CLINICO-LABORATORY CHARACTERISTICS AND PATHOGENETIC FEATURES OF THE MORGAGNI-STEWART-MOREL SYNDROME

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The syndrome was described by the pathoanatomist Morgagni for the first time in 1765, and was thereafter supplemented by Stewart in 1928 and by Morel (cited by 4) in 1930. The clinical picture is characterized by the following signs: intracranial hyperostosis, most frequently frontal, headache, systemic obesity, moderate hirsutism, very seldom virilism, systolic-diastolic hypertension, reduced carbohydrate tolerance to strongly manifested diabetes mellitus and nerve-mental illness (1—10). Many authors report additional symptoms, e. g. increased susceptibility to infectious diseases (8), higher glaucoma development incidence (2), menstrual disorders (2, 3, 11), vertigo (11, 13), goiter (5), hypo- or hyperthyroidism.

The Morgagni-Stewart-Morel syndrome (MSM) is rarely diagnosed in the clinical practice. However, whenever one is familiar with the condition and it is specifically looked for, its incidence proves much higher (4, 10). Twelve patients with MSM syndrome were observed over the past two years.

Clinical Material

Our series comprises female patients aged 34 to 64 years. The onset of the disease in eight of them was in the period 21—38 years, after 40 years — in three, and above 50 years — in one patient only. Intracranial hyperostosis is present in all cases, in nine it is a matter of hyperostosis frontalis interna, and in three — hyperostosis frontoparietalis. In one case alone the hyperostosis is asymmetric, with mainly leftside bony substance ammassment. Sella turcica is normal in eleven patients, and only in one its sizes are at the uppermost limit of the norm. Headache is a constant symptom, displaying chiefly frontal and occipital localization, and hardly responding to analgetics treatment. All patients have generalized obesity with formation of a pseudoclimacteric gibbus and pendulous abdomen (the form of apron), I degree in 1 patient, II degree — 6, III degree — 4 and IV degree — one patient. Single striae cutis distensae with a livid hue in the abdominal region are seen in three of the patients only. Discrete hirsutism is present in five patients, and rather marked — in seven. Three cases have manifested diabetes mellitus. The glucose tolerance test disclosed a diabetic blood-sugar curve in eight patients. Arterial hy-

pertension with values ranging from 140/100 to 180/120 is recorded in eight cases.

Nerve and mental disorders are established in all the patients of the series under review. Most frequently, they are characterized by vertigo, quick mental fatigability, depressive-hypochondriac experiences and anisoreflexia. A more thorough study of the vegetative nervous system reveals a number of functional variations. The Aschner Danini's test, performed in five cases, produced a positive reaction (pulse rate slowing with more than 12 beats/min and blood pressure fall with more than 10 mm Hg) in two of them, and negative paradoxical result in the remainder (3 patients). The epigastric reflex of Thoma and Roux proved to be strongly positive in three patients, positive — in two, and negative — in one. The clinostatic test elicited a positive reaction in two, and a paradoxical reaction in three cases, whilst the orthostatic test was positive in all patients under investigation. Skin thermometry showed asymmetry in five instances. Oscillometry of the forearm and legs was found to be normal in one female patient, whereas in the remainder it showed slight lowering of the oscillatory index with a varying degree of asymmetry, however, within the limits of physiological variations. The capillaroscopic picture disclosed variations in all seven patients subjected to examination. It is a matter of reduced in number, at some points kinked capillaries, with single or multiple arborizations, «fork»- or «cork-screw»-like forms against a pale, limpid background. EEG was made in four patients, and revealed varying degrees of disorganization of the normal alpha-rhythm and presence of theta-waves, as well as a marked diffusion of the changes, without delineation of foci and asymmetry.

Serum phosphorus is at the uppermost normal limit in two of the patients, and increased in one. The basal secretion of 17-KS is within normal limits, with intermittent rises being observed in two cases. Elevated 17-KS level is observed in three patients. Following ACTH stimulation they show more than a twofold increase in two cases, while in two the response is very weak. Hypophyseal ACTH storage, estimated with the aid of the metopirone test, is strongly reduced in ten of the eleven patients examined.

In part of the patients of our series, also some of the rare signs described in patients with the MSM syndrome are present, namely: enhanced liability to infectious diseases (2 cases), euthyroid goiter (similarly 2) and glaucoma (1 case). Exophthalmos is observed in one patient without clinical evidence of hyperthyroidism, at normal BSI values.

