over, we could not find another example of the uniformly expansile intradiploic cystic appearance of the temporal bony lesion seen in our patient. A similar appearance has been reported by Abdelwahab et al.,¹³ but their patient had an occipital bony lesion. CT has been recognised to be the mainstay of diagnosis of this entity,⁹ particularly when it reveals the "ground glass" appearance in typical locations. Whereas this observation appears generally valid, there may be a role of MR imaging in certain uncommon situations. For example, in cystic fibrous dysplasia, the radiolucent appearance especially in long bones may mimic malignant lesions such as intraosseous sarcomas.¹⁷ MR imaging clearly distinguishes these 2 entities.¹² In our patient, both the CT and MR studies unequivocally suggested a benign, long-standing disease process. Numerous entities need to be considered in the differential diagnosis of such intradiploic cystic lesions, including aneurysmal bone cyst, unicameral cyst, nonossifying fibroma, osteochondroma, giant cell and eosinophilic granulomas, intradiploic epidermoids, osteoblastomas as well as sarcomatous neoplasms.8,9,11,14,15

Cystic degeneration is an uncommon but well-described phenomenon in fibrous dysplasia. Cystic degeneration may place these lesions somewhere along the spectrum between simple and aneurismal bone cysts, as suggested by Ferretti et al.¹⁸ The clinical importance of this phenomenon in fibrous dysplasia is the relatively rapid increase in size that may be observed, and was also evident in our patient. This may lead to the lesion being misdiagnosed on clinical grounds as an aneurysmal bone cyst, simple cyst or even sarcomatous change in a pre-existing benign bony tumour. Cystic degeneration in fibrous dysplasia needs to be better recognised as a cause for this.

The management clearly rests on the recognition of this entity as a benign one. This may not always be straightforward, with atypical clinical and radiological presentations such as cystic change. Although it has been reported that there is no firm evidence for clinical arrest or resolution with onset of bony maturation,⁴ most do recommend postponing surgical intervention till late adolescence.^{4,5,15} Even then, it should be limited to cosmetic debulking.¹⁹ In case of diagnostic doubt, or when considering a potentially malignant lesion it makes sense to achieve a histological diagnosis. This should be especially borne in mind if one is contemplating any radical surgery for a suspected malignant swelling. Even when one is suspecting monostotic fibrous dysplasia, it is recommended to have a biopsy.^{4,5,15} In the polyostotic form, the multiple lesions make a presumptive diagnosis of fibrous dysplasia much more accurate. Additional measures necessary for all patients having fibrous dysplasia of the temporal bone include periodic otological examinations, owing to the high probability of compromise of the external canal, and other secondary complications.⁵ It should be noted that there is no role for radiation therapy, owing to the high incidence (upto 44%) of malignant transformation.²⁰

To conclude, cystic fibrous dysplasia should be considered in the case of any temporal intradiploic expansile lesion. Cystic degeneration may result in the rapid growth, and this may be misleading and result in suspicion of malignant transformation. Currently available CT and MR imaging techniques help in clarifying the benign nature of the disease and distinguishing this benign disorder from other numerous and malignant lesions with somewhat similar appearance.

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Dyke-Davidoff-Masson syndrome manifested by seizure in late childhood: a case report

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Summary The patient was a 19-year-old woman who presented with hemiatrophy and diminished superficial sensation on the left side of her body including her face. She had a past history of tonic-clonic seizures accompanied by left hemiparesis in late childhood.

Brain CT demonstrated dilatation of the frontal sinus, calvarial thickening, cerebral hemiatrophy and dilatation of the lateral ventricle on the right side. Brain MRI showed atrophy of the right cerebrum and midbrain and dilatation of the lateral ventricle on T1-weighted images, as well as a high signal intensity area from the parietal to the occipital lobe on T2-weighted images. These findings are suggestive of an episode that may have caused a transient ischemia through the right cerebral hemisphere after the intrauterine period.

