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Non-tumoral non-infectious intracranial calcifications

Intracranial calcifications are often an accidental finding on conventional radiograms or computed tomography (CT) scans. They can be physiologic or pathologic, accompanying various diseases of the central nervous system. Pathologic calcifications can be found in tumors as well as in other non-neoplastic diseases. In differential diagnosis of intracranial calcifications there should be mentioned two groups: infectious and non-infectious calcifications.

The aim of the paper was presentation of non-tumoral non-infectious intracranial calcifications on the basis of literature review and own observations.

Among non-infectious diseases producing intracranial calcifications there are: phacomatoses (Sturge-Weber syndrome, neurofibromatosis, tuberous sclerose, von Hippel-Lindau's disease), endocrine disorders (hypoparathyroidism, pseudohypoparathyroidism), idiopathic non-arteriosclerotic calcifications (Fahr's disease), nephrogenic diabetes insipidus, intoxications (lead, CO), postasphyxial enecephalopathy, bone abnormalities (Albers-Schoenberg disease, carbonic anhydrase II deficiency), vascular calcifications (arteriosclerosis, arterial aneurysms, cerebral vascular malformations), trauma, celiac disease, linear scleroderma as well as other rare syndromes (Sneddon's syndrome or Divry-Van Bogaert's orthochromatic leucodystrophy) (1-13).

PHACOMATOSES

Neurofibromatosis (von Recklinghausen's disease) is characterized by the presence of numerous neurinomas of cranial and spinal nerves. There is observed calcification of the choroid plexus to an unusual degree, often bilateral and symmetric, but also asymmetric and more pronounced in one of the lateral ventricles. Another find-

ing can be cerebellar calcification in the form of multiple nodular calcified lesions, some of which confluent, located peripherally in the folia of the cerebellar hemispheres. There can also be detected fine punctate calcified lesions of less than 0.5 cm in diameter on the cortical surface of cerebral hemispheres (7).

Sturge-Weber syndrome is a neurocutaneous syndrome characterized by leptomeningeal angiomatosis and facial port wine hemangioma (nevus flammeus) mostly located in the area of the first and second branch of the trigeminal nerve (6, 12, 13). Leptomeningeal angiomatosis is associated with cerebral angiomatosis in 10% of all children with facial port-wine stains, but leptomenigeal angiomatosis seldom occurs without facial hemangioma (13). Ipsilateral intracranial calcifications occur in tramlike patterns evident on skull radiograms with the advancing age (12, 13). CT is more sensitive in detection of calcifications before they become visible on conventional X-rays (13). Intracerebral calcifications are the result of thrombotic obliteration and ischemia (13). Focal or extensive cerebral cortical calcifications, often make the CT evaluation of changes in the cerebral gray and white matter impossible (6).

Tuberous sclerose (Bourneville's disease) is an autosomal dominant genetic disorder. It manifests itself in the central nervous system with isolated masses of gray matter made of abnormally developed nerve cells and neuroglia, especially around ventricles, rarely in cerebellum. Some of the nodules along the margins of ventricles undergo degenerative changes and can calcify. Often the lesions project into the ventricular cavities, thus in 50% of patients calcifications are evident on conventional skull radiograms, but rarely until 1-2 years of age. The radiopaque lesions are numerous (in 75%), bilateral (in 50%), small and rounded, however, due to coalescence of nodular calcifications, irregular forms may develop. Deep location corresponds to localization along the ventricular walls (3).

Von Hippel-Lindau's disease (neuroocular angiomatosis) is an autosomal dominant genetic disease with the presence of retinal and intracranial hemangiomas that can calcify (3).

ENDOCRINE DISORDERS

Hypoparathyroidism or Albright osteodystrophy is an endocrine syndrome in which calcifications involve mainly the basal ganglia, but also the falx cerebri. In this case intracranial calcifications are the result of disorders of calcium metabolism and biochemical test should be prescribed (9, 12).

Pseudohypoparathyroidism causes appearance of such skeletal abnormalities as brachydactyly, short and wide phalanges, bowing, exostoses and thickening of calvaria. In many cases calcification of the basal ganglia can be observed (8, 9).

METABOLIC DEFECTS

Ferrocalcinosis (Fahr's disease, Idiopathic nonarteriosclerotic calcifications) is a familial disease affecting small vessels with the depositions of iron and calcium involving mainly the basal ganglia, but also the subependymal areas surrounding the ventricles (12).

Cockayne's syndrome is a metabolic defect inherited as a recessive autosomal disease. Microcephaly and perivascular mineralization in the basal ganglia and in the dentate nucleus of the cerebellum are one of characteristic features (12). The shadows of calcifications are punctate and granular, resembling those in hypoparathyroidism.

Familial mitochondrial encephalopathy with lactic acidosis may be due to various defects of energy metabolism: pyruvate dehydrogenase complex, biotinidase, isolated or multiple deficiencies of the respiratory chain enzyme complexes or combinations of these defects (11). There were described periventricular calcifications capping the anterior horns of ventricules as well as gross calcification of necrotic areas in neonatal brain (11).

