cells appeared to be obviously malignant, occasionally in a form of multinuclear monstrosity.
Although the disease is not treatable, the correct diagnosis would be important for the patient and his relatives as well as for the physicians to avoid further expensive, complicated, and sometimes even invasive diagnostic and therapeutic procedures. The instructive photographs of the prominent features on MR imagines as well as on autopsy slices including GFAP staining will be presented.

P 2117

Severe hypophyseal disturbances with dwarfism due to craniopharyngioma

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Y. H. L., a 19-year-old male examined by us at the Department of Medical Neurology and the Epilepsy Center Yangon in February 2000 has shown severe dwarfish growth since his childhood. His appearance was that of an 8-year-old boy, at which age growth retardation was first observed. Body weight was 42 kg, head circumference 52 cm, height 134 cm. The patient showed highly disturbed secondary sex characteristics. There was a decline in intellectual performance. During the last two years the patient claimed increasing drowsiness, he developed diabetes insipidus. Three generalized seizures were observed since January 2000.
The neurological examination revealed polyneuropathy with severe muscle atrophy, marked flaccid paresis and typical sensitive disorders in combination with spastic symptoms and pseudobulbar paralysis. In addition optomotor disturbances (divergent position of the eye, upward gaze paresis) and marked frontal and cerebellar symptoms were seen. The patient showed hypophyseal dysfunction with alabaster coloured skin, hypogenitalism, diabetes insipidus and dwarfism. cCT showed a giant craniopharyngioma and hydrocephalus occlusus with signs of a transventricular CSF absorption. The EEG was pathological with delta rhythm over the temporo-occipital region with left sided accentuation.
The patient developed a coma state with symptoms of an acute midbrain syndrome followed by bulbar brain syndrome. He died from cardio-respiratory arrest. A postmortem examination was not accepted by the Buddhist family.
A giant craniopharyngioma with massive diencephalic-hypophyseal disturbances due to direct pressure on the surrounding area (upper brain stem, cerebellum) and an occlusion of CSF circulation with massive secondary intraventricular pressure seems to be a very rare condition. Early diagnostic measurements would have discovered the benign tumour much earlier. The patient died in a secondary tentorial and foraminal herniation after decompression of the severe hydrocephalus occlusus.

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Subtotal removal of acoustic neurinomas: causes and results

A. F. Smeyanovich, Y. G. Shanko
Minsk, Belarus

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Paraneoplastic dermatomyositis due to squamous cell carcinoma of sphenoid sinus

B. Zamini, K. Parsa
Tehran, Iran

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Cytology of cerebrospinal fluid leptomeningeal carcinomatosis

S. Miletic Drakulic, G. Toncev, J. Jevdjic
Kragujevac, Yugoslavia

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A rare tumour of the cranium basis (a case report)

Z. Jovanovic, D. Kozic, A. M. Pavlovic, J. Zidverc-Trajkovic, N. Sternic
Belgrade, Yugoslavia

P2122

Hospital-based study of neurooncological diseases in Georgia from 1987 to 1997

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Neurorehabilitation

P 2123

Changes of gross motor skills of patients treated with intensive neurophysiological rehabilitation system

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Introduction The Intensive Neurophysiological Rehabilitation System was worked out and successfully applied for the rehabilitation of patients with Cerebral Palsy. During 10 years more than 10 thousands patients including over six thousands from Germany, Austria, Switzerland and other Western European countries have been treated with good long-lasting results. The aim of the study was to measure the changes of gross motor skills of patients with Cerebral Palsy during the treatment.
Materials and Methods Gross motor skills of 2165 patients with Cerebral Palsy were assessed before and after the two-week course of intensive neurophysiological rehabilitation using the 56-items test of gross motor skills. The treatment includes biomechanical correction of the spine, extremity joints mobilization, reflexotherapy, mobilizing physical therapy, special massage system, rhythmical group exercises, mechanotherapy and apitherapy.
Results Improved head control in prone position was noted in 79% of patients unable to control the head before the treatment. Sitting abilities developed 65% of the patients, who could not sit before and 27% of the patients learned to crawl. Standing skills gained 40% of the patients, not able to stand before the treatment, and 18% of the patients, who could not walk, acquired this ability.
Conclusion The study suggests that the Intensive Neurophysiological Rehabilitation System have a positive influence on the development of gross motor skills of patients with Cerebral Palsy.

SEVERE HYPOPHYSIAL DISTURBANCES WITH DWARFISM DUE TO CRANIOPHARYNGIOMA

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1. Introduction

A 19 year old male (Y.H.L.) examined at the Department of Medical Neurology and the Epilepsy Centre, University Yangon in February 2000 showed severe dwarfish growth (Fig. 1), which had existed since his childhood, noted in the 6th year of life in form of retarded growth. The boy is the oldest child of a deeply religious Buddhist family living in the country near Yangon. During the last two years his main problems, progressive dementia and diabetes insipidus, became apparent. Since January 2000 three grand mal seizures had occurred. The cCT detected a giant craniopharyngioma.

2. Neurological findings

Optomotor disturbances (divergent position of the eyes (Fig 2) upward gaze and convergent paresis, cogwheel eye movements with horizontal nystagmic component, enlarged pupils with diminished reaction to light and to convergence).

