Suprasellar mature teratoma: case report

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Intracranial teratomas are tumors that occur mainly in childhood and extremely rare in adulthood. They account up to 50% congenital CNS tumors. In this article a case of mature teratoma in 33 year-old female patient with progressive visual impairment is presented. Differential diagnosis at preoperative stage was difficult. Outcome analysis of neuroimaging study method and clinical picture provided evidence of epidermal cyst. Transnasal endoscopic approach as a treatment method was chosen, but during the surgery the atypical tissue for epidermal cyst was identified with tight adhesion to the right internal carotid artery which limited the extent of surgical treatment. Pathohistological and immune histochemical study detected mature teratoma. Detailed visual impairment dynamic and instrumental methods of diagnosis during postoperative supervision are presented in the article. The choice of management, namely, surgical intervention using extended endoscopic transnasal approach is considered to be controversial and risky among different authors taking into account intraoperative characteristics of this tumor.

Keywords: teratoma; germ-cell tumors; suprasellar tumors; endoscopic neurosurgery; transnasal endoscopic neurosurgery; skull base surgery

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area of basal ganglia and thalamus (4–14%) [2,4,9], cerebellar vermis, third ventricle, cerebello-pontine angle, cavernous sinus [8].

Intracranial teratomas often result in the obstruction of cerebrospinal fluid, so their clinical manifestations are hypertension, visual impairment, endocrine functions (polydipsia, polyuria, hypopituitarism, panhypopituitarism, premature puberty).

Diagnostics of ICT involves computer tomography and magnetic resonance imaging (MRI) of the brain (without / with contrast agent), hormonal tests, tests for tumor markers (α-fetoprotein and chorionic gonadotropin in the blood and cerebrospinal fluid). Computed tomography scans represent ICT both solid and cystic-solid formations, with a large proportion of fat and calcifications, have a sharply heterogenous structure of the solid part and irregular shape of the cystic part. During MRI without intravenous contrast enhancement, on T1-weighted MR images inhomogenous signal is revealed: hyperintense from adipose tissue and protein-fatty fluid, isointense or moderately hyperintense from the soft tissue component, hypointense from calcifications and blood components. With intravenous contrast, the soft tissue component accumulates the contrast agent. Inhomogenous signal from different tumor components is also detected on T2-weighted images. During neuro-ophthalmological evaluation it is possible to detect loss of visual field, decreased visual acuity, congestion in the ocular fundus.

Treatment of ICT is combined and involves surgical removal in case of immature teratomas, chemotherapy (cisplatin, etoposide, ifosfamide for immature teratomas and teratomas with malignant transformation), radiotherapy.

Clinical case

Anamnesis. A 33-year-old female patient was admitted to the department of the Institute of Neurosurgery named after Acad. A.P. Romodanov National Academy of Medical Sciences of Ukraine with complaints of visual impairment, narrowing of the temporal visual fields, weight gain. These complaints have been troubling for about a year with a gradual increase.

Objectively: the general condition is satisfactory. Increased nutrition. Neurological status: clear consciousness. Hypertensive symptoms are presented by mild cephalgia. Visual impairment, bitemporal narrowing of visual fields.

Neuroimaging. According to MRI of the brain (Fig. 1) parasellarly on the right, a mass lesion of the heterogenous structure of small size with a clear contour was revealed which spread antero-, suprasellarly and caused chiasm compression, but without a significant mass effect on the basal parts of frontal lobes. Perifocal edema is absent.

Multiple areas of increased MR signal on T2- and T1-weighted images were dominated in the mass structure, which in sequences with attenuation of the MR signal from adipose tissue (STIR) had a signal void corresponding to the fat component. The mass caused a restricted diffusion, as evidenced by the increase of the signal on the diffusion-weighted imaging (DWI, b-factor 1000) and heterogeneous with isointensive and hypointensive areas of the MR signal on ADC maps. The signaling characteristics on the DWI resembled those of an epidermal cyst.

Multiple small «point» subcortical and subependymal foci were visualized in the projection of lateral ventricles (mostly on the left), the MR signal of which also corresponded to adipose tissue, without perifocal edema and mass effect.

On post contrast T1-weighted images, an intense uneven accumulation of paramagnet by peripheral parts of the tumor was noted.

There were no signs of perifocal edema, an increase in the size of the cerebral ventricles and periventricular edema.

According to MRI, the patient was diagnosed with congenital anomalies such as arachnoid cyst and venous angioma in the left fronto-parietal region, with their typical MR characteristics.