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INTRODUCTION

Dyke–Davidoff–Masson syndrome (DDMS) was first reported as a rare congenital malformation in 1933 by Dyke et al.¹ Major signs of this syndrome are cerebral hemiatrophy, contralateral hemiplegia and convulsions. Bone malformations, such as dilatation of the frontal sinus on the atrophied side and thickening or thinning of the calvarium, accompany this condition. We recently encountered a case of DDMS manifested by tonic–clonic seizure with left hemiparesis in late childhood.

CASE REPORT

The patient was a 19-year-old woman who complained of hemiatrophy and decreased superficial sensation on the left side of her body. Family history was unremarkable. She was born term delivery, weighing 3020 g and with a length of 50.5 cm. She had a normal course of development until reaching school age, although she had a slightly small build. Scoliosis began to be noted at age 9. At 10 years old, she experienced a tonicclonic seizure, lasting about 1 h. Soon after this episode, left hemiparesis was noticed and brain CT revealed atrophy of the right cerebral hemisphere. When she was assessed using the new K-type development test, her developmental quotient score was 60, suggesting developmental arrest. A few months later, she became aware of the atrophy and reduced sensitivity on the left side of her body including her face. Her epilepsy was well controlled with oral anti-epileptic drug therapy. When she visited our hospital in June 1999, hemiatrophy and diminished superficial sensation were perceptible on the left side of her body including her face. She was admitted for further examination. On admission, she was 149 cm tall and weighed 47 kg. Atrophy of the left side of her body including her face and marked scoliosis toward the right were observed. The maximum circumference of her left extremities was 2.0 cm smaller than that of her right extremities. Neurological examination showed left moderate hemiparesis and diminished superficial sensation of the left side of her body including her face. Although her deep tendon reflex (DTR) was normal, she showed positive pathological reflexes such as Babinski and Chaddock reflex on her left side. Barré sign was positive on the left side. Cerebellar examination was normal, she was able to stand and walk by herself, but her gait was unstable. Laboratory data including rheumatoid factor, anti-nuclear antibody and anti-phospholipid antibody were normal. Cerebrospinal fluid findings were



Fig. 1 Brain CT (axial section) demonstrated cerebral hemiatrophy (A), dilatation of the frontal sinus, and calvarial thickening on the right side (B).



Fig. 2 Atrophy of the right cerebrum and dilatation of the lateral ventricle on T1-weighted images (coronal section) (A). High signal intensity area in the deep white matter from the parietal to the occipital lobe (B), and cerebral cortical sulci and perimesencephalic cistern were more marked on the right than the left on T2-weighted images (axial section) (B and C).

normal. Myelin basic protein and oligoclonal IgG band were negative. Electrophysiological studies including electroencephalogram, acoustic brainstem response, nerve conduction studies and electromyograms showed no abnormal findings. Spinal radiography revealed scoliosis convex toward the right with a Cobb's angle of about 60°. Plain skull radiography showed a slight decrease in the skull volume and slight elevation of the upper edge of the right orbita. Brain CT demonstrated dilatation

of the frontal sinus, calvarial thickening, cerebral hemiatrophy and dilatation of the lateral ventricle on the right side (Figs. 1A and B). Brain MRI revealed atrophy of the right cerebrum and midbrain and dilatation of the lateral ventricle on T1-weighted images (Fig. 2A), as well as a high signal intensity area in the deep white matter from the parietal to the occipital lobe on T2weighted images (Fig. 2B). The cerebral cortical sulci and perimesencephalic cistern were more marked on the right than the left (Figs. 2B and C). When examined by brain SPECT using ¹²³I-IMP, the mean cerebral blood flow (CBF) was 30.4 ml/100 g/min on the left side and 23.2 ml/100 g/min on the right side, indicating reduced blood flow through the right cerebral hemisphere. Head and neck MRA and 3D-CT angiography revealed no abnormalities. The cervical-thoracic MRI revealed no abnormalities. Chest CT revealed atrophy of the left dorsal muscles on the concave side of the spine which had become scoliotic.

