EMPTY SELLA SYNDROME

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Abstract. *Empty sella syndrome is an enlargement of the sella turcica (the bone structure in the lower part of the brain in which the pituitary gland is located) when the pituitary gland is of normal size or when its size is reduced. Patients with empty sella syndrome have a defect in the cerebral canal barrier, which normally separates the cerebrospinal fluid located around the brain from the sella turcica.*

As a result, the cerebrospinal fluid puts increased pressure on the pituitary gland and the walls of the sella turcica. The sella turcica may increase in size and the pituitary gland may shrink.

Keywords: *"empty" Turkish saddle syndrome, pituitary gland the level of prolactin, hormonal imbalances, hypothalamus, Luteinizing hormone (LH) Oxytocin , Prolactin .*

"Empty" sella turcica syndrome (ESTS) is a combination of a defect in the diaphragm of the sella turcica with penetration of the cerebrospinal fluid system into its cavity with neuroendocrine pathology. To clearly understand the mechanism of occurrence of the "empty" sella turcica syndrome, it is necessary to briefly dwell on the anatomical features of the latter.

The dura mater at the entrance to the sella turcica splits into two sheets. One of them lines the walls and bottom of the saddle, and the second closes the entrance to it, forming the so-called diaphragm of the saddle. In the center of the diaphragm there is an opening only for the passage of the pituitary gland stalks. Normally, the arachnoid membrane and cerebrospinal fluid do not penetrate into the cavity of the sella turcica. Its diaphragm is a dense, fully formed arch above the sellar cavity or may be underdeveloped, with a sharply enlarged infundibular opening.

Three main types of sella turcica diaphragm have been described: the first - in the form of a connective tissue plate with a hole in the center, allowing only the pituitary stalk to pass through, the second - when the diaphragm is not completely closed and has a hole around the pituitary stalk 3 mm, the third - characterized by the presence of only a narrow strip (2 mm or less) duplication of the dura mater along the periphery of the sella turcica.

It is customary to distinguish between the symptom and the syndrome of the "empty" sella turcica. The symptom is not clinically manifested and is detected on section. It was first described by Bush in 1951, who identified an "empty" pituitary fossa in 5.5% of cases in a sectional study of 788 people who died from various non-endocrine diseases. The combination of the symptom of an "empty" sella turcica with a neuroendocrine disorder is called the "empty" sella syndrome [2]. If the sella diaphragm is insufficient, fluctuations in cerebrospinal fluid pressure can lead to expansion of the cervical opening of the diaphragm, through which the pituitary stalk passes. In this case, the membranes of the brain are introduced into the cavity of the sella turcica under the diaphragm. The resulting "cerebrospinal fluid sac" pushes aside the pituitary gland, flattening and pressing it to the bottom or to the wall of the sella turcica.

Congenital underdevelopment of the sella diaphragm with extreme variability of the central opening is not uncommon and, according to various authors, occurs in 40-50% of pathological studies. As a rule, the shape of the sella turcica becomes cylindrical, its size increases, and the

back becomes thinner. The described signs can be detected on conventional radiographs of the skull in 84% of cases.

Among the reasons leading to the occurrence of primary syndrome of the “empty” sella turcica, the following are distinguished: congenital underdevelopment of the diaphragm of the sella turcica in combination with increased intracranial pressure; local increase in pressure in the ventricles, including the third, possible due to impaired patency of the liquor-conducting tract; transient hyperfunction and hyperplasia of the pituitary gland with subsequent involution (multiparous women), long-term hormone replacement therapy that can cause hypotrophy of the pituitary gland and its legs (including long-term use of oral contraceptives), arachnoid cysts that developed as a result of optico-chiasmatic arachnoiditis, spontaneous necrosis of pituitary tumors.

There is a secondary “empty” sella turcica, which occurs after surgical removal of a pituitary tumor or radiation therapy [4].

Thus, the “empty” sella turcica is a consequence of a long process that depends on many etiological factors. The cause of neurometabolic and endocrine disorders in this syndrome is considered to be not compression of the secretory cells of the pituitary gland, but compression of its stalk and disruption of hypothalamic control over pituitary functions as a result of periodically occurring disturbances in the supply of neurohormones from the hypothalamus [1].

By a variety and varying severity of pathological symptoms, in particular frequent headache, which patients cannot accurately localize due to the fact that irritation of a small area of the dura mater is transmitted through the intero - baroreceptor system to the entire dura mater. Possible visual disturbances - changes in color fields of vision, diplopia, narrowing of peripheral fields of vision (bitemporal hemianopsia). Some patients experience spontaneous liquor rhea as a result of non-closure of the ducts between the sella turcica and the oral cavity. Headache and blurred vision are most typical for secondary “empty” sella turcica.

