Gelastic, quiritarian, and cursive epilepsy

A clinicopathological appraisal

P. K. SETHI AND T. SURYA RAO

From the Departments of Neurology and Neurosurgical Centre, Command Hospital, Lucknow, India

SYNOPSIS A patient is reported with episodes of epileptic laughter, crying, and running occurring alone or in combination. He was found to have a discrete, well-circumscribed tumour of the left temporal lobe. The neurology of these epileptic events is discussed in relation to the pathological lesion.

Laughter as an epileptic phenomenon is extremely uncommon and equally so is episodic running. The combination of these two forms of epilepsy—'gelastic' (laughter) and 'cursive' (running)—in the same individual is rare. There are only two earlier reports, of in all, three cases (Sisler *et al.*, 1953; Chen and Forster, 1973). All these cases resulted from diffuse cerebral disorder. We studied an individual with such episodic events of laughter, running and also crying occurring alone or in combination, associated with a discrete neoplastic lesion in the left temporal lobe. This case permits appraisal of cerebral localization of 'centres' for the physical expression of emotions. Crying as an ictal event has not received much attention in the literature. We coin the word 'quiritarian' epilepsy (*quiritare*: Latin: to cry/scream) to indicate this phenomenon.

CASE REPORT

A 33 year old infantry soldier was evacuated to our neurological centre with a history of episodic seizures for one and a half years. He was observed to have four to six seizures a day, lasting 0.5-1.0 minute. In some of these episodes he would burst into crying, while in others into laughter and sometimes there was a curious mixture of both. The laughter did not have an infectious quality. He was later observed to have, in addition, episodes of running. There was no con-

vulsive movement and he never fell unconscious to the ground. Sometimes these episodes could be induced by hyperventilation, but not invariably. One episode induced by hyperventilation has been documented by movie and serial still photographs (Figs. 1-4). He suddenly burst into laughter, with squeezing of the eyelids (Fig. 1) and, at the same time rubbing his upper abdomen, ran to the other end of the room near the door (Fig. 2), then turned and ran diagonally across with an expression of fear on his face (Fig. 3), followed immediately by a frozen expression of defiance, keeping both hands in the position of boxing, standing against the wall as if ready to fight (Fig. 4). The whole episode lasted for about 90 seconds. Postictally for about 30 seconds he would brush aside any attempts to take his pulse, very arrogantly. After that, he was his normal self with no memory for the whole episode.

There was no history of trauma or any febrile episode in the past and no history of epilepsy in the family. Systemic survey and neurological examination were within normal limits except for a right central type of facial asymmetry. Electroencephalography showed generalized spike discharges during seizures induced by hyperventilation with an epileptogenic focus in the left temporal lobe. Routine laboratory investigations and radiography of the skull did not reveal any abnormalities. Left carotid angiography showed changes consistent with a relatively avascular space occupying lesion in the anterior and middle temporal area (Fig. 5a, b).

OPERATIVE FINDINGS At left temporal craniotomy he was found to have a light yellowish-brown, well-circumscribed, partly amorphous and partly granular tumour, located in the middle and inferior temporal gyri, with compressed oedematous brain acting as a

Address for correspondence: Major P. K. Sethi, Command Hospital, Central Command, Lucknow 226002, India.

(Accepted 21 April 1976.)

FIGS. 1-4 Serial photographs of the patient in one of the epileptic episodes induced by hyperventilation.

FIG. 5 (a) Left carotid angiography anteroposterior and (b) lateral views.

false capsule. The mass extended subcortically inferiorly and medially towards the uncus. The tumour was excised *en masse*. Its total size was $4 \times 4 \times 3$ cm. It extended up to the pole just short of 2 cm, and up to the vein of Labbé. The extent of the tumour in the brain as seen by the operating surgeon is represented diagrammatically in Fig. 6. Histopathological examination revealed it to be an astrocytoma grade I. The patient made an uneventful recovery from the operation and has been asymptomatic on anticonvulsant therapy for the last six months.

DISCUSSION

The pathophysiology of emotional disorder has always fascinated neurologists. Focal lesions are the more interesting as they permit certain insights and surmises into the anatomical substrates of these emotions. Despite the arguments against localization, especially where disturbed behaviour is concerned, this approach has certain practical advantages. Moreover, the recent interest in split-brain functions has renewed interest in the exercise of 'localization'. It is admitted that higher cerebral functions are complex and their anatomical substrate is not as simple as a reflex arc, but this does not mean that they need be

regarded as less bound to structures than the 'lowest' functions.

