Dwarfism and severe endocrinological disturbances in a patient with a giant craniopharyngioma

Ch. Stepan*)**), Nyan Tun***), MiMi Cho***), H. Lukasser****), F. Gerstenbrand**)

*) Neurological Centre/ Otto Wagner Hospital, Vienna, Austria**) Ludwig Boltzmann Institute for Restorative Neurology and Neuromodulation, Vienna, Austria

***) Department of Medical Neurology, Yangon, Myanmar

****) EEG Department, Neurological University Clinic, Innsbruck, Austria

Abstract:

Craniopharyngeoma is a benign epithelial tumor of the central nervous system. Four clinical stages in the development of the tumor can be differentiated: direct pressure to the surrounding brain, endocrine dysfunction, compression at the hypothalamic hypophyseal axis and increased intracranial pressure due to the compression of the aqueduct. Mostly the diagnosis of a craniopharingeoma is done, during the first two phases. In the history of the demonstrated patient all four phases were observed. The main symptom was dwarfism. Out of the special situation of the patient this benign tumor caused a fatal course.

Key words:

Craniopharyngeoma - endocrinological dysfunction with dwarfism - increased intracranial pressure – acute brainstem symptoms

Craniopharyngeoma is a benign epithelial tumour of the central nervous system. Only a few cases with malign course are reported (1). Mostly craniopharyngeoma is a tumor of the childhood. The peak of incidence is

between the 6-29 years (3,4,5,6,7). A second peak can be seen in the age range of 40 - 50 years (4,5). The tumor tissue develops out of the adamantinomatous type and the papillary variant. Frequently a mixture of both types is possible (1).

Rathke`s pouch, a residue of the Ductus craniopharyngeus (2, 3). Histologicaly two types of craniopharyngeoma can be differentiated, the adamantinomatous type and the papillary variant. Frequently a mixture of both types is possible (1).

In children, craniopharyngeoma is the third most common brain tumor (4). In the literature the frequence is quoted between 5% and 9% (8,7) of all intracerebral tumors in childhood. Of all supra- and parasellar tumours 10% are craniopharyngeomas (6, 8). In its defelopment the Craniopharyngeoma can invade neighbouring brain tissue. In the first stage the hypituitary gland, hypothalamus and the hypituitary stalk (1,6,8). In further course the optic pathway and the upper brain stem.

Patients with craniopharyngeoma generally present four major clinical syndromes related to visual problems resulting due to direct compression of the optic pathways, endocrinological dysfunction caused by compression of the hypothalamic - hypophyseal region and increased intracranial pressure (1). First clinical signs of an increased intracranial pressure are headache, nausea, visuell disorders (1,2,4,5,9).

In the following course a hypophysar and hypothalamic insufficiency is developing. A reduction of growth hormone (GH), luteotrop hormone (LH), follicle stimulating hormone (FSH), thyreotrop hormone (TSH), adrenocorticotropes hormone (ACTH) somatotropin and vasopressin are to observe (6, 10,11). The first clinical sign of the endocrinological dysfunction is a disorder of growth (5,6,12,13,14,15).

Clinical history:

The 19 year old man was admitted at the Department the Neurological clinic and in the Epilepsy Center, University Yangon in February 2000. He lived in a mural area. As main symptoms a dwarfish growth was observed. In the case history the growth disturbance could be proved since his 6th year of life. The paitend was admitted because of generalized seizure. The adult patient looked like an eight year old boy. His height was 134 cm, the weight 42 kg, circumference of the head 52 cm. During the last two years symptoms of a progressive dementia were observed. Since one year a diabetes insipidus was known. From January 2000 three grand mal seizures occured.

The neurological examination showed optomotoric disturbances, a divergent position of eyes, paresis of upward gaze and of convergence, coghwheel movements and a horizontal nystagmus, enlarged pupils with diminished reaction to light and to convergence. On all extremities increased tendon reflexes with absence of ancle jerk, hypaestesia, severe muscle atrophy, clear frontal lobe signs (palmo-, pollico-, mentomental reflexe, grasp and oral reflexes), cerebellar symptoms (atactic gait and posture hemispheric ataxia), spastic signs, Parkinson symptoms (amimia, akinesia, vegetative signs and rigidity) and marked dementia was to recognize.

Endogrinological symptoms were marked with dwarfish feature, hypogenitalism absence of secondary sex characteristics, diabetes insipidus (8 liter per day). A detailed hormon status could not be examinated.

The EEG was abnormal with delta rhythm, focused in the temporo - occipital region and left sided accentuation. In the cCT a giant craniopharyngeoma was found with hydrocephalus occlusus and signs of transventricular CSF absorption.

Ten days after the admission to the hospital the patient died inner two hours.

