EPIDEMIOLOGY OF SUBACUTE SCLEROSING PANENCEPHALITIS (SSPE) AND OTHER ENCEPHALITIDES IN KARACHI AREA; A PROGRESS NOTE

Pages with reference to book, From 169 To 173

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Abstract

Relative frequency of neurologic diseases in Karachi, with special reference to inflammatory disorders has been estimated by using the comprehensive data from the Deptt. of Neurology, Dow Medical College, Karachi,
The incidence of subacute scierosing pan encephalitis has been estimated to be approx. 10 per million. Figures for multiple sclerosis appear comparable to those from India, Turkey and Israel. Rates for active encephalitides and postencephalitic sequelae are unbelievably higher than in the developed countries (JPMA 36:169 1986).

INTRODUCTION

This is a part of comprehensive collaborative studies on encephalitides in Karachi area. Its purposes include: 1) clinical patterns and the relative frequency of the diseases, 2) estimation of their prevalence rate, 3) morbidity and mortality patterns in Karachi area due to various neurological disorders, particularly encephalitides, 4) evaluation of the risk factors for SSPE.
Thrget diseases are inflammatory disorders of the cerebral parenchyma of all kinds.

TABLE - I

Applicable Codes in the 9th Revision of International Classification of Diseases (ICD–9) in the Survey of "Encephalitides" in Karachi, Pakistan.

Code	Disease	Exclusion
046	Slow virus infection ofcentral nervous system (CNS)	
049.8 - 049.9	Other non-arthropod-borne viral diseases of CNS	Pure meningitides
054.3	Herpetic meningoencephalitis	
056.0	(056.01 in ICD-9CM) Encephalomyelitis due to rubella	
062 - 064	Mosquito-borne and tick-borne viral encephalitis, and encephalitis transmitted by other and unspecified arthropods	
072.2	Mumps encephalitis	
323*	Encephalitis, myelitis and encephalomyelitis	Pure myelitides
324.0	Intracranial abscess	
046 - 072.2	Selected from the Chapter I – Infectious and Parasitic Diseases (001 – 139)	
323 - 324.0	Selected from the Chapter VI - Diseases of the Nervous System and Sense Organs (320 – 389)	

Table I summarizes such diseases with the applicable codes in the 9th Revision, International Classification of Diseases (ICD-9).

TABLE - II

Diagnostic Criteria of SSPE for Epidemiological Studies in Karachi, Pakistan.

Definite:

Those verified pathologically, and/or by either one or more of

1) measles virus isolation from the brain
2) demonstration of the measles virus antigen in the brain
3) demonstration of the measles virus nucleocapsids in the brain cells

Serological probable:

Those clinically compatible with SSPE, showing increased anti-measles titer in the cerebrospinal fluid

Clinical Probable:

Those satisfying all of the following clinical features and for which no evidence more strongly suggested other diagnosis than SSPE

1) afebrile onset*
2) onset before 15 years of age
3) initial incipient mental deterioration combined or followed by myoclonus which soon generalizes
4) ADL-dependent within 1/2 year and/or death within 2 years after onset
5) EEG showing brief, recurrent, synchronous, high-voltage slow waves, at a fairly regular interval

Possible:

All others with clinical features suggestive of SSPE and with no counterevidences for the diagnosis

* fever preceding the onset of otherwise totally acceptable case as Clinical probable does not immediately cause the rejection of the case.

Table II describes diagnostic criteria for SSPE.

MATERIALS AND METHODS

Materials include ; 1) cases, their histories, and the outpatient (OPD) and discharge records of the Department of Neurology, Civil Hospital; 2) the death certificates in the Abbasi Shaheed Hospital in which death certification is well organized according to the rules proposed by the World Health Organization.

Prevalence rate of various groups of the target diseases was measured indirectly based on the number of the OPD patients with the diseases relative to motor neuron disease (MND), assuming that the rate from MND is 3-5/100,000 uniformly throughout the world.

Patterns of the morbidity of the target diseases and other disorders were evaluated on the basis of statistics of the OPD and the discharged patients, based on a classification by ICD-9.

Patterns of the mortality were determined by indirectly calculating age-and-sex specific mortality rates due to the target diseases and various other causes. The Abbasi Shaheed Hospital material was used instead of the regional certificates which were unsatisfactory.

