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Epileptic laughter with precocious puberty

Gelastic (laughing) epilepsy may be defined as complex co-ordinate movements with grinning, giggling, or joyful weeping. The disorder is rare and may arise from lesions in the temporal lobe, limbic system, or hypothalamus. We report a case of gelastic epilepsy which preceded the onset of precocious puberty.

Case report

A girl aged 2.2 years was referred with a long history of bouts of unusual behaviour. The perinatal and neonatal period were normal. From age 3 weeks she had had repeated episodes of crying during which she

would draw up her knees, clench her fists, and micturate frequently. As the child grew older the nature of the episodes changed; they would begin with a characteristic cry or giggle, she would then frown, appear vacant, and become pale and there would be slow rotatory eye movements, drooping of the eyelids, and repeated blinking. These episodes lasted a few seconds only but would recur about every 2 min for up to 12 hours. Throughout them she would be overexcited, aggressive in her behaviour, and repeatedly incontinent of urine. Once free of the attacks she would drink excessively and sleep for between 12 and 24 hours. Otherwise she was a happy, contented child with no other problems.

Examination was normal. However, the EEG showed paroxysmal moderate to high amplitude sharp and slow wave complexes in all areas but these were particularly pronounced in the central region. An air encephalogram showed slight hydrocephalus and a space-occupying lesion on the floor of the 3rd ventricle. On ventriculography a tumour occupied the whole of the floor of the 3rd ventricle. A biopsy was attempted but the tumour was too hard and fibrous to get adequate material. A bilateral ventriculocisternostomy (Torkildsen's operation) was performed to lessen the intraventricular pressure. Radiotherapy and anticonvulsant drugs were given and during the next year there were fewer fits.

During her 3rd year of life the child developed precocious puberty. On examination height was 108.6 cm (+2.1 SD) and weight 22 kg (>97th centile), pubertal development was Tanner P₄G₃ with menarche. Bone age 8.7 years. Plasma gonadotrophins were in the pubertal range; basal luteinising hormone (LH) 11.4 U/l, and follicle stimulating hormone (FSH) 4.4 U/l, after 100 μ g intravenous luteinising hormone releasing hormone (LHRH) plasma LH rose to a peak greater than 50 U/l and FSH to 24.5 U/l. Plasma oestradiol was 150 pmol/l (41 pg/ml). Growth hormone, adrenocortical, and thyroid studies were all normal. She was treated with cyproterone acetate 50 mg three times a day. Clinically, sexual maturation slowed and she lost her axillary and pubic hair. Height has continued to increase and her bone age at a chronological age of 5.8 years was 10.8 years.

At 5 years the seizures increased in frequency to at least one an hour. She would giggle or hiccup and this was followed by a brief period of apparent insecurity and uncertainty about her surroundings. Initially, these episodes occurred when she was tired or emotionally upset but later they became more common and were associated with deterioration in her school work and refusal to leave the house to play with her friends. The episodes have responded well to clonazepam 2.5 mg daily and this has

Table *Reports of cases of gelastic epilepsy and precocious puberty*

Authors	Date	Sex	Age at onset (months)		Clinical findings	Disease
			Of seizures	Of precocious puberty		
Dott	1938	F	Birth	6	Autistic; death, aged 8 years, of unrelated cause	Astrocytoma in region of mammillary bodies
List <i>et al.</i>	1958	F	2	33	Obesity, episodes of high fever; death at 15 years, of unrelated cause	Hamartomatous mass attached to mammillary body
Money and Hosta Case 1	1967	M	3	24	Normal EEG and AEG facial flushing*	Idiopathic
2	1967	M	81	24	Attacks of hiccups, facial flushing. Abnormal EEG; normal AEG, and brain scan*	Idiopathic
Bierich <i>et al.</i> (reported in Money and Hosta)	1967	M	72	Birth	Mental retardation*	Tumour on floor of 3rd ventricle
Gascon and Lombroso	1971	F	24	48	Mental retardation*	Hypothalamic astrocytoma

*Alive at publication of case.

improved her school performance, and increased her security and happiness.

Discussion

Laughter is a complex phenomenon involving a variety of sensory stimulants—visual, auditory, tactile, and memory. Affectively it involves the muscles of facial expression and respiration. Laughing seizures are characterised by the sudden onset of involuntary laughter which may be associated with a variety of repetitive movements. The laughter is inappropriate, stereotyped, and may be muted or uproarious. The prolonged attacks of giggling and other movements in our patient appears to have been gelastic status epilepticus. This has been reported previously in two cases (Gumpert *et al.*, 1970).

Lesions of the temporal and frontal lobes, and the hypothalamus have been associated with epileptic laughing (Ironside, 1956). The limbic system unites areas on the cerebral cortex with the hypothalamus and midbrain, and it seems probable that lesions in any part of this system may result in gelastic epilepsy.

Differences have been found in the nature of the epileptic laughter according to the site of the lesion. Diencephalic lesions result in laughter without affect, whereas laughter due to temporal lobe lesions has been associated with an affective component (Gascon and Lombroso, 1971).

Childhood gelastic epilepsy is often caused by a tumour of the posterior hypothalamus. Onset in the neonatal period is rare. In four previous case reports Sher and Brown (1976) recommend a conservative approach to treatment unless there is evidence of an enlarging mass or neurological deterioration.

The mammillary bodies and posterior hypothalamus are recognised as being involved in the synthesis and secretion of LHRH and therefore in the control

of puberty. The association of gelastic epilepsy and precocious puberty has occasionally been reported (Table). Although a number of lesions in this area are hamartomas, it is clear that the onset of gelastic epilepsy in childhood must be recognised as of potentially serious significance.

Summary

A child with associated epileptic laughter and precocious puberty is reported. The significance of epileptic laughter as a possible marker of hypothalamic disease is noted.

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