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症　　例

Precocious Puberty with Fits of Laughter and with a Large Cystic Mass on the Floor of the Third Ventricle (Case Report)

by

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INTRODUCTION

Precocious puberty with fits of laughter is a rare clinical phenomenon. In reviewing literature, MONEY and HOSTA (1967)[1] stated that they found out only 2 previously reported cases (the case of DOTT, et al. (1938)[2] and of LIST, et al. (1958))[3] and added their 2. BIERCH, et al. (1967)[4] also reported one case and in Japan, there have been 2 cases[5][6] published. We are reporting another case recently encountered in which radiological examination revealed presence of a cystic mass of a size of golf-ball on the floor of the third ventricle.

CASE REPORT

K.F., a male, aged 9, was born on May 3, 1959 and admitted to the Neurosurgical Clinic, Kyoto University Medical School on April 8, 1968. He was the first child of healthy parents, without any familial background of neurological disorder. The patient had been a full term infant, delivered spontaneously. There were no childhood diseases or history of meningitis.

At the age of 3 years, his mother noticed the enlargement of penis and also about that time, he had the first generalized convulsions. Thereafter, unusually rapid skeletal growth with marked development of genitalia had been noted. At age 6, there developed pubic hair. From the age of 8 years and 7 months, he began to have attacks of uncontrollable laughter which occasionally followed by generalized convulsions.

When admitted, at the chronological age of 8 years 11 months, he had the appearance of a 14~15 year adolescent boy. The head was comparatively large with circumference of 57 cm., height 153 cm. (against 121.9 cm. by normal at his age) and body weight 45 kg. (against 23.2 kg.). The bone age was between 13 and 14 years. The external genitalia were adult type with a marked development of penis and testes accompanied by growth of the pubic hair (Fig. 1). Physical features were otherwise normal and his

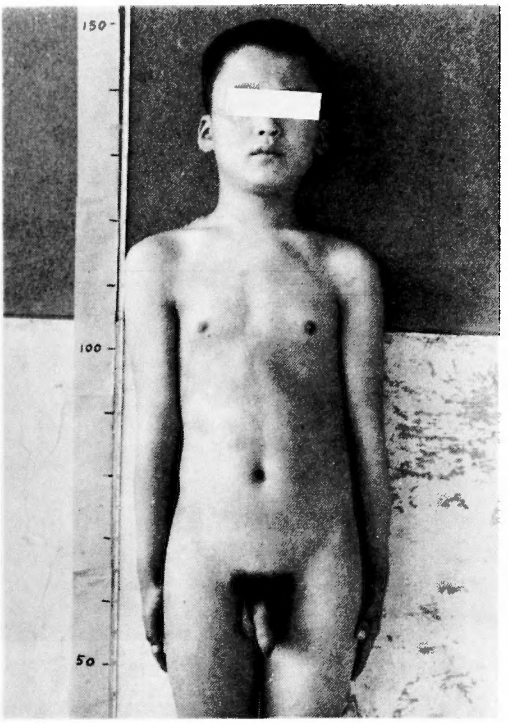

Fig 1

mental activity seemed slightly retardated. IQ was 71 (WISC). Neurological examination disclosed no abnormality including negative Parinaud's sign.

Laboratory examination : Table 1 summerizes the various results obtained by laboratory examinations. The cerebrospinal fluid was normal. The successive measurements of 17-ketosteroid in urine also gave normal results (between 1.3 mg. to 3.98 mg. in 24 hours). Friedmann' reaction in urine was negative (50 k.u.) and estrogen amounted to 5.8 γ/24 hours.

Electroencephalogram : Interseizure EEG showed paroxysmal bursts of medium voltage 5 to 7 per second waves which occurred synchronously in all leads with emphasis on parietal and occipital regions, but no definite spike pattern was seen.