Risk factors for SSPE were evaluated on the pattern of an ongoing Case Control Study being done in Japan by one of the authors (K.K.). Design of the Study was similar to those of Detels et al[1] and Halsey et al.[2]

INTERIM RESULTS AND DISCUSSION

Collected materials and on-going surveys are:

1) Statistics of the neurological patients: The following three statistics have been collected: a) OPD, selected diagnoses, all patients, April 1974 -March 1984; b) OPD, all diagnoses 2,000 consecutive patients around 1980 and c) Inpatients, all discharge diagnoses, all patients, March, 1975-March 1984.

TABLE - III

Number of Out-Patients with selected Diagnosis, 9 April, 1974 - 4 March, 1984, Department of Neurology, Civil Hosptial, Karachi, Pakistan

Diagnosis	Male	Female	Total
Motor neuron disease	24	2	26
Kugelberg-Welander's disease	2	1	3
Parkinson's disease	80	32	112
Huntington's chorea	3	1	4
Senile/presenile dementia	3	1	4
Friedreich's ataxia	4	3	7
Spinocerebellar degenerations, NOS	15	8	23
Syringomyelia	2	1	3
Multiple sclerosis	15	7	22
Myasthenia gravis	16	6	22
Polymyositis	16	2	18
Charcot-Marie-Tooth disease	4	2	6
Duchenne muscular dystrophy	15	1	16
Becker-type	2	0	2
Facioscapulohumeral	2	0	2
Limb-girdle	3	1	4
Distal	2	0	2
Ocular	0	1	1
Muscular dystrophies NOS	23	9	32
Myotonic dystrophy	1	0	1
Encephalitides	26	16	42
Acute	1	0	1
Subacute	0	1	1
"Persistent"	2	0	2
Measles or post-measles	4	5	9
Viral	5	1	6
Toxoplasma	1	0	1
Postvaccinal	2	1	3
Brainstem	0	0	0
Cerebellar	0	1	1
NOS	11	7	18
Meningoencephalitis	1	0	1
Viral	1	0	1
Encephalomyelitis	3	2	5
Mumps	1	1	2
NOS	2	1	3
SSPE	7	5	12
Postencephalitic	70	45	115
mental retardation	10	6	16
epilepsy	15	8	23
mental retardation and epilepsy	2	3	5
hemiparesis	5	3	8
otherwise-specified	6	3	9
NOS(state, syndrome, brain damage, etc)	32	22	54
Postmeningoencephalitic	2	0	2
Postencephalomyelitic	0	1	1

NOS : not otherwise specified

Table III shows the numbers of patients based on the statistics. They provide a basis for an indirect estimation of the prevalence rate of SSPE. Table III also discloses unusual male! female ratios in all diseases. Even in multiple sclerosis or myasthenia gravis, which are known for a female excess world wide, males are preponderant. It is possible that female patients are disadvantaged in Pakistan in asking

for a medical consultation.

2) Cases with SSPE

Table N summarizes cases. Suspected as SSPE by the second author either in the Civil Hospital or in his private clinic. Clinical diagnosis is tentative and the cases were classified by the criteria in Table II according to the nature of the evidences that support the diagnosis. The Definites and the Serological probables meet international diagnostic standards, but a category for the Clinical probables has been proposed to separate cases lacking in laboratory data, but satisfying clinical criteria from the rest of "SSPElike" cases. In epidemiological studies, the possibles are not accepted as SSPE.

There are, based on the clinical case ascertainment as of 22 March 1984, and on the serological data subsequently reported, no Definite, 8 Serological probables, 9 clinical probables along with one possible case. Total of the cases in three Original diagnosis of SSPE includes suspected cases. 3 unadmitted cases in the first group were not classified for which adequate clinical records and EEGs were not available. Of 8 Serological probables, 6 cases (0014, 0086, 0090, 0130, 0132, 0134) were included in the clinical case series of the present collaborative study. One each of male and female cases were reported from the Department of internal medicine of CHK and the Abassi-Shaheed hospital, which were not included in this table. acceptable categories were 11 males and 6 females, indicating a striking male predominance. These numbers are preliminary and changable as new cases or new laboratory data emerge.

