Nigerian Journal of Paediatrics 1987; 14(2): 51-57

Subacute Sclerosing Panencephalitis in Enugu

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Summary

Iloeje SO and Izuora GI. Subacute Sclerosing Panencephalitis in Enugu. *Nigerian Journal of Paediatrics* **1987; 14:51.** Five cases of subacute sclerosing panencephalitis (SSPE) in children aged 3 - 13 years, seen at the University of Nigeria Teaching Hospital, Enugu, between November 1977 and October 1985, are presented. This number represents 0.1 % of the total number of cases reviewed for the period under study. Suggested possible reasons for the low incidence include genetic and racial factors, and failure of the people to seek medical help in hospital whenever they are ill.

Introduction

SUBACUTE sclerosing panencephalitis (SSPE) is a rare and devastating degenerative disease of the nervous system which most commonly affects children and young adults. It has an annual incidence in Britain, of about 1/million childhood population, and 5–10/million children with measles[1]. Ever since its first description by Dawson[2] in 1933, intensive research has been directed towards understanding the pathogenesis of the disease. Presently, it is believed to be associated with a previous infection by measles or some measles-like virus[3-6]. Although it usually runs a chronic and relentlessly progressive course, with an almost uniformly fatal outcome, various clinical stages of the disease have been described for both diagnostic and therapeutic purposes[7 8].

In 1981, a case of the disease was reported at the University of Nigeria Teaching Hospital (UNTH), Enugu[9]. Since then, an attempt has been made to keep an SSPE register in the Paediatrics Department of the hospital. The purpose of this paper is to report the present stage of the register for the 8-year period, November 1977 to October 1985.

Patients and Methods

All children with neurological disorders presenting at the consultants' clinics and children's outpatient and emergency departments were referred, for the purpose of this study, to the paediatric neurology clinic where they were seen by one or both authors. Those admitted to the paediatric medical wards were also included in the study. Detailed history, including history of seizures, measles or other viral or bacterial infections and immunization,

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was obtained. Detailed clinical examination, with emphasis on the nervous system was carried out.

Investigations were done as indicated; these included a complete blood count, serum urea and electrolytes, cerebrospinal fluid studies, mantoux test, blood sugar estimation, skull and chest X-ray and electroencephalography (EEG). The EEG machine used was a 10 channel 'Grass' equipment, model 8–10C, and the international (10/20) system of electrode placement was used. A diagnosis of SSPE was made on the basis of strong clinical evidence, in addition to the typical EEG features. Clinical staging of the disease was based on the following criteria proposed by Jabbour *et al* in 1969[8]:

Stage 1: Cerebral signs (mental, behavioural)

Affectionate displays, drooling, forgetfulness, indifference, irritability, lethargy, regressive speech, slurred speech, withdrawal.

Stage 2: Convulsive, motor signs

Dyskinesia, choreoathetoid movements and postures, inco-ordination of trunk and limbs, myoclonus.

Stage 3: Coma, opisthotonus

Decerebrate rigidity, extensor hypertonus, irregular stertorous respiration, no responsiveness to any stimulus.

Stage 4: Mutism, loss of cerebral cortex function, myoclonus

Flexion of upper and lower limbs, hypotonia, occasional limb myoclonus, pathologic laughter, crying, startled by noise, wandering of eyes.

Results

A total of 1,637 cases with neurological disorders were seen in the paediatric neurology clinic, while 2,799 others presented at the outpatient clinic and Children's Emergency. Some of these were admitted into the paediatric medical wards. Thus, the total number of neurological cases reviewed for the period under study was 4,436, while 4,488 EEGs were obtained and evaluated. Of these cases, only 5 patients met the diagnostic criteria stated above.

