly

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TABLE 1

Features	Patient 1	Patient 2
Clinical Data		
Sex and age	Male 8 years	Female 8 years
Past history of measles	At 18 months of age	At 7 years of age
Presenting complaints	Gradual scholastic deterioration Jerky movements left side Inability to walk Loss of speech	Gradual scholastic deterioration Jerky movements whole body Repeated falls Response in monosyllables Bowel, bladder incontinence
Duration of illness on admission	2 months	1½ years
Immunisation history	DPT and Measles	Only DPT
Clinical findings	Aphasia Spasticity three limbs, decorticate posturing (Fig. 1) Myoclonus Exaggerated tendon reflexes Planters upgoing	Dysarthria Flaccid paralysis both lower limbs (Fig. 2) Myoclonus Exaggerated tendon reflexes Planters upgoing Chorioretinitis
EEG	Bilateral epileptiform discharge	Abnormal record suggestive of generalised epilepsy and burst suppression
CSF		
Appearance	Clear	Clear
Protein	50 mg/dl	50 mg/dl
Sugar	54 mg/dl	80 mg/dl
(Random blood sugar)	(98 mg/dl)	(117 mg/dl)
Globulins	Elevated	Elevated specific
Measles antibody titre		
(ELISA units per ml)		
Serum	107.9	175.9
CSF	77.7	117.7

Fig. 1 : Decorticate posture.

the most important clinical finding (4) was present in both. EEG was not classical in our patients since the expected typical periodic complexes were absent. It is the experience of

Fig. 2 : Flaccid paralysis of both lower limbs.

other authors that typical first EEG is found in only 79% of cases, strict periodicity tending to occur with progression of disease [6].

Haemagglutination inhibition assays have been used by many to detect measles antibodies (6,9) and qualitative ELISA by some [7]. Many authors believed, mere demonstration of measles antibody in CSF is enough to clinch diagnosis of SSPE [9]. The patients of SSPE are known to have antibody titres 10-100 times higher than is expected in convalescent samples of sera [1]. High CSF antibody titres dictate intrathecal synthesis. We have put up atraumatically collected normal CSF samples as controls and found that none gave titres > 20 EU/ml (the cut off value for positivity in the kit used - Diamedix, USA). It is also known that CSF antibody levels may fluctuate during SSPE with periods of total absence [6]. Therefore antibody alone is not enough to confirm or refute a diagnosis of SSPE and the totality of clinical and other criteria are to be relied upon [4].

SSPE is generally a fatal disease. Some may die in few months, others may remain in a decerebrate state for years. Focal retinitis occurs in a majority causing blindness [1]. We found chorioretinitis in our second patient. There are reports claiming isoprenazine (Inosiplex) prolongs life in SSPE [10].

That measles infection gives rise to a life long immunity is well known. The fact that

the virus may remain sequestered in brain to produce a late sequel of SSPE have raised questions about the efficacy of viral clearance by a normally functioning immune defence.

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