NEPHROGENIC DIABETES INSIPIDUS

Intracranial calcifications are an unusual complication of nephrogenic diabetes insipidus. They occur in the form of symmetrically distributed calcification of basal ganglia as well as calcifications in the frontal, temporal, parietal and occipital lobes near the gray and white matter junction (8).

INTOXICATIONS

CO intoxication – in the course of the intoxication basal ganglia calcifications appear as a result of the toxic effects of carbon monoxide inhalation (3).

Lead intoxication – intracranial calcifications are due to chronic lead exposure (3).

TRAUMA

Chronic subdural hematomas (2) of at least several years of duration may undergo calcification or even ossification (Fig. 1). Usually the calcium deposits are curvilinear and parallel to the contour of the cranial vault. Between the calcium lines there can be visible irregular plaques of calcium. Fortunately, due to the attention paid nowa-

Fig. 1. Gross shadow of calcification in cranial vault on the right is a sequel to a conservatively treated chronic subdural hematoma

days to patients with cranial trauma, such long standing, calcified subdural hematomas are rarely observed.

BONE ABNORMALITIES

Albers-Schonberg disease is a syndrome of osteopetrosis and intracranial calcifications associated with renal tubular acidosis due to carbonic anhydrase II deficiency. Intracranial calcifications are usually bilateral and located in the basal ganglia and the white matter, especially in the white-gray junctional area. The tentorium, the falx cerebri, the cerebellum, the thalamus and the corpora colliculi can also be affected. There was described the image of ventricules lined by globules of calcifications extending into the white matter (9, 10).

Metaphyseal dysplasia can be accompanied by calcifications of the corpus callosum as well as of periventricular and subcortical white matter with no reported CNS symptoms. Thus differential diagnosis of intracranial calcifications should include bone X-ray examination (9).

VASCULAR CALCIFICATIONS

Arteriosclerosis causes visible vascular calcifications in many aging patients. Calcification is a prominent feature of the disease of cavernous and siphon parts of the internal carotid artery (Fig. 2). Lateral skull X-rays reveal calcified vessels as transverse

Fig. 2. Calcifications of walls of the distal part of left internal carotid artery

lines projecting upon the sella turcica or sphenoid sinus. Extensive calcifications may extend upwards above the anterior clinoid processes and in severe cases even branch into the anterior and middle cerebral arterial patterns (3).

Arterial aneurysms are often affected by arteriosclerosis causing the appearance of curvilinear calcifications. Such calcifications are thinner and smoother than in tumors. Calcified aneurysms are most often found along the internal carotid artery and the circle of Willis (3).

Cerebral vascular malformations such as previously described Sturge–Weber syndrome.

CELIAC DISEASE

Calcifications in celiac disease have been described in sporadic cases since 1976. The association between cerebral calcifications and celiac disease is peculiar and pathogenetic relationship between the two pathologies is unknown. The calcifications are extensive, usually cortico-subcortical, at the junction of the white and gray matter. They may be present both in the brain tissue and in the walls of cerebral vessels. They appear as free sharply demarcated psammoma-like bodies and small granular calcium depositions along venules and capillaries or as heavy calcifications in cortical veins causing severe stenosis (1, 4). There were also described bilateral occipital calcifications resembling tram tracks with no nevus flammeus and normal angiogram.

LOCALISED SCLERODERMA

In linear form of localised scleroderma when skin lesions involve the face and the scalp causing the *en coup de sabre* deformity, ipsilateral intracranial calcifications of the frontal lobe can be found. The etiology of intracranial calcifications is unknown, although some researchers have postulated that they may be due to calcified hemangiomas. No clinical or neurological signs and symptoms accompany the lesions (5).

CONCLUSIONS

It can be concluded that although more sporadic than tumoral or infectious calcifications, calcified lesions in non-infectious diseases should also be taken into account in differential diagnosis of intracranial calcifications.

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SUMMARY

Intracranial calcifications are often an accidental finding on conventional radiograms or computed tomography (CT) scans. They can be physiologic or pathologic, accompanying various diseases of the central nervous system. Pathologic calcifications can be found in tumors as well as in other non-neoplastic diseases. In differential diagnosis of intracranial calcifications there should be mentioned two groups: infectious and non-infectious calcifications. In the paper there were discussed non-tumoral non-infectious intracranial calcifications. Although more sporadic than tumoral or infectious calcifications, calcified lesions in non-infectious diseases should also be taken into account in differential diagnosis of intracranial calcifications.

Nieguzowe zwapnienia śródczaszkowe pochodzenia niezakaźnego

Zwapnienia śródczaszkowe są częstym przypadkowym znaleziskiem na konwencjonalnych zdjęciach rentgenowskich i w badaniach metodą tomografii komputerowej. Mogą być fizjologiczne lub patologiczne – towarzyszące różnym nowotworom i chorobom nienowotworowym. W pracy omówiono nieguzowe zwapnienia śródczaszkowe występujące w przebiegu niezakaźnych schorzeń ośrodkowego układu nerwowego. Chociaż są one znacznie rzadsze niż zwapnienia guzowe lub w chorobach zakaźnych, to należy je również brać pod uwagę w diagnostyce różnicowej zwapnień śródczaszkowych.