Their clinical data are shown in the Table, while their EEGs appear in Figs 1–5. The table shows that the patients were all males, and their

TABLE

Clinical Data of 5 Children with Subacute Sclerosing Panencephalitis

Patient	*Sex*	*Usual Residence (Urban/Rural*	*Age at Presentation (years)*	*Measles Immunization*	*Age at Measles Infection (years)*	*Age at Onset of SSPE (years)*	*Interval between Measles Infection and Onset (years)*	*Clinical Stage**	*EEG (Fig)*
EA	M	rural	3½	-	2	2½	½	III	1
IR	M	rural	8	-	½	7½	7	II	2
IS	M	rural	9	-	1	8	7	II	3
OA	M	rural	10½	-	1	10	9	II	4
OT	M	urban	13½	-	-	13	-	II	5

*Jabbour *et al*[8]

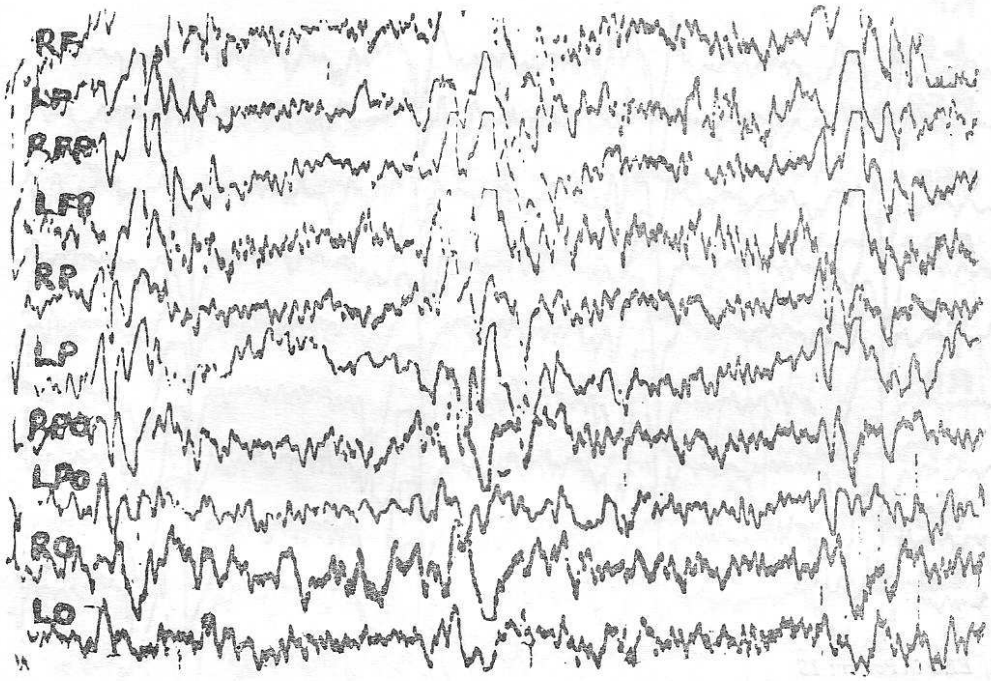

Fig 1. EEG in Patient EA

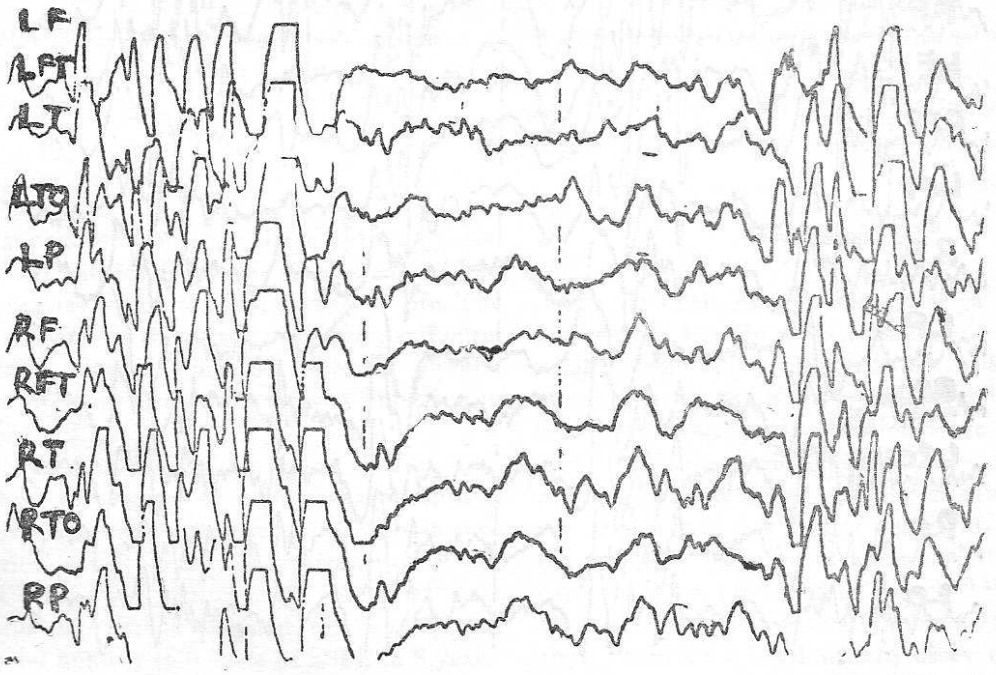

Fig 2. EEG in Patient IR