Radiological examination : Skull plain radiograms were normal. Air encephalogram showed moderate dilatation of the lateral and third ventricles. The chiasmal and inter-

Table 1

1. Hematological examination
   R.B.C. 481×10^4 Hb 87% W.B.C. 4,700 thrombocytes 25.4×10^4 coagulation time 10'
2. Urinalysis
   volume 500~1,800 ml/day specific gravity 1017
3. Liver function test
   icterus index 3 Co 3 Cd 7 T.T.T 1 $ZnSO_4$ 8
4. Blood chemistry
   alkaliphosphatase 18.5μ acidphosphatase 2.5 cholesterol ester 113 mg/100ml GOT 29.5μ
   GPT 19.0μ blood glucose 88 mg/100ml serumprotein 4.22 mg/dl
5. Electrolytes
   Na 138 mEq/L K 4.0 mEq/L Cl 97 mEq/L Ca 10.0 mg/100ml
6. Immunological examination
   ASLO 625μ CRP (−) RAT (−) serumprotein 7.2 g/100ml A/G=1.28
   Alb. 56.2% α-glob 10.7% β-glob 11.6% γ-glob 21.5%
7. CSF examination
   watery clear pressure 140 mmH_2O Nonne Apelt (−) Pandy (−)
   protein 33.0 mg/100ml glucose 36 mg/100ml cell counts 5/3
8. Hormonal test
   urinary excretion of 17-KS 1.3~3.98 mg/day
   urinary excretion of 17-OHCS total 1.9~9.8 mg/day free 0.01~0.127 mg/day
   urinary excretion of estrogen 5.8 γ/day metopiron test normal
   feed back test suppressed by 1mg dexamethasone triosorb-resin 30.9 % (normal)
   urine Friedman' reaction 50 k.u. (negative)

peduncular cisterns were not obliterated and there was no finding to be suspected of a mass in the pineal region (Fig. 2).

In order to visualize selectively the anterior part of the third ventricle, a rubber catheter was introduced in direction to the third ventricle through the foramen of Monro. After confirming drainage of fluid (which, at that time, was thought to be the cerebrospinal fluid and no detailed examination of fluid was made), a small amount of positive contrast media (60% Meglumin iothalamate solution) was injected through the catheter. It was clarified that the tip of the catheter was not in the third ventricle but in a cavity and a large cystic mass estimated to be a size of golf-ball, located on the floor of the third ventricle (Fig. 3). In the lateral view, a part of the cavity was filled with air (Fig. 4).

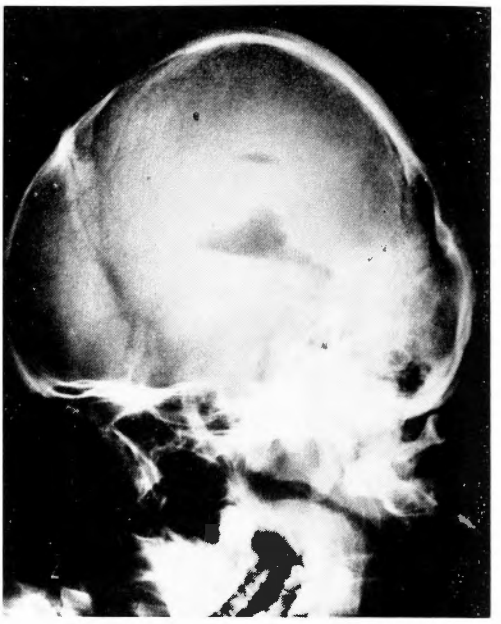

Fig. 2

Fits of laughter: While he was under admission, series of attacks of laughter were observed by the author. Each attack came on without any recognizable reason. Duration

