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AUTONOMIC SEIZURES AND AUTONOMIC *STATUS EPILEPTICUS* IN EARLY ONSET BENIGN CHILDHOOD OCCIPITAL EPILEPSY

Panayiotopoulos syndrome

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ABSTRACT - To study clinical and EEG features of children with ictal vomiting and no underlying brain lesions (Panayiotopoulos syndrome). The subjects were 36 children aged 2-13 years. The onset of seizures occurred between 1 and 5 years of age. Fourteen children (38.8%) had a single seizure. Fourteen children (38.8%) had autonomic *status epilepticus.* Impairment of consciousness was reported in 30 (83.3%) children, eye deviation in 10 (27.7%) other autonomic symptoms and head deviation in 9, generalization in 8, visual symptoms in one child, and, speech arrest or hemifacial motor symptoms in 8 cases. The EEG showed occipital spikes or spike-wave complexes in 27 (75.0%) children, blocked by opening of the eyes in 8 (22.2%) cases. Nine patients (25%) also had rolandic spikes and 3 had extraoccipital spikes. Six (16.6%) patients had normal EEG. No clinical differences were observed between patients having occipital or extraoccipital spikes. In children only with autonomic seizures, the spikes are predominantly occipital but blockage by opening of the eyes is a less frequent feature. In some children there is an overlapping of different focal childhood idiopathic syndromes.

KEY WORDS: benign childhood epilepsy, eletroencephalography, epileptiform activity, ictus emeticus, occipital spikes, Panayiotopoulos syndrome.

Crises autonômicas e *status epilepticus* autonômico na epilepsia occipital benigna da infância de início precoce (síndrome de Panayiotopoulos)

RESUMO - Estudar aspectos clínico-eletrencefalográficos de crianças com vômito ictal e sem sinais de lesão cerebral (síndrome de Panayiotopoulos). Foram estudadas 36 crianças na faixa etária de 2-13 anos. O início das crises ocorreu entre 1 e 5 anos de idade. Quatorze crianças tiveram crise única. *Status epilepticus* foi observado em 14 (38,8%) casos. Distúrbio da consciência foi relatado em 83,3% das crianças, desvio ocular em 27,7%, outros sintomas autonômicos e desvio da cabeça em 26,4%, generalização em 23,5%, bloqueio da fala ou sintomas motores da hemiface em 23,5% das crianças e sintomas visuais em um caso. O EEG mostrou pontas ou complexos de ponta-onda em 27 (75,0%) casos, bloqueados pela abertura dos olhos em 8 (22,2%) pacientes. Nove pacientes tiveram também pontas rolândicas e 3, pontas extraoccipitais outras. O EEG foi normal em 6 crianças. Não houve diferença clínica entre as crianças com pontas occipitais e extraoccipitais. Em crianças com crises autonômicas as pontas foram de predomínio occipital, mas bloqueio pela abertura dos olhos foi pouco freqüente. Em alguns casos houve sobreposição de diferentes síndromes idiopáticas focais da infância.

PALAVRAS-CHAVE: epilepsia benigna da infância, eletrencefalografia, atividade epileptiforme, ictus emeticus, espículas occipitais, síndrome de Panayiatopoulos.

Panayiotopoulos (1988)[1] described a form of childhood benign focal epilepsy corresponding to about 6% of the epilepsies occurring in this age range, with an incidence peak at 5 years of age and affecting both sexes equally. The prognostic is good and seizure remission frequently occurs within 1 or 2 years of the onset. The crises are focal, initially characterised by a complaint from the child of "not feeling well", followed by autonomic signs or symptoms frequently characterised by nausea or vomiting. During the seizure evolution, the child becomes flaccid and unresponsive, with a tonic eye and/or head deviation, and,