It is of special interest to note the pathologo-anatomical finding in one of the female patients, displaying changes in all endocrine glands: slightly enlarged hypophysis ranging from marked hypercrinism to formation of adenomatous structures of the eosinophilic and basophilic cells; enlarged thyroid revealing a histological appearance of a mainly macrofollicular struma, without evidence of hormonal activity; reduced weight of the pancreas with fibrosis, lipomatosis and scarce insular apparatus; the ovaries are enlarged with cystic-proliferative structure. Along the inner surface of the frontal bone stalactite-like bony processes with the dura mater adherent to them are present.

Discussion

Our observations show that young women are also affected by the MSM syndrome. The latter fact is in disagreement with the classical concept according to which this syndrome affects females chiefly in the fifth decade of life, but complies with more recent reports in the literature (1, 4, 8, 11). Intracranial hyperostosis is a symptom sine-qua-non in establishing the diagnosis. Such a hyperostosis is demonstrated roentgenologically in 1 per cent of the healthy women beyond 40 years of age (5, 7) — a fact considered as one of the essential arguments against the nosological differentiation of the syndrome (12). The diagnosis MSM syndrome is possible only provided the hyperostosis is associated with the listed above endocrine-vegetative, metabolic and nerve-mental disorders.

The adenomatous hyperplasia of adenohypophyseal eosinophilic and basophilic cells, observed in one of the female patients, confirms the finding made by Jules and Holo, respectively their viewpoint concerning the total hyperfunction of this particular lobe. The changes in the other glands represent a further support of such a possibility. The increased basal values of 17-KS, and the stormy reaction after ACTH stimulation recorded in some of the patients of the series point to a hyperfunction of the adrenal cortex. The grossly reduced ACTH reserve of the hypophysis indicates a hypophyseal dysfunction. The latter is diminished both in hypopituitarism, and in ACTH-secreting hypophyseal adenomas (cited by 5). Of no less interest is the fact that in one of the female patients elevated serum phosphorus values are present, associated with simultaneous increase of the sella turcica sizes to the uppermost limit of the norm, while in two other patients the phosphorus shows borderline values — 4.5 mg %. The data just referred to, compared with the eosinophilic cells' hyperplasia, render imperative the discussion of the role played by STH in the development of intracranial hyperostosis (10). Experimental evidence in this line is already available. Mortimer (cited by 7) was successful in producing a thickening of the cranial bones after injecting an extract from the anterior hypophyseal lobe. These observations prove the pathogenetic relationship between hypophyseal changes and intracranial hyperostosis.

The results of the vegetative-cardiac test indicate a state of vegetative dysregulation with a prevalence of parasympathicotonic reactions. The disturbance of thermoregulation which, in the series under review, presents in the form of asymmetry of skin temperature, constitutes a frequently met with symptom of the diencephalic syndrome. The capillaroscopic changes observed are likewise in compliance with those in the diencephalic syndrome. EEG changes are characteristic primarily of the functional involvement of deep truncal structures (the rostral part of the reticular formation and posterior hypothalamus). The data reported once again corroborate the role played by diencephalic structures in the pathogenesis of the syndrome.

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КЛИНИКО-ЛАБОРАТОРНАЯ ХАРАКТЕРИСТИКА МОРГАНЫ—СТЮАРТ—МОРЕЛЛЯ И ПАТОГЕНЕТИЧЕСКАЯ ОБОСОБЛЕННОСТЬ СИНДРОМА

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РЕЗЮМЕ

Исследованы 17 больных с синдромом Морганьи—Стюарт—Морелля в возрасте от 34 до 64 лет. У всех их было налицо ожирение различной степени, головные боли и внутричерепный гиперостоз. У 11 больных наблюдалась пониженная переносимость углеводов, вплоть до явно выраженного диабета у 3 из них. У 8 больных была установлена артериальная гипертония. Базальная секреция 17-кетостероидов (КС) была нормальной, но у 2 больных наблюдались периодические повышения выше нормы. 17-ОС были выше границы нормы у 3 болъных. После стимуляции АКТГ, 17-ОС повысились более чем в 2 раза у 5 больных, а у 2 — реакция была очень слабой. Гипофизный резерв АКТГ оказался сильно уменьшенным у 10 из исследованных 11 больных. У одной больной было отмечено увеличение фосфора в сыворотке крови, а у 2 других он был на верхней границе нормы. Турецкое седло оказалось на верхней грани нормы у одной больной, а у остальных оно было в пределах нормальных размеров. У всех больных был налицо вегетативно-дистонический синдром. ЭЭГ — показывает наличие диффузных изменений с наличием тета-волн и полиморфного альфа-ритма.

Обсуждается возможный патогенез синдрома, причем подчеркивается участие диэнцефальных структур. Высказывается воззрение о нозологической обособленности этого синдрома.