Yakovlev (1948) in a masterly article on 'motility, behaviour, and the brain' has convincingly deduced on a teleological basis that there are three levels of biological functions in the neuraxis. He ascribed these levels of total neuronal functions to ento-, meso- and ectopallium subserving spheres of motility, serving visceral, expressive and affective functions, respectively. The temporal lobe is in a unique position as it embraces all three systems concerned with these three spheres of motility. In this presentation we are predominantly concerned with the second and third spheres—that is, expression and affect. The second sphere, biologically and morphologically in the middle, is the 'sphere of motility of the outward expression of internal states' (Williams, 1968). The internal states of pleasure or happiness, frustration, pain or grief, fear or terror, may respectively be exteriorized as smiling or laughter, crying or sobbing, attempt at flight (running) or fight. Although these three systems are independent peripherally, centrally they are intimately related and interdependent (Williams, 1968). The temporal cortex is, therefore, in a unique position for an epileptic event there to be exteriorized in

FIG. 6 Diagrammatic representation of the extent of the tumour (hatched areas).

one or more of these three spheres. Therefore, the occurrence of epileptic laughter, crying, and running are easily explained in this patient with a discrete temporal lobe lesion.

Involuntary (pathological) laughter (or for that matter crying) may occur as a release phenomenon due to a destructive lesion, as in pseudo-bulbar palsy, or as an epileptic event (gelastic epilepsy). In distinguishing between them, epileptic laughter is usually a short-lived paroxysm of laughter, with the patient having complete amnesia for the whole event. A patient with 'release' laughter, on the other hand, has longer periods of laughter (actually peals of laughter) and the patient is not only aware of it but, at times, actually embarrassed about his disorder. This laughter may be provoked by the slightest stimulation, while provocation of epileptic laughter may require techniques normally required for provoking epilepsy, such as hyperventilation. The patient with released laughter may have the corresponding emotional component—that is, a feeling of happiness or joy, while it is not known whether a patient suffering from true gelastic epilepsy also feels elated or not as there is total amnesia for the episode.

The question arises whether the temporal lobe is the exclusive site for such epileptic events. A few cases have been reported with temporal lobe tumour (Daly and Mulder, 1957) and several with temporal or frontotemporal electroencephalographic foci (Ironsides, 1956; Druckman and Chao, 1957; Daly and Mulder, 1957; Weil *et al.*, 1958; Fukuyama *et al.*, 1959; Lehtinen and Kivalo, 1965; Roger *et al.*, 1967; Zecchini and Cecotto, 1967). Other reports have described lesions of the basal ganglia, hypothalamic region, and in the wall of the third ventricle (Dott, 1938; Ironsides, 1956; List *et al.*, 1958; Gumpert *et al.*, 1970; Gascon and Lombroso, 1971). Somehow the hypothalamus has come to be regarded as of special importance (Masserman, 1941). Martin (1950) called it a 'laughter centre'. The trend in this direction has been such that in some cases of gelastic epilepsy with a temporal lobe lesion, indirect pressure on the hypothalamus was implicated as a cause of epilepsy due to the proximity of the infero-medial surface of the temporal lobes to the hypothalamus. The same arguments might well be used in the reverse direction to explain at least some of the cases of gelastic epilepsy

originally attributed to a hypothalamic lesion.

Careful study of some of the other reported cases of gelastic epilepsy associated with a lesion at another site shows that there was involvement of the temporal lobe as well. Two examples may be quoted. In the case reported by Wood *et al.* (1958), gelastic epilepsy occurred in association with a neurofibroma of the trigeminal nerve which had also deformed the inferomedial aspect of the left temporal lobe. In a case of gelastic epilepsy reported by Weil *et al.* (1958), a papilloma of the third ventricle had extended to the temporal lobe. Furthermore, a careful study of many of the reported cases of laughter occurring with a hypothalamic disorder and some of those of basal ganglia and midline structures, such as the third ventricle, reveals as has been rightly pointed out by Swash (1972) that the laughter in these cases is more like the pseudo-bulbar type. Another argument in support of this contention is that one generally regards epilepsy as a cortical phenomenon. The hypothalamus is only one of a series of stations concerned with control of emotional feeling and behaviour (Alpers, 1940). Therefore, we feel that the role of the hypothalamus in the genesis of gelastic epilepsy is over-emphasized in the literature. At the same time, it has to be conceded that our explanations do not account for all the reported cases of gelastic epilepsy. It seems that other structures in the limbic system may also at times give rise to similar phenomena, but the temporal lobe, because of its unique biological and morphological endowments, is in an especially unfavourable position for these epileptic events.