Discussion:

Craniopharyngeoma large size before becoming clinically symptomatic, but it remains the possibility of local pressure on blood vessels and nerves. A craniopharyngeoma can pass four phases of symptoms. In the first phase visual disturbances can be observed due to direct compression of the optic nerve, the second phase is characterized by endocrinological dysfunction caused by compression of the hypothalamic - hypophysial region. In the third phase intracranial pressure is increasing. In the fourth phase symptoms of brain stem lesion in form of a midbrain and a bulbarbrain syndrome can occure. In childhood the first clinical signs are headaches, vomiting and vision disorders. The hypophysaer and hypothalamic insufficiency is producing endocrionological signs which can be objektivated by examing the level of growth hormone (GH), luteotrop hormone (LH), follicle stimulating hormone (FSH), thyreotrop hormone (TSH) adrenocorticotrop hormone (ACTH), somatotropin and vasopressin and its clinical pictures like dwarfism, diabetes insipidus and other insufficiancy.

In the following course the space occupying effect of the tumour develops with an occlusion of the aqueduct and an increased intracranial pressure with progressive dementia and finally a tentorial herniation.

A craniopharyngeoma in adult developes different clinical signs, with high risk of cardiovascular problems. Hormonal dysfunction leading to impotence in males and amenorrhea in females. The relation from muscle tissue to fatty tissue is shifting from muscle in fabour of fat. (6, 11).

To verify the diagnosis the insufficience of the hypophysär - hypothalamic hormones is an important criteria. On the other hand the possibility of radiodiagnostic tests. In radiodiagnostic x-ray show the calcification of the tumour tissue (2,4). In international literature the insulin - hypoglycaemic test and the aginin - load test are recommended to support the diagnosis (6). As differential diagnosis a glioma of the nervous opticus or the optic chiasm, a Corpus pinealis tumour and a tumour of the wall of the third ventricle have to be taken into consideration (2).

The therapy of craniopharyngeoma is a combination of surgical and radiotherapeutic treatment (8). The strategy of surgical intervention depends on the localisation of the tumour and the problem of a residua of the tumour. For that reason a radiotherapy is necessary after an incomplete resection of the tumour. After gross total resection the recidive rate is approximately 20%. Tumours incompletely respected have recidive rate up to 60%. After radiation incomplete respected craniopharyngeoma show 30% of remission (1). A postoperative growth spunt was observed by the use of growth hormone (GH) (5,8,12). In some cases a normal growth without aplication of GH was reported after craniotomy (5,13). Successful treated patients with craniopharyngeoma and similar clinical courses are reported by *Frasier et. al.* (13)

Summary:

In the demonstrated case a rare course of a patient with a giant craniopharyngeoma is presented. From his 6th year of life, of the 19 year old patient dwarfish growth was observed. Optomotoric disturbances in form of a divergent position of the eyes were known for several years, a diabetes insipidus since two years. When the patient was 17, progressive dementia could be observed together with additional neurological symptoms such as Parkinson symptoms, spastic signs and cerebellar symptoms. Occlusion of the aqueduct lead in a final stage to tentorial herniation, with an acute midbrain syndrome followed by an acute bulbar brain syndrome. The young man died after respiratory and cardiac arrest. In the first neurological examination, after three grand mal epileptic seizures at the age of 19, a giant craniopharyngeoma with massive space occupying effect was discovered. The giant craniopharyngeoma must have existed for many years.

Four phases in the clinical course are to analyse. 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 and cerebellar symptoms). The thired phase was characteristed by a progressive diffuse brain damage with dementia based on a stenosis of the aqueduct. In the fourth phase with volume increase and tentorial and foraminal herniation with secondary acute midbrain and irreversible acute bulbar brain syndrome.

The cours of a giant craniopharyngeoma discovered in a 19th year old man has to be seen against the Buddhistic background and the special health situation in a developing country

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Organizing Secretary,
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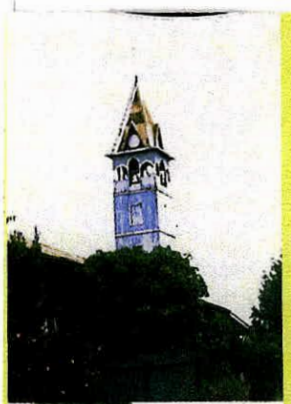

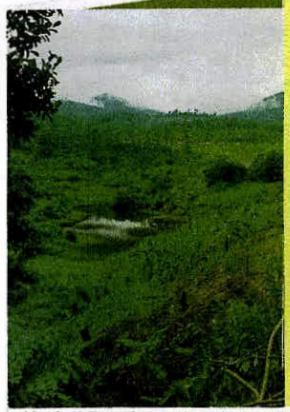

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