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in some cases, followed by a generalised convulsion or motor hemiconvulsion. The seizures are frequently prolonged and in one third of the cases there is just one single seizure. In approximately 60% of the children, the seizures occur while sleeping. Some of the children present other autonomic manifestations such as: paleness, cyanosis, pupil dilation, facial blushing, coughing, urinary and/or faecal incontinence and even irregular breathing. The background activity (BA) of the electroencephalogram (EEG) is normal and in 90% of the cases, the epileptiform activity (EA) is characterised by spikes or spike-wave complexes of great amplitude and a functional nature, with multifocal localisation predominating in the posterior regions. Occipital localisation of the EA occurs in approximately 68% of the children. The EA frequently disappears at about thirteen years of age but may persist after remission of the clinical seizures[1,2].

Recently this form of epilepsy was recognised as early-onset benign occipital childhood epilepsy (EOOE) in the proposal of the Task Force on Classification and Terminology of the International League Against Epilepsy (ILAE)[3]. However, several publications question the classification of this epileptic syndrome as occipital epilepsy, since the commonest ictal characteristics suggest an extra-occipital location and the EEG alterations are frequently multifocal, despite the elevated occurrence of occipital-located spikes[2,4-6]. Since autonomic expressions are frequent amongst the critical manifestations of EOOE, various recent studies have specifically approached this clinical aspect[6-8].

Thus the objectives of the present research were to study the correlation between the electroencephalographic characteristics and the clinical aspects of children with autonomic focal seizures with Panayiotopoulos syndrome.

METHOD

In a retrospective way, the medical registers of children examined in the Electroencephalographic Service of the Celso Pierro General & Maternity Hospital (PUC-Campinas) and in the private clinics of the authors, presenting focal epileptic seizures starting with nausea and/or vomiting followed by other critical autonomic symptomatology or even that of a generalised secondary nature with diagnosis of early onset benign childhood occipital epilepsy (Panayiotopoulos syndrome), were analysed.

The inclusion criteria were: critical autonomic symptomatology, normal neurological examination, normal neuropsychomotor development, no past history suggestive of neurological disorders, normal computerized tomography (CT) scan, and normal intercritical EEG background activity[7].

The age and gender of the children were noted from the registers and the type, total number and duration of the seizures and the age when they occurred, noted and analysed.

From the EEG the following were studied: presence of EA according to type - spikes (S) or spike-wave complexes (SWC) -, location, extension and reactivity to the opening and closing of the eyes. The presence of spikes evoked (ES) by hand and foot stimulation was also studied.

The research plan was approved by the Ethics Commission for Research in Human Beings of PUC-Campinas.

RESULTS

Thirty-six children presenting autonomic epileptic seizures were included in this study. The initial manifestation was nausea and/or vomiting in all the cases. Other autonomic manifestations (paleness, abdominal pain and cyanosis) were observed in 50% of the cases.

During the sequence of the seizures, eye deviation occurred in 10 (27.7%) cases and head deviation in 9 (25%). Complaints about vision were made in one case and 8 (22.2%) had seizures with speech arrest or hemifacial motor symptoms, choking sensation or parestesias of the hands. Impairment of consciousness was noted during the seizure in 30 (83.3%) of the children, hemiconvulsions in 5 (13.8%) cases, generalised convulsions in 3 (8.3%) and postictal symptoms were referred to in 11 (30.5%) children.

The seizures occurred while sleeping in 26 (72.2%) cases.

The seizures started between 1 and 5 years of age in 86.1% of the cases (average age 4 years). The female gender showed a discreet, non-significant predomination (58.3%). In 80% of the children the total number of seizures was up to 3 episodes, 14 children only had one single seizure and seizures were numerous in 7 children. In 14 children (38.8%), the seizures lasted more than 20 minutes, characterising autonomic status epilepticus.

Electroencephalographic aspects – As the inclusion criteria all the children showed normal BA in the EEG.

Thirty children (83.3%) presented EA.

