ANNALES
UNIVERSITATIS MARIAE CURIE-SKŁODOWSKA
LUBLIN — POLONIA
VOL. LI, 22 SECTIO D 1996

Katedra i Klinika Neurologii Akademii Medycznej w Lublinie
Kierownik: prof. dr hab. Wiesław Kawiak

ANDRZEJ FIDOR, HALINA KRASIŃSKA-CZERLUNCZAKIEWICZ, WALDEMAR FRĄK, EWA WRZOS

Clinical Analysis of Massive Calcifications within Basal Ganglia and Cerebellum

Kliniczna analiza masywnych zwapnień w obrębie jąder podstawy mózgu i móżdżku

Since the introduction of brain CT intracranial calcifications have been quite frequently detected. This method is over ten times more sensitive than traditional X-rays of the skull.

Intracranial calcifications are found in numerous pathologic conditions of intracranial structures, such as aneurysms, angiomas, neoplastic tumours, intracranial haematomas, tuberculomas, in the course of facomatoses, syphilis of the brain and meninges, and also parasitic diseases of the brain, such as cysticercosis, toxoplasmosis (5, 8, 10, 11).

Cases of massive, quickly forming calcifications after acute cerebral ischaemia have been described in scientific papers (6).

There is often found the presence of intracranial calcifications in cases with symptoms of hypoparathyroidism (9).

There are also known cases of familial calcifications within the brain, and most of them are localised in the basal ganglia and cerebellum (2, 3).

Lowenthal and Bruyn suggested a division of diseases with familial calcifications of basal ganglia and dental nucleus of the cerebellum into calcifications resulting from disturbances of calcium-phosphate metabolism associated with the function of parathyroids and spontaneous calcifications independent of parathyroidal function (2, 5).

In patients with intracranial calcifications there are often observed

mental disability, pyramidal and extrapyramidal symptoms, as well as epileptic seizures, but there were also found patients with intracranial calcifications without neurologic symptoms (4, 10, 12).

The authors observed in Neurology Clinic of the Medical School in Lublin patients with dysfunctions of the pyramidal and extrapyramidal system and also with the symptoms of psychoorganic syndrome, in whom CT-imaging revealed symmetric calcifications within basal ganglia and cerebellum.

CLINICAL OBSERVATION

A patient A. I., aged 54, was admitted to the Clinic due to quickly increasing symptoms of psychoorganic syndrome. They were accompanied by deterioration of motor function. These symptoms were preceded by two episodes of losing consciousness with head injury. It was, however, impossible to determine the sequence of these events. Several years earlier the patient had sustained head trauma and abdominal injury in a road accident. He had not been treated for any diseases of the thyroid and the parathyroid glands. Neurologic examination showed features of extrapyramidal symptoms such as fine tremor of the limbs and muscular rigidity. These were accompanied by pyramidal symptoms such as hyperreflexes and irregular occurrence of the Babiński sign. Neuropsychologic examination revealed general decrease in intellectual capacity mainly in the sphere of thinking, memory and criticism. EEG examination showed the presence of medium degree generalised changes such as slowing down of basic brain electric function. No asymmetry, focal changes or discharges were found on EEG examination. CT-imaging of the brain showed symmetric calcifications in both hemispheres of cerebellum and vermis, and homogenous calcifications in both caudal nuclei, in central parts at both lateral ventricles, and also bilaterally centrum semiovale. The ventricular system of the brain was symmetric. Slight cortical-subcortical atrophies were observed. Angiographic picture of the brain showed atheromatic changes, especially in extracranial vessels, but the lumen was retained. No other vascular pathology was found. Apart from a small increase of protein concentration (52 mg%), no other abnormalities were observed in cerebro-spinal fluid. Basic blood and urine tests were normal. Parameters of calcium-phosphate metabolism, that is calcium concentration (4.9 mEq/l) and phosphorus concentration (4.1 mEq/l) were within normal limits. This was also the case with the activity of thyroid hormones. Vasoactive drugs and medicines affecting the extrapyramidal system were used achieving improvement in the physical and mental activity of the patient. The above presented findings let recognise extensive symmetric periventricular intracranial calcifications with the coexistence of psychoorganic and pyramidal-extrapyramidal syndrome.

A patient H. B., aged 50, was hospitalised in Neurologic Clinic because of headaches, lowered mood, difficulties in concentration and memory disturbances. The case history did not reveal the presence of earlier diseases of the thyroid gland and parathyroids. Neurologic examination showed a generalised increase of the muscular tension (muscular rigidity). No pyramidal signs were observed. Neuropsychological evaluation revealed the presence of psychoorganic syndrome. On EEG examination the patient was normal. CT-imaging of the head revealed the presence of periventricular calcifications, calcifications of the basal ganglia and both hemispheres of the cerebellum. Blood and urine tests did not show any pathologic changes. Blood calcium concentration was 5.2 mEq/l (normal value 4.5 — 5.5 mEq/l), and phosphorus concentration was 3.9 mEq/l (normal value 2.4 — 4.4 mEq/l). The activity of thyroid hormones was normal. It was not possible to perform the Ellworth-Howard test consisting in the assessment of cAMP excretion in urine after parathormone administration. Basing on the above presented abnormalities there were diagnosed massive intracranial calcifications of unclear aetiology with the suspicion of the Fahr's syndrome with late beginning and dominating neurologic signs and progressive dementia.