DISCUSSION

DDMS is a malformation first reported in 1933 by Dyke et al.¹ Its major signs are unilateral cerebral atrophy, contralateral hemiparesis and seizures. It is accompanied by an expansion of the frontal sinus on the atrophic side, decrease in the cranial volume, elevation of the petrous pyramid and the upper edge of the orbita, thickening or thinning of the cranial vault, etc. on encephalograms.1 DDMS is a rare malformation and few cases have been reported to date.¹⁻¹⁰ Reports of cases in which brain CT or MRI revealed cerebral hemiatrophy^{11,12} showed that these cases also had features similar to those reported by Dyke et al., dilatation of the frontal sinus, decrease in the skull volume, elevation of the petrous ridge and thickening of the calvarium. This case is noteworthy because of the one sided atrophy which included the face. Hozumi et al. reported a case of DDMS presenting with total hemiatrophy. Because their case showed atrophy of the midbraincerebral peduncle, which seemed to be secondary to Wallerian degeneration, as in the present case. It was thought that unilateral brain atrophy was responsible for the total hemiatrophy of the body in their patient.⁵

DDMS can be etiologically divided into 2 types: the congenital or primary type and the acquired or secondary type. In the former type, some vascular factors during the intrauterine life damage the brain and symptoms appear at birth or shortly thereafter. Regarding the vascular factors responsible for brain damage, Stred et al. reported a case in which coarctation of the midaortic arch reduced the unilateral carotid flow, leading to cerebral hemiatrophy.⁹ In the latter type, trauma, infection, vascular abnormalities and haemorrhage during delivery or in childhood may be responsible for the disease.^{2,7,12} As a factor responsible for the latter type of DDMS during delivery, a case of brain damage caused by forceps delivery has been reported.⁶

The types of causative factors can be inferred from the variable features of the imaging findings of patients with DDMS. The compensatory skull changes, such as calvarial thickening and enlargement of the paranasal sinuses on the atrophied side, suggest that the insult to the brain occurred in early life, usually before 2 years after birth.¹² In the present case, T2-weighted images revealed not only hemiatrophy but also a high signal intensity area in the deep white matter from the parietal to the occipital lobe. Cases with similar features have also been reported by Dix and Cail.¹³ Lee et al. compared MRI of normal and abnormal heads of infants and neonates and reported that a 2 year-old infant with cerebral infarction showed a high signal intensity area on T2-weighted images of the deep white matter of the parietal and occipital lobes, in addition to cerebral hemiatrophy. They inferred that this infant's change probably represented secondary demyelinating change induced by ischemia.14 Sener and Jinkins reported brain MRI showing cerebral hemiatrophy occurring in a 4-year-old child who had developed stroke at age 20 months, and they emphasised the presence of prominent cerebral cortical sulci and perimesencephalic cistern, which were also observed in the present case.⁷ This finding may be interpreted as suggesting that the episode responsible for brain damage took place immediately before or after birth, after completion of the sulcus formation. In other words, if the damage occurs during the embryogenesis, sulci are unlikely to become prominent because the gyri and sulci have not been completed at this stage.^{2,7} Taken together, these radiographic findings suggest an episode which could have caused a transient hypoperfusion in the right cerebral hemisphere had taken place after the late intrauterine period. However, in the present case, symptoms such as seizure and hemiparesis occurred relatively late; the timelag between completion of cerebral hemiatrophy and the onset of symptoms was long. Since then a case manifested by late onset seizure similar to this case has been reported.⁶ Cerebral hemiatrophy in such cases could take several years to complete. As for the mechanism of cerebral hemiatrophy, we speculated that focal ishaemic change within a certain period lowered the neural activity and synaptic plasticity lead to a decrease in neurotrophins, as a result, the poor development rather than atrophy of the cerebral hemisphere might have occurred.^{15,16} Moreover, the brain damage by tonic-clonic seizure at 10 years of age might have accelerated the reduction of neural activity and synaptic plasticity in the present case. Therefore, it is desirable that the data such as nerve growth factor, brain-derived neurotrophic factor, and cerebral blood flow be measured soon after ischaemic episode in patients of DDMS or cerebral hemiatrophy.