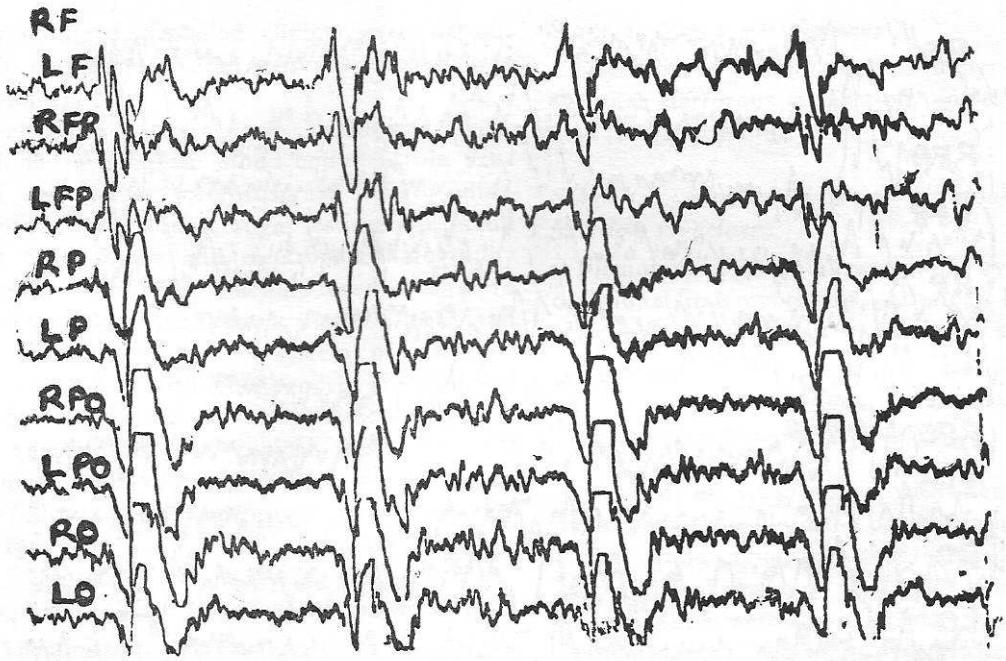

Fig 3. EEG in Patient IS

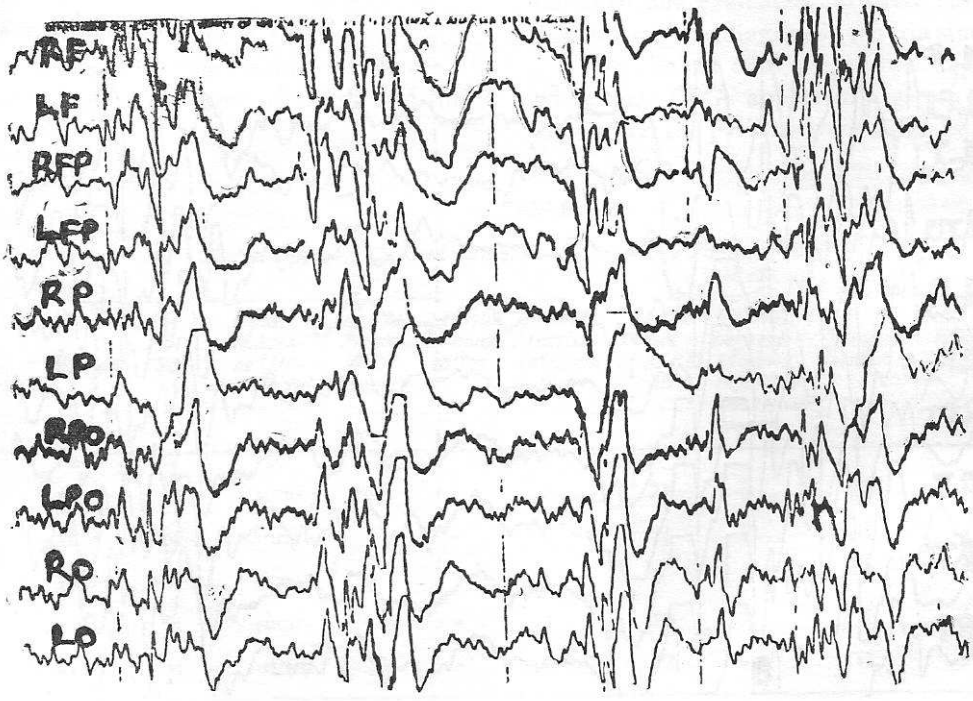

Fig 4. EEG in Patient OA

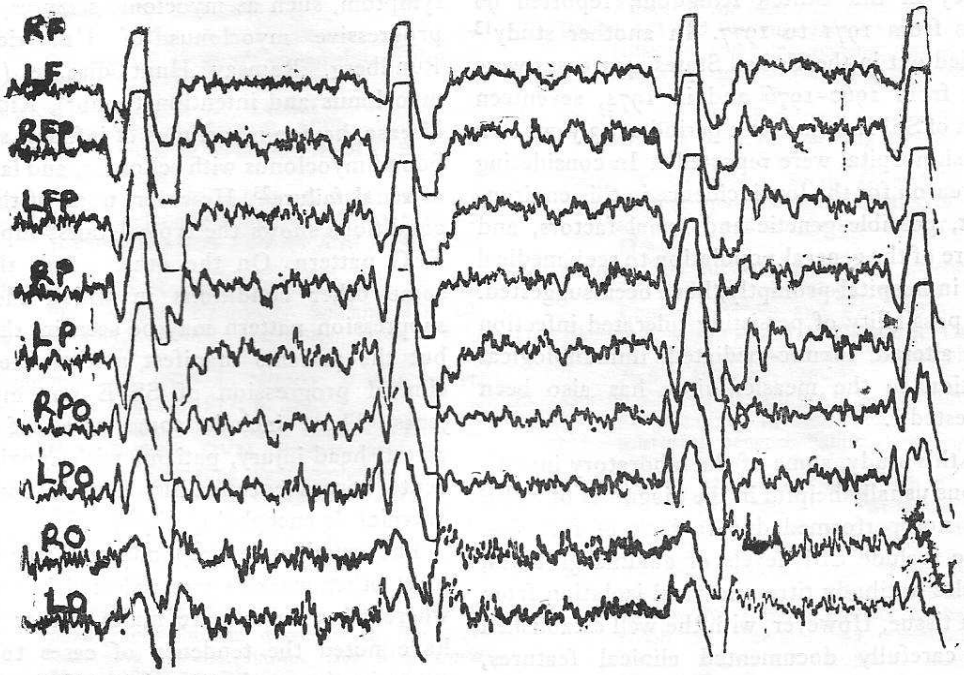

Fig 5. EEG in Patient OT

The classical burst suppression pattern (described in the text) is seen in all the EEG records. Note that in Figures 2, 3 and 5, only nine channel records were obtained, as the pen in channel I did not write.

