Dacrystic epilepsy

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SYNOPSIS We observed a 69 year old patient who had spontaneous paroxysmal attacks of weeping with lachrimation, contorted facies, and (usually) head-turning to the right. These episodes were followed by confusion and amnesia. During a pentylenetetrazol-evoked attack the EEG showed a right temporal delta rhythm. Atrophy, particularly of the right temporal regions, was noted on pneumoencephalography. We propose the term 'dacrystic epilepsy' for this rare type of seizure disorder.

No one today would dispute that laughter can be a manifestation of epileptic activity. The phenomenon has been documented for over 100 years since Trousseau (1873) observed laughter during an ictal episode. Since then there have been upwards of 120 similar case reports (Rey-Pias, 1972; Chen and Forster, 1973). But crying—which might be looked upon as the expressive antithesis of laughter—is curiously absent from most descriptions of epilepsy. Indeed, an extensive search in the literature revealed only five case reports in which weeping might be construed as epileptiform (Zilgien, 1906; Purves-Stewart, 1927; Davison and Kelman, 1939; Efron, 1961; Stutte, 1963).

Recently, we had the opportunity to study a patient with a seizure disorder in which episodes of paroxysmal crying were prominent. The phenomena were unusual and intriguing. Because of the rarity of this type of seizure, we feel that it warrants reporting. We propose that the term 'dacrystic epilepsy' [Greek *dakryon*, tear] be used to describe this type of seizure disorder.

CASE HISTORY

In January 1975 a 60 year old, right-handed man was hospitalized because he suffered from seizures which

were nearly continuous despite the administration of diphenylhydantoin and phenobarbitone. His mental state precluded us from obtaining a thorough medical history, but the following is known about his past:

In 1954 the patient was treated for syphilis of the central nervous system. The diagnosis was supported by cerebrospinal fluid that showed a positive serological test and a first zone colloidal gold curve. At that time he received penicillin for 20 days. Some time after this he developed epilepsy. Initially the seizures were grand mal in type, but in subsequent years the pattern changed, so that on separate occasions he would experience one or more of the following: grand mal convulsions, focal motor seizures involving the left-sided limbs, laughing spells, and episodes of crying. In recent years, most episodes were left-sided focal motor in type.

In 1958 he was seen by a physician for a complaint of generalized convulsions which occurred once or twice a month. These were sometimes preceded by a 'bad taste' in his mouth. Examination revealed only a left superior incongruous quadrantanopsia; no further investigations were carried out at that time. Three years later, however, re-examination of the visual field revealed that the visual loss had progressed and that he now had a complete left homonymous hemianopsia. Although the EEG demonstrated spike discharges in the right parietal region, right carotid arteriography was normal. An examination in 1964 disclosed a mild hemiparesis, the first focal deficit other than hemianopsia to be noted. This is the last record we have for the patient until the hospitalization described below.

At the time of the present admission there was almost constant twitching of the left lower facial musculature. We observed that the patient had a

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severe organic mental syndrome, and this was subsequently confirmed by psychometric testing. The following deficits were noted: a dense left hemianopsia; a mild left hemiparesis which involved the face, tongue, arm, and leg, and which was accompanied by slightly increased tone, hyperreflexia, and an extensor plantar response; moderate left hypaesthesia.

At times, without any apparent provoking stimuli, he would begin to weep. These episodes seemed to be like normal crying because they were accompanied by lacrimation, contorted, mournful facies, and sobbing sounds, and because they were capable of evoking an emotional response in observers. The weeping usually lasted about 30 seconds and was often, but not always, followed by head-turning to the right side. During the attacks the patient was inattentive to his environment and unresponsive to command; afterwards he was slightly confused for a few seconds. The patient was amnesic for events occurring during these episodes. At other times he experienced periods of giggling which lasted a few

seconds. Except for these paroxysmal disturbances, the patient was pleasant and co-operative. His behaviour was not grossly inappropriate nor was his affect otherwise labile.

Skull radiographs were normal. Four vessel cerebral angiography revealed diffuse intracranial atherosclerotic changes and evidence of lateral ventricular enlargement. While pneumoencephalography demonstrated enlargement of both lateral ventricles, the right frontal and temporal horns were considerably larger than those on the left. The cerebrospinal fluid VDRL test was reactive, although the fluid was acellular and had a protein concentration of 0.57 g/l.