- Spastic symptoms (hyperreflexia, pyramidal signs, spasticity, pseudobulbar paralysis)
- Polyneuropathy (dysesthesia on the lower part of the extremities, muscular atrophy, diminished tendon reflexes, positive Lasègue)

Frontal lobe signs (palmo-, pollico- and mentomental reflex, grasp and oral reflexes)

Cerebellar symptoms (atactic gait, hemispheric ataxia)

Parkinson symptoms (amimia [Fig 2] akinesia, autonomic signs, rigidity)

Dementia

Signs and symptoms of an initial state of acute midbrain syndrome (tendency to flexed- stretched position of the extremities [Fig 2], tendency to somnolence, reduced vigilance)

3. Endocrinological signs

Dwarfish feature (19 year old male looking like an 8 year old boy [Fig 1], height 134 cm, weight 42 kg, head circumference 52 cm

Decline in intellectual performance

Absence of secondary sex characteristics (Fig 1)

Diabetes insipidus

Various hypophysial signs (Fig. 1, 2)

Details of the hormonal status were not available

4. cCT and EEG

CCT: giant craniopharyngioma, hydrocephalus occlusus with transventricular CSF absorption (Fig. 3)

Abnormal EEG, delta rhythm, focused in the temporo-occipital region with left sided accentuation (Fig.4).

5. Clinical course

Severe dwarfish growth together with other hypophysial symptoms had been observed since the sixth year of life of a 19 year old Burmese man. Only in February 2000 during his first neurological examination a giant craniopharyngioma was discovered which had had a severe space occupying effect for more than two years due to direct pressure of the tumour on the surrounding area. Parkinson signs, cerebellar and spastic symptoms, optomotor symptoms and diabetes insipidus developed. A progressive stenosis of the aqueduct caused disturbances of CSF circulation with progressive dementia and epileptic seizures The occlusion of the aqueduct lead to rapid increase of intraventricular pressure with tentorial hemiation and an acute midbrain syndrome. The patient died with respiratory and cardiac arrest in an acute bulbar brain syndrome.

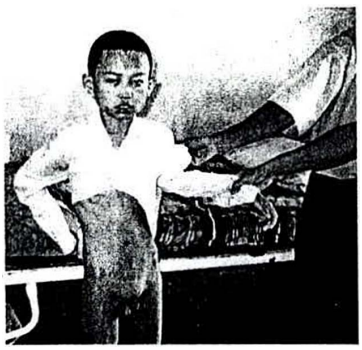

Fig.1: 19 year old male with dwarfish growth based on craniopharyngioma

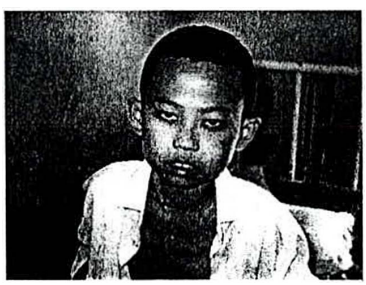

Fig.2: Optomotor disturbances (divergent position of the eyes)

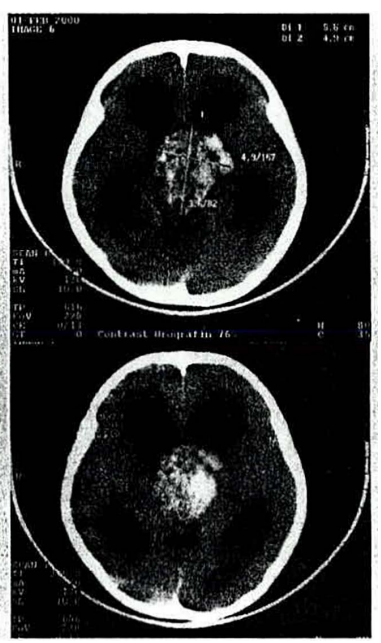

Fig.3: Hydrocephalus occlusus based on a giant craniopharyngioma

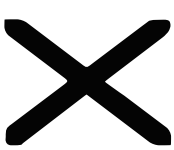

Fig.4: EEG with delta rhythm, focused in the temporo-occipital region

6. Conclusion

10% of all supra- and parasellar tumours are craniopharyngiomas. They develop out of Rathke's pouch, a residue of the ductus craniopharyngeus. 2/3 of all craniopharyngiomas occur in children until the age of 16 (1). Based on a reduction of gonatotropin, patients do not develop secondary sex characteristics. Also, a reduction of TSH, ACTH, somatotropin and vasopressin has been observed in patients with craniopharyngioma (2). The five year survival rate after surgery reaches 100%(1). There are only few cases in the literature with malign course (3). Mostly combined with cystic involvement, usually discovered after a short time (4).

The dwarfishness of the 19 year old man was observed since his 6th year of life. The boy passed the elementary school programme with partly good results, later on he was not able to work in his father's small rice farm. Various endocrinological disturbances beside dwarfishness had developed. When the boy was 17 progressive dementia was observed together with additional neurological symptoms such as optomotor disturbances, Parkinson symptoms, spasticity and cerebellar symptoms. Because of epileptic seizures and progressive dementia, the patient was brought into the Neurological University Clinic, Yangon, where a giant craniopharyngioma with massive space occupying effect was discovered. Occlusion of the aqueduct lead in the final stage to tentorial hemiation, with an acute midbrain syndrome followed by an acute bulbar brain syndrome. The young man died after respiratory and cardiac arrest.

The history of this unique course leads to the conclusion that the giant craniopharyngioma must have existed for many years with dwarfishness having been the first symptom at the age of 6.

The clinical course passed three phases. The first phase was characterised by endocrinological disturbances. In the second phase, neurological symptoms due to direct lesion of the surrounding brain region (optomotor disturbances, Parkinson symptoms etc.)and progressive diffuse brain damage (dementia) based on a stenosis of the aqueduct were observed. In the third phase decompensation of the space occupying lesion lead to tentorial and foraminal hemiation with secondary acute midbrain and irreversible acute bulbar brain syndrome.

The special health situation in a developing country with Buddhist background and lack in the health care system may cause such a fatal course of a benign tumour.

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