R	= *right*	*T*	= *temporal*	*FP*	= *fronto-parietal*
L	= *left*	*O*	= *occipital*	*PO*	= *parieto-occipital*
F	= *frontal*	*FT*	= *fronto-temporal*	*TO*	= *temporo-occipital*
P	= *parietal*				

ages at presentation ranged from $3\frac{1}{2}$ to $13\frac{1}{2}$ years. Four of them resided in the rural area and had had measles within the first 2 years of life. The interval between the attack of measles and onset of SSPE ranged from 6 months to 9 years. There was no history of measles infection in one patient.

All the EEGs show the typical paroxysmal, synchronous bursts of high voltage diphasic activity followed by periods of diminished electrical activity—usually referred to as the burst suppresion pattern. If the first case reported in 1981[9] is included, this would bring the total number to 6 cases of SSPE in 8 years.

Discussion

The UNTH Enugu, serves as a referral centre for Anambra State and several other neighbouring states. With such a wide catchment area of about 14 million people, it is surprising to find such a low incidence of SSPE over an 8–year period, especially as measles, which is strongly linked with the disease, is still very common in this part of the world. Prior to the first case recorded in our hospital, only 4 cases had been published in Nigeria[10 11]. In the United States of America, Jabbour *et al*[12] in 1972, reported a total of 219 cases over the

period 1960–1970. Bellman and Dick[1] in a survey in the United Kingdom, reported 96 cases from 1971 to 1977. In another study[13] carried out in the United States, 453 cases were seen from 1960–1976 and in 1974, seventeen cases of SSPE seen over a period of 13 years in a British hospital were reported[14]. In considering the reason for the low incidence in this environment, possible genetic and racial factors, and failure of the general population to seek medical help in hospital promptly, have been suggested. The possibility of persisting tolerated infection and altered thymic-mediated immunological reaction to the measles virus has also been suggested[15].

In this study, some of the laboratory investigations usually helpful in the diagnosis of SSPE were not performed due to lack of facilities. These include CSF levels of gamma globulin, measles antibody titre and viral isolation from brain tissue. However, with the well established and carefully documented clinical features, accompanied by the typical EEG pattern, we have no doubt about the diagnosis of SSPE in the 5 cases. In the study by Mackenzie *et al* in South Africa[16], cases were diagnosed on strongly presumptive clinical evidence only. A past history of measles lends further support to the diagnosis, although it is known that this may not necessarily be present in all cases[17-19]. Four of our 5 cases had clinical measles.

Case 1 in this report had SSPE only 6 months after clinical measles infection. Even though the interval is rather short when compared to the more frequently reported intervals of 2 years and above[1 20], it is not unique, as Bhettay *et al* have reported an even shorter latency period of 2 months[21]. It could be that the latency period is related to the severity of the immunological derangement, since immunological abnormalities have been reported in patients developing SSPE after measles infection[13 22].

Differential diagnosis of SSPE include certain degenerative or progressive neurological disorders in which myoclonus is a prominent symptom, such as myoclonic seizures, familial progressive myoclonus of Unverricht and Rundberg, Ramsay Hunt disease (seizures, myoclonus and intention tremor), Kinsbourne disease, benign myoclonus of infancy, stimulus-bound myoclonus with echolalia, and late stages of renal failure[23]. However, none of the above conditions shows the typical burst-suppression EEG pattern. On the other hand, there are some other conditions in which the burst-suppression pattern may be seen in the EEG, but the patients manifest neither the typical clinical progression of SSPE nor myoclonic jerks. These include some cases of cardiac arrest, head injury, patients with anoxic states, acute herpes encephalitis, hepatic coma, and Wernicke's encephalopathy[24].

The usual male preponderance reported by most other workers was reflected in our series where all 5 cases were males. Other workers have noted the tendency of cases to cluster more in the rural areas than urban centres[12]. This trend was also observed in our series, where 4 of the 5 cases were normally resident in rural areas.

Most registries usually have the shortcoming of representing only a sample of the population, and ours is no exception. We hope that this report will highlight the need for a more extensive epidemiological study of this disease in other parts of Nigeria.

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Accepted 14 January 1987.