The mechanism of occurrence of visual disorders also depends on the nature of previous therapy. In case of surgical intervention for a tumor, sagging of the optic nerves into the “empty” sella turcica may subsequently occur [3]. If a secondary “empty” sella turcica is formed after radiotherapy for a pituitary tumor, then subsequent changes in visual function may be due to the negative effect of radiation on the vascular system. Radiation therapy can provoke vasculitis and impair vision due to narrowing of blood vessels [3].

The clinical picture is determined by changes in the function of the pituitary gland in the form of panhypopituitarism , including isolated ACTH deficiency. Patients turn to a gynecologist about the consequences of changes in the functional state of the pituitary gland, most often its anterior lobe, in the form of menstrual dysfunction such as oligo- opsomenorrhea , amenorrhea-galactorrhea , infertility. Cases of myxedema, acromegaly, false hypoparathyroidism , obesity, etc. have been described. When the posterior lobe of the pituitary gland is compressed, diabetes insipidus is possible.

Diagnosis of “empty” sella turcica syndrome is quite difficult. Clinical manifestations of this syndrome are very similar to a tumor of the anterior pituitary gland. On survey craniograms in the lateral projection, an increase in the vertical and sagittal dimensions of the pituitary fossa, osteoporosis of the parts of the sella turcica, pressing of the bottom of the pituitary fossa into the main sinus are noted, that is, symptoms typical of a pituitary tumor. It is possible to use pneumoencephalography, computed tomography and MP tomography.

Fluctuations in neurometabolic and endocrine manifestations, the absence of unambiguous changes in pituitary hormones from analysis to analysis, a history of numerous pregnancies, traumatic brain injuries, and arterial hypertension should alert us to this pathology even in the absence of radiological signs of enlargement of the sella turcica [1]. The prognosis for “empty” sella syndrome is generally unfavorable, especially in the presence of visual impairment, amenorrhea, and symptoms of hypopituitarism.

The treatment uses complex resorption, dehydration, and vascular therapy. Treatment of neuroendocrine disorders should be based on the clinical picture, the dynamics of the level of pituitary tropic hormones and agreed with an endocrinologist. The question of the need for neurosurgical intervention with plastic surgery of the sella diaphragm is considered with the progression of visual disorders. Patients with menstrual dysfunction or who have undergone surgery for a pituitary tumor if hormonal therapy is ineffective should be referred to radiologists and endocrinologists for additional examination to identify the “empty” sella syndrome.

Purpose of the study: To study the neurological manifestations of the “empty” sella syndrome.

Materials and methods of research: Over the past 1 year, we have observed patients at the TashPMI clinic (9 people) aged from 10 to 18 years with headaches of unknown etiology. We studied the neurological status, carried out a survey radiography of the skull, ultrasound of the genital organs and thyroid gland, and, if indicated, computed tomography and magnetic resonance imaging of the skull and ultrasound of the adrenal glands. In 5 patients it was detected macroprolactinoma, 3 had microadenoma, and one had “empty” sella syndrome.

Here is our own observation.

Patient B., 17 years old, came to the clinic because of headache, blurred vision, cold intolerance, fatigue, and amenorrhea for 10 months. History: menarche at 12 years old, menstruation for 5 days every 26-28 days, regular, painless, moderate, body weight increased by 10 kg over 10 months. Macromastia, headache, mood instability, irritability, and increased premenstrual syndrome appeared.

In the neurological status, decreased vision, muscle hypotonia were noted, tremor of the fingers was detected in the Romberg position, the finger-nose test is performed with slight intention on both sides, tendon reflexes BR, PR are animated, ANS: the distal parts of the arms and legs are sweaty, cold to the touch, VKF are age appropriate.

Clinical and laboratory examinations were carried out: the general condition was not impaired; height - 154 cm, body weight - 83 kg; mixed type obesity; the skin is clean, no striae are detected; mammary glands are enlarged to Ma 3, milky discharge from the nipples; No pathological anatomical changes were detected on the part of the genitals.

Research results: On the craniogram: the shape and structure of the sella turcica is without features, dimensions - 12x11 mm. In the frontal bone, a rounded area of rarefaction is identified, from the lower contours of which canals of diploic veins measuring 12x16 mm can be traced.

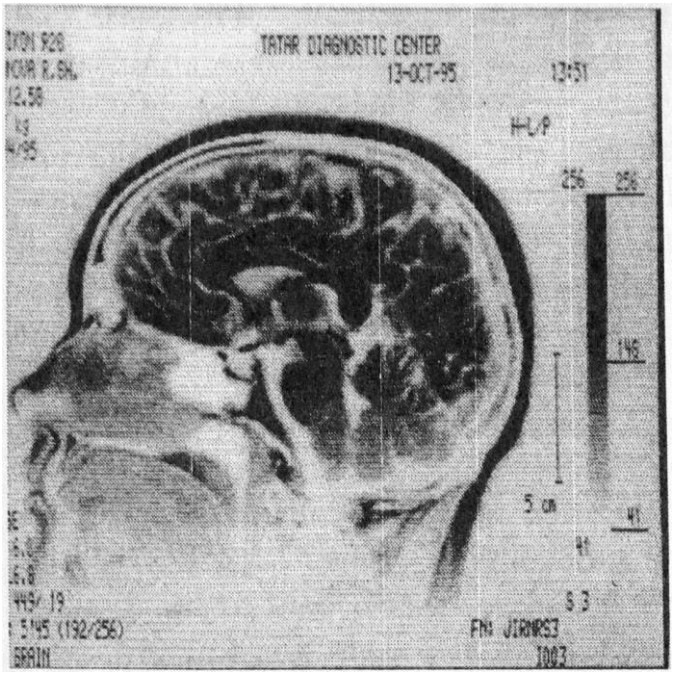
ECHO-encephalography: the cerebrospinal fluid pathways are not dilated, there is no displacement of the third ventricle. Electroencephalography: no pathology. Rheoencephalography: obstruction of venous outflow. Venous angiopathy.