With regard to lateralization, the left temporal lobe seems to be a favoured site. Poeck and Pilleri (1963) found a left-sided lesion in six cases and right-sided lesion in four cases. The tumour in the present case was also on the left side.

In laughter as well as in crying, in addition to a play of specific facial muscles, a series of respiratory arrests and accelerations occur. Two facio-respiratory pathways have been postulated. Neural mechanisms involved in emotions like laughter and crying seem to have a higher control with its integrating mechanisms and a 'final executive pathway' for the corresponding emotion. The latter is dependent on neural mechanisms arising in close association with telencephalic and diencephalic centres concerned

with respiration (Haymaker, 1969). The higher mechanism has already been discussed briefly. The whole concept fits well with the concepts of Papez (1937) and MacLean (1955). In their scheme, the temporal lobe acts as higher integrating cortex within the limbic apparatus and is closely connected through this system with hypothalamic cingulate, and brain stem pathways. Theoretically, a strategically placed lesion anywhere in these pathways may give rise to involuntary laughter and crying. This may occur as a release phenomenon (removal of inhibition) or as an epileptic discharge (facilitation) as in the present case. Released involuntary laughter is generally due to bilateral lesions, though, rarely, a unilateral lesion may give rise to it (Swash, 1972).

Crying as an epileptic phenomenon has received little attention in the literature. Most authors dealing with temporal lobe epilepsy emphasize that emotions occurring as warning of minor attacks are unpleasant (Penfield, 1955). Thus crying as compared with laughter would be expected to be a more common epileptic event. In crying, as in laughter, a series of respiratory accelerations and arrests occurs in addition to a different play of facial muscles. The neuro-anatomical substrates of 'final executive pathways', although akin to that of laughter, cannot be the same as the total outward expression is of two entirely different moods. Not only do specific facial muscles related to a specific emotion come into play, but the pattern of respiratory acceleration and arrests may also be different, to account for such things as sobbing. In addition, the neural cells responsible for highest controls must necessarily be distinct, though closely related anatomically to explain why a specific emotion evokes a specific facial expression. In spite of this, there may be some common neuronal mechanisms sharing a number of synaptic pathways in the midbrain and brain stem, or interconnections of these two sets of pathways, or both. This would explain why some normal people at times exhibit incongruity of emotional expression, such as feeling so happy that they burst into tears and *vice versa*, or laughing themselves into crying. This might explain some of the episodes in which the patient reported here laughed or others in which he cried or in which there was a curious mixture of laughing and crying.

Only a few of the reported cases of running (cursive) epilepsy have had an associated pathological study (Lennox, 1960; Chen and Forster, 1973). Chen and Forster (1973) reviewing the available EEG findings in all reported cases concluded that a temporal lobe focus is essential for the genesis of running epilepsy. We concur completely with their conclusions because of the unique position of the temporal lobe to be the site of these epileptic discharges. Running may be regarded as a dynamic physical expression of an internal state of fear.

The inferior and medial portion of the temporal lobe has come to be regarded as a seat of emotions. In this case the tumour also extended inferiorly and medially. The precise location of laughter, crying, and running individually in the temporal lobe is uncertain. The clinical information in our case is not sufficient for such precise localization but the role of the temporal lobe in producing these epileptic discharges is clearly demonstrated.

We are grateful to Col A. K. Ghose, Officer Commanding, and the Director General, Armed Forces Medical Services, for permission to publish this paper, and to Sister Mary Ross, MA, BEd, for advice on Latin etymology.