DISCUSSION

In the patients calcium was deposited symmetrically within basal ganglia and in the hemisphere of the cerebellum and vermis. Analysing clinical picture and examination results the above described complex of signs seems to be most corresponding to the Fahr's syndrome. This interpretation is also supported by calcium and phosphate content in the blood. However, the authors did not have any possibility of a more detailed examination od parathyroideal function, and especially determining of parathormone activity. Therefore, a definitive and conclusive diagnosis could not be made.

Calcium may get deposited in the basal ganglia in extravascular spaces and consist of conglomerates of calcium, iron, zinc and magnesium salts. The ground for these changes can be precipitation of blood serum proteins in perivascular spaces. The forming of protein-multipolysaccharide complexes can make up the matrix for depositing of inorganic substances (4, 5).

In patiens with the Fahr's syndrome calcium salts deposits can be affected by local factors. The basal ganglia are the site of special biochemical activity of the nervous tissue. It is also the place where the

greatest amounts of metal compounds and other microelements occur, which function as catalysers. Changes of physico-chemical properties of tissues in the presence of various pathogenic factors can be responsible for metabolic disturbances, and consequently for calcium deposits (1, 6, 7).

In the Fahr's diseases calcium is deposited in the walls of blood vessels unchanged by atherosclerosis (5, 11).

The depositing of calcium-phosphate in soft tissues depends on the ratio of calcium content to phosphates in extracellular fluids. Only the exceeding of certain values of this ratio predisposes to the formation of calcium deposits (5, 8).

There are also spontaneous calcifications in soft tissues, in which it is impossible to show disturbances of calcium to the phosphates ratio. In the mechanism of developing symptoms of CNS dysfunctions in patients with intracranial calcifications one should consider the presence of vascular factors. A decrease in blood flow through the basal ganglia may affect development of neurologic signs. Head injury with marginal cerebral oedema may compensate for the functional efficiency of the central nervous system and predispose to the formation of calcium salts deposits (1, 12).

REFERENCES

1. Bartecki B. F., Kamieniowski J.: Zwalniające niedokrwienie ogniskowe w przypadkach choroby Fahra. Neur. Neurochir. Pol., 4, 443, 1979.
2. Flint J., Goldstein L. H.: Familial calcification of the basal ganglia: a case report and review of the literature. Psychol-Med., 22/3, 581, 1992.
3. Forstl H., Krumm B., Eden S.: Neurological disorders in 166 patients with basal ganglia calcification: a statistic evaluation. J. Neurol., 39, 36, 1992.
4. Koeller W. C.: Calcification of the basal ganglia. Computerized tomography and clinical correlation. Neurology, 29, 3, 1979.
5. Lowenthal A., Bruyn G. W.: Calcification of the striopallidodentate systemj. [In:] Handbook of Clinical Neurology, Amsterdam 1968.
6. Midroni G., Willinsky R.: Rapid postanoxic calcification of the basal ganglia. Neurology, 42, 2144, 1992.
7. Morgante L., Vita G.: Fahr's syndrome: local inflammatory factor in the pathogenesis of calcification. J. Neurol., 233, 19, 1986.
8. Mossakowski M. J.: Podstawy neuropatologii, PZWL, 379, Warszawa 1981.

9. Paradowski B., Nikiel M.: Rozległe zwapnienia śródmózgowe w przebiegu pooperacyjnej niedoczynności przytarczyc. Neur. Neurochir. Pol., 1/2, 94, 1990.
10. Puramendram K.: Basal ganglia calcification on computer tomografic scan. Acta Neurol. Scand., 3, 66, 1982.
11. Taxer F.: Klinische Fruhsymptome und CT-Befunde beim Fahrschen Syndrom. Nervenarzt, 10, 583, 1986.
12. Ziąber J.: Zwapnienia jąder podstawy i móżdżku. Neur. Neurochir. Pol., 5, 721, 1993.

Otrz. 1996.09.29

STRESZCZENIE

Autorzy przedstawili 2 chorych z obecnością zwapnień wewnątrzczaszkowych o lokalizacji w obrębie jąder podstawy mózgu oraz w móżdżku. Chorzy cierpieli z powodu objawów zespołu piramidowo-pozapiramidowego oraz postępującego otępienia. Nie wykazano obecności ogólnoustrojowych zaburzeń gospodarki wapniowo-fosforanowej.