Thyroid scintigraphy: within normal limits. Thyroid hormones: T3 - 1.73, T4 - 121.4, TSH - 0.15, TSH - 2.59 nmol / l, prolactin - 1616.72 μIU / ml.

Ultrasound of the thyroid gland: the gland is symmetrical, the contours are even, the left lobe is 37x17x19 mm, the right lobe is 36x16x20 mm, the bridge is 3 mm, in the posterior lobe there is a node up to 9 mm.

A neurosurgeon diagnosed prolactinoma : treatment with parlodel up to 7.5 mg per day was prescribed, with a positive reaction in the form of regular menstruation. The general condition improved, body weight decreased by 4 kg, and tissue swelling decreased. After 4 months, the dose was reduced to 5 mg per day and discontinued when the prolactin level reached 88.4 μIU /ml. Subsequently, after discontinuation of the drug, the prolactin level increased to 1527.79 μIU /ml, which again required the administration of parlodel. During treatment, prolactin levels quickly decreased, and menstruation returned to normal.

Subsequent magnetic resonance imaging showed the absence of focal changes and displacement of the midline structures. The ventricles are symmetrical and not enlarged. The sella turcica is enlarged and deepened, the phenomenon of an “empty” sella turcica and the replacement of the sella by the chiasmal cistern due to hypotrophy of the pituitary tissue are observed (see Fig.). This observation confirms the need to study the dynamics of prolactin levels during treatment in order to exclude the “empty” sella syndrome and the subsequent unjustified prescription of large doses of dopaminergic drugs. It is recommended to correct the sella diaphragm in consultation with a neurosurgeon.



MRI patient:

Conclusions: As such, there is no treatment method for empty sella syndrome. Drug therapy is aimed at eliminating the symptoms of the syndrome. Therefore, if the pathology is discovered completely by chance and does not bother the patient in any way, then it does not require treatment. In our case, the elimination of disorders of the endocrine and nervous system led to an improvement in the clinic.

It is impossible to predict the course of the disease. It all depends on the course of SPTS, concomitant diseases and the condition of the gland itself. Constant monitoring of hormone levels in order to eliminate complications in time. Engage in health promotion to help the body fight disease.

REFERENCES

1. Agarwal JK, Sahay RK, et al. Empty sella syndrome. Per. from English N. D. Firsova (2018)
2. Artykova D.M. Integrated assessment of risk factors for the formation of primary “empty” sella turcica syndrome link to source: https :// mrtpetrograd . ru
3. Babarina M. B., Kadashev B. A. “Empty” sella turcica (PTS) // Pituitary adenomas: clinical picture, diagnosis, treatment / Ed. prof. B. A. Kadasheva . — M.:: Triada Publishing House LLC, 2007; — P. 173—179. — 368 p. — ISBN 978-5-94789-258-1.
4. Egorov E. A., Tagirova S. B., Alyabyeva Zh. Yu. The role of the vascular factor in the pathogenesis of glaucomatous optic neuropathy // Klin . o phthalmology. - 2002; - No. 2: - P. 61-65.
5. Efimov A. S. Small encyclopedia of an endocrinologist. — 1st ed. - K.: Medkniga , DSG Ltd., Kiev, 2007; - pp. 334-335. — 360 s. — (“Library of a practicing physician”) — ISBN 966-7013-23-5.
6. Diseases of the hypothalamic-pituitary system // Materials of the Russian Scientific and Practical Conference, Moscow, 2001 / Ed. I. I. Dedova. - M.., 2001; — pp. 10–15, pp. 46–51.
7. Kirilyuk M.L. “Empty” sella turcica syndrome // Clinical endocrinology and endocrine surgery . 2012; No. 4 (41):
8. Monarch Disease Ontology release 2018-06-29sonu - 2018-06-29 - 2018.
9. “In the cranial cavity there are visual ...” // Normal anatomy of the human visual organ Archival copy dated January 4, 2010 on Wayback Machine
10. “Evaluation of the sella turcica...” // Evaluation of craniograms in patients with neuroendocrine syndromes Archived copy dated December 3, 2011 on Wayback Machine