REFERENCES

- Alpers, B. J. (1940). Personality and emotional disorders associated with hypothalamic lesions. In *The Hypothalamus. Proceedings of the Association for Research in Nervous and Mental Diseases*, 20, 725-752.
- Chen, R., and Forster, F. M. (1973). Cursive epilepsy and gelastic epilepsy. *Neurology (Minneapolis)*, 23, 1019-1029.
- Daly, D. D., and Mulder, D. W. (1957). Gelastic epilepsy. *Neurology (Minneapolis)*, 7, 189-192.
- Dott, N. M. (1938). Surgical aspects of the hypothalamus. In *The Hypothalamus, Morphological, Functional Clinical and Surgical Aspects*, pp. 179-181. Edited by W. E. Clark, Beattie, J., Le Gros, G. Riddoch, and N. M. Dott. Oliver and Boyd: Edinburgh
- Druckman, R., and Chao, D. (1957). Laughter in epilepsy. *Neurology (Minneapolis)*, 7, 26-36.
- Fukuyama, S., Arima, M., Nagahata, M., and Okada R. (1959). Pathological laughter as an epileptic manifestation. *Recent Advances in Research in the Nervous System (Tokyo)*, 3, 675, Cited by Chen, R., and Forster F. M. (1973). *Neurology (Minneapolis)*, 23, 1019-1029.
- Gascon, G. G., and Lombroso, C. T. (1971). Epileptic (gelastic) laughter. *Epilepsia*, 12, 63-76.
- Gumpert, J., Hansotia, P., and Upton, A. (1970). Gelastic epilepsy. *Journal of Neurology, Neurosurgery, and Psychiatry*, 33, 479-483.

- Haymaker, W. (1969). In *Bing's Local Diagnosis in Neurological Diseases*, pp. 167. Mosby: St. Louis.
- Ironside, R. (1956). Disorder of laughter due to brain lesions. *Brain*, **79**, 589–609.
- Lehtinen, L., and Kivalo, A. (1965). Laughter epilepsy. *Acta Neurologica Scandinavica*, **41**, 255–261.
- Lennox, W. G. (1960). *Epilepsy and Related Disorders*. Vol. 1, pp. 280. Little Brown: Boston.
- List, C. F., Dowman, C. E., Bagchi, B. K., and Bebin, J. (1958). Posterior, hypothalamic hamartomas and gangliogliomas causing precocious puberty. *Neurology (Minneapolis)*, **8**, 164–174.
- MacLean, P. D. (1955). The limbic system ('visceral brain') and emotional behavior. *Archives of Neurology and Psychiatry (Chic.)*, **73**, 130–134.
- Martin, J. P. (1950). Fits of laughter (sham mirth) in organic cerebral disease. *Brain*, **73**, 453–464.
- Masserman, J. H. (1941). Is the hypothalamus a centre of emotions? *Psychosomatic Medicine*, **3**, 3–25.
- Papez, J. W. (1937). A proposed mechanism of emotion. *Archives of Neurology and Psychiatry (Chic.)*, **38**, 725–743.
- Penfield, W. (1955). The twenty-ninth Maudsley lecture: The role of temporal cortex in certain psychical phenomena. *Journal of Mental Sciences*, **101**, 451–465.
- Poeck, K., and Pilleri, G. (1963). Pathologisches Lachen und Weinen. *Schweizer Archiv für Neurologie und Psychiatrie*, **92**, 323–370.
- Roger, J., Lob, H., Waltregny, A., and Gastaut, H. (1967). Attacks of epileptic laughter: on 5 cases. *Electroencephalography and Clinical Neurophysiology*, **22**, 279. (Abstract).
- Sisler, G. C., Levy, L. L., and Roseman, E. (1953). Epilepsia cursiva, Syndrome of running fits. *Archives of Neurology and Psychiatry (Chic.)*, **69**, 73–79.
- Swash, M. (1972). Released involuntary laughter after temporal lobe infarction. *Journal of Neurology, Neurosurgery, and Psychiatry*, **35**, 108–113.
- Weil, A. A., Nosik, W. A., and Demmy, N. (1958). Electroencephalographic correlation of laughing fits. *American Journal of Medical Science*, **235**, 301–308.
- Williams, D. (1968). Man's temporal lobe. *Brain*, **91**, 639–654.
- Wood, M. W., Svein, H. J., and Daly, D. (1958). Involuntary laughter. *Proceedings of the Mayo Clinic*, **33**, 266–275.
- Yakovlev (1948). Motility, behavior and the brain. *Journal of Nervous and Mental Disease*, **107**, 313–335.
- Zecchini, A., and Cecotto, C. (1967). Crisi parossistiche di riso e di natura epilettica. *Minerva Medica*, **58**, 3405. Quoted by R. Chen and F. M. Forster (1973). *Neurology (Minneapolis)*, **23**, 1019–1029.