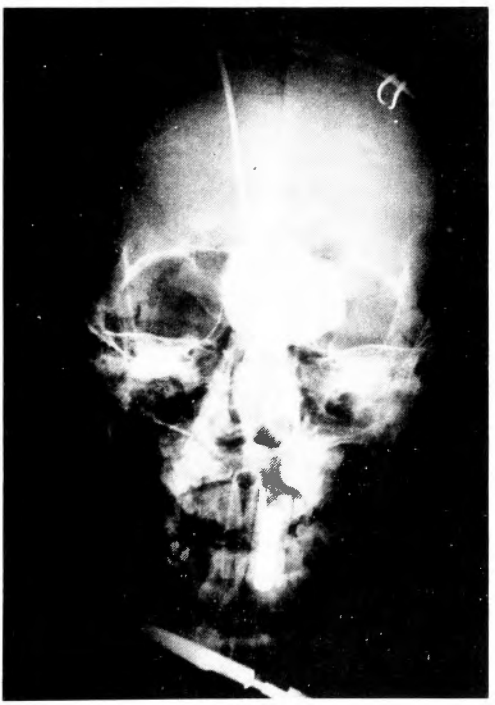

Fig. 3

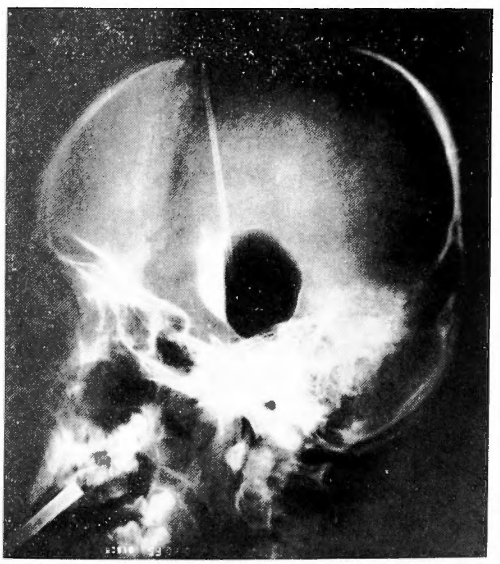

Fig. 4

of the whole episode was less than 30 seconds and the attack occurred as often as 2 to 3 times a day. While he was laughing, his eyes were fixed and vascular flushing of the face and myoclonic twitching of the cheek muscles were often noticed. There was no definite sign of loss of attentive contact with his environment, though, he could not continue talking. He could keep standing and also could respond to simple orders such as to rotate his head to one direction. On most occasions, he did not seem to feel any emotion appropriate to laughter. Immediately after the seizure, we often inquired him as to what had happened, and the boy said that there had been nothing funny, but he had been unable to stop laughing. Only on several occasions, it was also noted that the boy began to laugh with the word "funny". However, even in such instances, there was no evidence that, apart from his laughter, his behaviour exhibited any sign of amusement and such fits of laughter were often followed by the generalized convulsions which indicated strongly that laughter occurred as a part of an epileptic fit.

Clinical course : He showed abnormal sexual interests which seemed exclusively heterosexual. He cast his eyes at a nurse, often attempted to tackle her from the rear. At times, erection and nocturnal emission were noted. His affection was instable with a tendency to emotional outbursts. He was averse to being examined by a doctor and acted violently to his mother.

The operative manipulation to the cystic mass was refused and the patient was discharged in July 1968.

DISCUSSION

Precocious puberty with fits of laughter is a rare clinical phenomenon. Furthermore, only in 2 (the case of Dott and of List) of these reported cases, the precise location and nature of pathology have been ascertained by autopsy. Both were hamartoma in the hypothalamic region. In other cases, however, except that of Bierch, et al. in which some supporting findings were demonstrated in selective pneumoencephalogram, an assumption was made that the site of the lesion might be at or near the hypothalamus mainly from the clinical evidence of sexual precocity coupled with fits of laughter.

In our case, as a large cystic mass was clearly demonstrated on the floor of the third ventricle, it would seem far more likely to assess the direct hypothalamic involvement (presumably chronic irritable state of the hypothalamus) which was responsible for provoking both conditions. As regards to the histological nature of the mass, nothing can be stated for the present.

CONCLUSIONS

One rare case of sexual precocity with fits of laughter was presented. Radiological examinations revealed that a cystic mass of a size of golf-ball located on the floor of the third ventricle.

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和文抄録

笑い発作を伴ない第三脳室底に Cystic Mass を認めた青春早発症（Pubertas Praecox）の症例

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8才11ヵ月の男子，3才より性器および身体の発育が目だち，一方，同じころ全身性痙攣発作が初発．さらに8才7ヵ月よりは特異な笑い発作をも明らかに認めるようになり，感情障害と軽度の知能障害を伴なつていた．X線検査により第三脳室底にゴルフ球大のcystic massがあり，視床下部の直接の障害（慢性刺激状態）に基づくPPおよび笑い発作であることを強く示唆していた．

笑い発作を伴なうPPの症例報告は極めて稀でMoney及びHosta（1957）は1956年より1966年までの文献を渉猟して既報告例は，わずかに2例にとどまるとのべ自験例の2例を追加しているにすぎない．また間脳性PPの成因に関して多くの示唆を与える症例と考える．