When the dosage of anticonvulsant medication was increased, this patient's seizures eventually stopped. The focal motor episodes proved easiest to control; the laughing and crying spells were more refractory to treatment. However, in order to document the ictal nature of our patient's crying and to verify that his emotional lability did not just reflect bihemispheric disease, we performed a pentylene-tetrazol test. The baseline EEG (Fig. 1) had shown a

FIG. 1 Bipolar EEG recording shows disorganization of basic pattern on right side with absence of alpha rhythm. The pattern consists of theta with some delta activity. The left side shows alpha rhythm with occasional theta activity. Eye blink artefact present in first two channels.

diffusely abnormal record with asymmetry of the intrinsic pattern of electrical activity over the two hemispheres. The most organized tracings were recorded from the posterior region of the left side and consisted of alpha, theta, and beta waves. The homologous area on the right side was dominated by theta and delta components; alpha activity was not present.

With continuous 16 channel EEG and cinematographic monitoring, we administered pentylenetetrazol intravenously at a rate of 50 mg/min. When 100 mg of the drug had been infused, a low voltage (50 μ V) rhythmic 2.0 Hz delta rhythm developed in the right posterior temporal-parietal-occipital region and subsequently rose in amplitude to 150 μ V. Concurrent with the inception of the delta activity, the patient's eyelids began to quiver. Fifteen seconds later this was followed by a left-beating horizontal nystagmus. When 175 mg of the convulsant drug

had been administered (and the injection terminated) the patient began to cry (Fig. 2). This was accompanied by lacrimation and appeared to be identical with the spontaneous episodes observed previously. At the same time we observed in the EEG an electrical seizure characterized by diffuse rhythmic high voltage (50–100 μ V) slow (1.5–2.0 Hz) discharges (Fig. 3). The patient continued to weep for 40 seconds, then the eyes deviated to the right, the left facial musculature contracted tonically, and the left-beating nystagmus returned. The seizure seemed to end when the facial musculature relaxed and the patient salivated; but there was a second episode characterized by rightward ocular deviation, sustained tonic contraction of the left face followed by deviation of the head to the left and clonic activity of the left arm. At this point we terminated the episode with 10 mg intravenous diazepam. The patient was completely amnesic for the event.

FIG. 2 Serial photographs of patient during pentylenetetrazol-induced seizure (times indicated are after start of infusion). (1) Control; (2) Two minutes, 15 seconds. (3) Three minutes, 30 seconds. Crying began. (4) Three minutes, 50 seconds. (5) Four minutes. Crying accompanied by sobbing and tears.

FIG. 3 Same EEG montage as Fig. 1. After pentylenetetrazol (175 mg) was administered intravenously, rhythmic slow wave epileptic activity appeared on right side especially in posterior areas, but also reflected to a lesser extent over the entire cortex.

DISCUSSION

Although pathological crying is a frequent accompaniment of certain types of neurological disease, epilepsy is not usually one of them. But here we have a case of paroxysmal crying which we believe to be epileptic in origin. There is considerable support for our conclusions. For example, the man's weeping appeared spontaneously or in response to administration of the convulsant drug pentylenetetrazol; moreover, his emotional changes and facial expressions were usually inappropriate to his situation or environment. The episodes were accompanied by paroxysmal alterations in the EEG, and frequently by other epileptiform manifestations such as head and eye movements. When the patient regained consciousness he was always confused and amnesic. And most importantly, all these

phenomena ceased when the proper dose of anticonvulsant medication was administered. All of these features indicate a convulsive process.

The precise nature of the underlying pathological process in the present subject is obscure. One possible aetiology is neurosyphilis. Our patient did have both a history of syphilis and a reactive CSF VDRL test. The symptoms of neurosyphilis are not always indicative of diffuse brain disease; focal symptoms are possible (Merritt and Springlova, 1932). Epileptic attacks and focal signs such as hemianopia and hemiparesis can characterize the Lissauer type of general paresis (Lissauer and Storch, 1901). Furthermore, atrophy, particularly of the temporal and parietal lobes, is a prominent feature of this disease. A second possible cause of the patient's weeping is progressive vascular disease.

The angiogram performed on the present admission did show evidence of diffuse vascular pathology, but one performed 14 years previously was normal. Finally, trauma could produce a similar clinical picture, but the lack of a definitive history makes this most speculative.

Involuntary and irresistible crying—so-called forced crying—after organic brain disease is widely recognized as a symptom of several neurological states. One is pseudobulbar palsy, whether it results from amyotrophic lateral sclerosis or is an effect of bilateral cerebral damage from vascular disease (Wilson, 1924; Davison and Kelman, 1939). The syndrome has also been observed in advanced multiple sclerosis, mesencephalic (Wilson, 1924) and hypothalamic tumours (Davison and Kelman, 1939); on occasion unilateral hemispheric tumours, infarcts, or Wilson's disease have been implicated as causative factors. In addition, although it has been often denied, pathological crying frequently accompanies bulbar palsy.