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θ Oscillation in the anterior cingulate and β -1 oscillation in the medial temporal cortices: a human case report

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Summary In previous studies we observed γ (30–150 Hz) and β -1 (10-20 Hz) oscillations in the medial temporal lobe (MTL) using subdural electrodes. The β -1 was present during wake and REM sleep while γ was present in all states. Recently we studied a patient (35 years M) with electrodes attached to the anterior cingulate cortex (ACC). This structure showed regular θ (5–6 Hz) oscillations. In the first recording, electrodes were attached to the MTL, that demonstrated the β -1 and γ oscillations. Two months later, electrodes were placed on orbitofrontal and ACC and an all night sleep recording was carried out. The ACC exhibited a highly regular and continuous θ oscillation during wakefulness and REM sleep, but not during NREM sleep. Since this same subject showed β -1 oscillations in the MTL, it is probable that the θ in the ACC is independent of β -1 in the MTL. This single case suggests the existence of two different frequency oscillators in the human limbic system. Elucidating their functional roles will be an interesting challenge for future studies.

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INTRODUCTION

It has been suggested that hippocampal θ rhythm or rhythmic slow activity (RSA)^{1,2} plays an important role in inducing long term potentiation (LTP),3-5 which is considered as a physiological base of memory. However, it has not been clear whether the human hippocampus also manifests θ oscillation. In our previous studies, we reported the presence of γ (30–150 Hz) and β -1 (10–20 Hz) electrocorticogram (ECoG) oscillations in the human medial temporal lobe (MTL) using subdural electrodes.^{6–8} The β -1 oscillation was regular and continuous during wake and REM sleep.⁸ We therefore speculated that β -1 may be functionally equivalent to the hippocampal θ rhythm observed in rodents. Recently, we have studied a patient who had electrodes attached to the anterior cingulate cortex (ACC). This structure exhibited a regular θ oscillation. This was the same patient who previously manifested γ and β -1 oscillations in the medial temporal cortex.

CASE AND RECORDINGS

The patient was a 35-year-old male suffering from partial epilepsy. His first epileptic event occurred at age 14 years and was characterised by abnormal behaviour and a generalised seizure that accompanied a high fever. Subsequent seizures have been poorly controlled by medication, and he has since been admitted to hospitals in *status epilepticus* on several occasions. He was therefore referred to the department of neurosurgery for possible surgical treatment of his epilepsy.

Subdural electrodes were surgically attached to the MTL, but failed to detect an epileptic focus. An all-night sleep recording of MTL activity was made (recording I). The subdural electrode positions of MTL electrocorticogram (ECoG) and the montage employed are indicated in Fig. 1A. Two months later, subdural electrodes were placed on the orbitofrontal cortex and ACC. An all-night sleep recording was also made (recording II). The electrode positions and the montage employed for this recording are indicated in Fig. 1B. Both all-night recordings were made approximately 1 week after electrode placement surgery, when the patient was fully alert and could perform daily hospital activities. In addition to the ECoG, a surface Cz-A1 electroencephalogram (EEG), an oblique electrooculogram (EOG), and a chin electromyogram (EMG) were continuously recorded to identify sleep stages. By the second EEG recording and a recording during the surgical operation, the patient's epileptic focus was tuned out to be on the right frontal lobe (fronto-polar region).

All signals were recorded by a 32-channel amplifier on a digital tape recorder with a sampling rate of 3000 Hz. These signals were downloaded to a computer hard disk. On downloading, sampling rate were reduced to 750 Hz through antialiasing filter. Sleep stages were scored on a computer display using the Cz EEG, EOG and EMG, following standard criteria.⁹ Fifty 2048 point (= 2.73 s) artifact free epochs of each wake, NREM and REM states were selected and subjected to FFT analysis.

This protocol was approved by the Tokyo Institute of Psychiatry's ethics committee.

RESULTS

In recording I (Fig. 2), γ (all states) and β -1 (wake and REM) oscillations were observed in the MTL as we previously reported.⁶⁻⁸ In recording II (Fig. 3), ECoG signals from ACC showed very regular and continuous θ oscillations during wakefulness