3) Prevalence rate of SSPE

An exhaustive case finding is necessary for a direct measurement of the prevalence rate of a given disease in a given community, which is not feasible in Karachi in this project. An indirect method, therefore, has been proposed to estimate the rate relative to that of MND based on the numbers of the OPD patients with the diseases. Excluding high risk foci in Guam or Kii peninsula of Japan, prevalence rates of MND range 0.8-6.7 in areas of the world, the average being 3.7 per 100,000[3]. As seen in Table Ill number of MND is 26 and the acceptable cases of SSPE in the first group of

TABLE - IV

Tentative Classification of Cases as of March, 1984, Suspected as SSPE by Ahmed in Karachi, Pakistan, grouped by the Sources of the Cases.

Sources of cases recorded	Definite	Serological probable	Clinical probable	Possible	Rejected	Records unavailable	Total
1. Cases in the OPD register with an initial diagnosis of SSPE	0	5	2	0	2	3	12
2. Referred cases directly admitted and discharged with a diagnosis of SSPE	0	1	5	1	5	0	12
3. Private cases with SSPE examined by Ahmed	0	2	2	0	1	0	5
Total	0	8	9	1	8	3	29

Table IV is 7, pooling both the sexes. Prevalence rate of SSPE in Karachi is, simply multiplying 3.7 by 7/26, 1.0 per 100,000, or 10 per million. Cases in the first group only was used for this calculation, because other cases have different motivations in seeking consultation.

TABLE -V

Average annual Incidence Rates of SSPE in 13 countries.

Country	Report	Years studied	Number of cases	Population (million)	Average incidence per year ($X 10^{-6}$)
North Island, New Zealand	Baguley & Glasgow, 1973[4]	1956-59	27	?	7.7**
Israel	Soffer et al, 1976[5]	1966-73	52	3.33 (1970)	1.95
Algeria	Baugermough*	1967-74	21	1.9 (1970)	1.6
Quebec, Canada			12	5.7 (1965)	1.0
NE Region, North Scotland	Clark & Best 1964[6]	1952-61	4	?	<1.00
Iran	Mirchamsy*	1976	15	33.0	0.45
Finland	Donner & Haltia, 1969[7]	1960-69	20	4.65 (1965)	0.43
Belgium	Canal & Torck, 1964[8]	1939-61	79	9.6 (1965)	0.28
South Africa	Mackenzie et al, 1975[9]	1955-74	79	17.8 (1965)	0.22
Cape Province only	Ditto	1970-74	30	10.6 (1965)	(0.57)
Sao Paulo state, Brazil	Canelas et al, 1967[10]	1954-66	25	13.0 (1960)	0.15
U.S.A.	Modlin et al, 1977[11]	1960-77	475	203.2 (1970)	0.13
United Kingdom	Dick, 1975[12]	1961-75	49	55.5 (1970)	0.06
Japan	Okuno et al, 1976[13]	1966-76	45	103.5(1970)	0.04

* Personal information ** for population aged 5-20

Table V summarizes available incidence rates of SSPE in 13 countries. Such rate is not directly available in Karachi, but may be about 7-10 per million, based on the equation that prevalence rate incidence rate X average duration the latter being 1-1.5 years in SSPE IN MOST COUNTRIES. A follow-up study is being planned to obtain the average duration in Karachi cases. Validity of these estimations is debatable, but if we accept this tentative figure, it justifies clinical impressions of the second author that SSPE is common in Karachi.

Incidentally, figures for multiple sclerosis range 3.4-4.2 X l0,[5] being comparable to those reported from India, Turkey and Sephardim Israelis. Rates for active encephalitides range 6.4-8.0 for postencephalitic sequelae, 18.4-23.0 being unbelievably higher than the values in developed countries.

CONCLUSION

Even at this preliminary stage of the study convincing data indicated that inflammatory afflictions of the central nervous system predominate in the neurological diseases in urban Karachi. SSPE appeared to be frequent which is, at least in communities in the temperate countries, usually considered to occur one per million per year. Regional factors, particularly climatic and socioeconomic, may be responsible for such a pattern. Results of the case-control study are awaited to evalnate the factors.

ACKNOWLEDGEMENTS

This study was supported by the Japan Ministry of Education. Cordial thanks are due to the Medical Superintendent, Civil Hospital for permitting the use of clinical data, and to Mr. Aftab Ahmed, Department of Neurology, Civil Hospital, for secretarial help.

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