The EA was occipital-located in 27 (75%) of the children. It was exclusively occipital in 18 cases but there was also generalised SWC in 3 cases and extra-occipital (central and mid-temporal) in 9 children. In 8 cases, on opening the eyes, blocking or attenuation of the occipital EA occurred.

Three children presented EA in the central and mid-temporal regions and in 6 cases there was no register of EA on the EEG.

ES was found in 1 case (2.7%) due to stimulation of the feet and hands.

Table. Clinical characteristics of the children with autonomic seizures according to the EEG findings.

Electroencephalogram	Total nº of seizures			Critical characteristics								
	1	2-3	>10	dev.	imp. con.	mot	oro-phar	vis	gen	sleep	post-ictal	status
Normal n=6	4	–	2	3	5	2	–	1	–	3	3	2
EA occ. n=18	8	7	3	8	13	3	5	–	2	16	6	9
EA occ. & centro-temporal n=9	–	8	1	3	9	1	2	–	1	5	2	3
EA centro-temporal n=3	2	–	1	2	3	1	1	–	–	2	–	–
Total	14	15	7	16	30	7	8	1	3	26	11	14

n, number of cases; occ, occipital; nº, number; dev., eye and/or head deviation; imp. com., impairment of consciousness; mot, focal or hemi-generalised motor seizure; oro-phar, oro-pharyngeal manifestation; vis, focal with visual symptoms; gen, seizure followed by tonic-clonic generalisation; sleep, seizure while sleeping; status, seizures lasting >20 minutes.

No significant differences were observed between the clinical and critical aspects amongst the children with occipital or extra-occipital EA or with normal EEG (Table).

DISCUSSION

The findings of this study with respect to the age of the child and the number, duration and characteristics of the seizures, corroborated the data found in previous publications involving approximately 800 cases of EOOE described in the literature[1,4,5,9-11].

As already mentioned[2,4,9-12] some cases (22.2%) were observed involving only a single seizure, in which the autonomic symptomatology was followed by oro-pharyngeal manifestation, characteristics suggesting the overlapping of two benign childhood epilepsies, the early onset occipital one and that with centro-temporal spikes.

With respect to EA, the predominant region was occipital in the majority of cases, exclusively centro-temporal in some and inexistent in others. In a significant way, differences were not observed between the children presenting occipital or centro-temporal focal EA or normal EEG with respect to the total number of seizures, the duration or the type of seizure. These findings are consistent with the initial description of Panayiotopoulos and that of other authors[1,2,5,9].

In 37% of the cases, the occipital EA was characterised by spikes, a finding similar to that of other authors, who also suggested similarity between the neurophysiological characteristics of this occipital EA with those of the rolandic spikes of BIECT[1,2,9]. In 29.6% of the cases, blockage or attenuation of the occipital EA was caused by opening the eyes, a lower percentage than the 80% and 100% of cases reported respectively by Caraballo et al.[9] and Verroti et al.[13], but similar to that reported by other authors[5,14].

In the children presenting occipital EA, 11.1% of the cases were observed to present generalised SWC, in agreement with other studies[2,9].

Analogous to the study of Covanis et al.[8], some of the children with autonomic clinical manifestations presented exclusively centro-temporal EA in the EEG.

ES by foot stimulation was observed in one (2.7%) child. In previous studies[15] it had already been observed that ES could appear on the EEG due to several types of epileptic syndrome, particularly with benign focal epilepsy in childhood, but that it could also appear in normal children with no epileptic manifestations[16].

In conclusion, in the children studied a focal epileptic state is relatively frequent. The EEG showed that the EA was preferentially occipital, consisting of spikes and/or SWC that could, in some cases, be blocked by opening the eyes. Centro-temporal EA occurred in a significant percentage of the cases and could be the only alteration seen on the EEG. There was overlapping of the clinical and electroencephalographic characteristics in some cases, suggestive of two benign childhood epileptic syndromes, the early onset occipital one and that with centro-temporal spikes.

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