Forced crying may resemble our patient's

condition to some extent in that such periods of crying are involuntary, stereotyped, and uncontrollable. But in contrast with our patient's episodes, forced crying is usually, but not always, precipitated by events or objects in the external environment (even though the precipitating cause may be inappropriate or non-specific). Furthermore, the patients are neither unresponsive during attacks, nor amnesic for them afterward; they do not respond to anticonvulsant medication. Thus, despite certain similarities, it should be possible to differentiate between forced crying and epileptic crying.

Common sense may dictate that laughter and weeping are closely related—they laughed until they cried—but gelastic seizures crop up frequently, while if we are to judge from published accounts, dacrystic seizures must be rare or at least infrequently recognized. Before the present case there have been descriptions of five cases in which pathological crying appeared to have an epileptic basis (Table). Of these five, all but Stutte's (1963) lack sufficient detail. None

TABLE
PATIENTS WITH CRYING SEIZURES

Reference	Age, sex (y)	Clinical features	Pathology or clinical findings
Zilgien (1906)	52 M	Aura: 'dizziness'; then production of whistling sound, fluttering of lids, opening and shutting of mouth, contraction of L face, turning of head to L; finally violent sobbing after which patient brought L hand to mouth and then wiped eyes with handkerchief	Luetic pachymeningitis; softening of R parietotemporal cortex
Purves-Stewart (1927)	11 F	No aura; uncontrollable attacks of weeping culminating in loss of consciousness and falling	Glioma of interpeduncular space infiltrating upper pons, third nerve, and optic chiasm
Davison and Kelman (1939)	51 M	Aura: dimming of vision and vertigo; then tonic deviation of eyes to R; pupil fixed and dilated; then uncontrollable weeping lasting 2–15 min; postictal R-sided dysaesthesias; no loss of consciousness	Encephalomalacia L parietal cortex and medial nucleus of R thalamus
Efron (1961)	28 M	Aura: 'dizziness'; 'throbbing sensation in head'; loss of consciousness and weeping. Later: complex seizures including at various times auditory, olfactory, and formed visual hallucinations: sensory Jacksonian march, crying, crawling on hands and knees	R cerebral arteriovenous malformation
Stutte (1963)	12 M	No aura; deviation of head to R; flexion of extremities; rotation of body to R, crying	Neonatal asphyxia; mental retardation
Present	60 M	No aura; weeping and lacrimation followed by head turning to R; inattention to environment, amnesia	Ventricular enlargement especially of R frontal and temporal horns

L: left. R: right.

provide EEG tracings taken during a weeping episode.

In the absence of more adequate data, any generalizations based on clinical observations have to be offered with obvious reservations. But it is of interest that four of six patients had head and/or eye deviation preceding the episodes of weeping. In most cases, there was amnesia for the crying. Davison and Kelman's case (1939) is an obvious exception because the patient apparently maintained consciousness during his seizures; a postictal affective component was not described.

The dacrystic seizures that are documented vary from simple (Purves-Stewart, 1927; Stutte, 1963) to very complex automatisms (Zilgien, 1906; Efron, 1961). In some instances a definite pathological localization is available; even so, the data do not permit identification of a specific locus responsible for weeping itself. However, as with gelastic seizures, it seems probable that a pathological substrate for dacrystic phenomena will ultimately be demonstrated.

At present, the neurophysiological basis of 'normal' crying is still unknown. It is a complex neurophysiological event which involves the association of affective changes, facial and respiratory movements, and vasomotor and secretory alterations. Clearly many neural structures must participate. But as yet, there is no evidence either for an accurate localization or representation of this complex pattern within the brain. To our knowledge, experimental stimulation of brain structures has not evoked weeping. Lacrimation by itself has been produced, reportedly as part of diffuse autonomic discharges caused by lesions which involved the structures surrounding the third ventricle (Penfield, 1930; Haymaker 1958). Stimulation of the amygdala and other parts of the limbic system (Anand and Dug, 1955) is another source of tearing. The facial patterns associated with crying can probably be elicited by brain stem stimulation (Weinstein and Bender, 1943). However, the most obvious difficulty in understanding crying is the lack of

a suitable experimental model, as humans are apparently unique in this regard. It is not unreasonable to expect that further clinical investigations of patients with crying epilepsy may produce information concerning the underlying mechanisms of crying.

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