Preoperative multispiral computed tomography (MSCT) was less informative and confirmed the presence of a well-defined supra-ante-parasellar mass lesion on the right, represented by areas of varying density, including adipose tissue, that did not accumulate contrast material. Multiple point subcortical and subependymal foci on MSCT were not clearly defined (Fig. 2).

Hormonal studies of blood serum: prolactin - 14 ng / ml (reference values - 1–27 ng / ml), growth hormone - 0 ng / ml (age-appropriate normal value - 0–20 ng / ml).
ml), cortisol (morning) - 612 ng / ml (normal range - 260–720 ng / ml), free T4 - 17.6 ng / ml (normal range - 11.5–23.0 ng / ml), α-fetoprotein in the blood - 10.2 ng / ml (normal range - <15 ng / ml), α-fetoprotein in the cerebrospinal fluid - 1.0 ng / ml (normal range - <1.5 ng / ml). Chorionic gonadotropin - 2.1 IU / ml (normal range - <5.0 IU / ml), chorionic gonadotropin in CSF - 0.18 IU / ml (normal range - <0.5 IU / ml).

Neuroophthalmological examination was performed by standard methods. Visual impairment in both eyes was shown (Vis OD = 0.4; OS = 0.3).

Examination of the visual fields revealed partial bitemporal hemianopsia (main defect OD = 9.17; OS = 4.31), examination of ocular fundus - a simple atrophy of both optic nerves (Fig. 3).

Surgical treatment. The analysis of clinical and instrumental methods of diagnosis showed that the patient had the highest probability of an epidermal cyst (cholesteatoma) of chiasmosellar area. Considering the localization and anatomic features of the tumor, transnasal transphenoidal binocular extended transtubercular endoscopic approach was chosen. Intraoperatively (Fig. 4), a large amount of adipose tissue was found in the lower (endoellar) part of the tumor, which also

**Fig. 2.** Preoperative MSCT

**Fig. 3.** Automated perimetry, which shows tunnel vision by the type of partial bitemporal hemianopsia
indicated in favor of cholesteatoma (epidermal cyst). However, during the internal decompression of the tumor while transitioning to the removal of the suprasellar part, a glandular yellow-pink anemic tumor was detected. The tumor tissue was tightly adjacent to the supraclinoid segment of the right internal carotid artery. The pituitary stalk was visualized and preserved. Subtotal resection was performed. Multilayer plastic repair of suprasellar cisterns with nasoseptal flap.

**Postoperative period.** In the first day after surgery, the patient is alert, without any motor deficit. Subjectively, she noted visual impairment. According to data of the controlled neuro-ophthalmological study the next day after surgery, visual acuity is reduced (OD = 0.2, OS = 0.4). In the field of vision of the right eye - absolute upper hemianopsia and relative temporal hemianopsia, in the field of vision of the left eye - upper quadrant temporal hemianopsia (Fig. 5). On the ocular fundus there is a primary atrophy of both optic nerves.

3 days after surgery, improvement in visual function was noted: visual acuity (Vis OD = 0.3; OS = 0.4-0.5). In the field of vision of the right eye - expansion of the upper border, in the field of vision of the left eye - relative upper quadrant temporal hemianopsia. On the ocular fundus there is the primary optic atrophy of both optic nerves.

Histological conclusion: mature teratoma (ICD-O code 9080/0) (Fig. 6).

Immunohistochemical study: CD45 (DAKO, clone 2B11 & PD7 / 26) - positive reaction (see Fig.6) in areas of small cell infiltrations, S-100 (DAKO, polyclonal) - negative reaction in areas of small cell infiltrations, p53 (DAKO, clone DO-7) - negative reaction in areas of small cell infiltrations, Ki-67 (DAKO, clone MIB-1) - positive reaction in single cells among lymphoid infiltrations.

After 2 months, during follow-up examination an improvement in visual acuity was revealed (Vis OD = 0,6; OS = 0,6), but the loss of visual fields remained unchanged (Fig. 7).

The mechanism of development of visual disturbances in the postoperative period, in our opinion, was associated with disturbed circulation in a chiasm.

One year after surgery, the general condition of the patient is satisfactory.

In postoperative period, the patient did not receive adjuvant therapy due to the results of histopathological examination.

Improvement in vision was noted. According to the control MRI of the brain 1 year after surgery, there were no signs of continued tumor growth (Fig. 8).

**Fig. 4.** Stages of surgical intervention: 1 - sphenoidal stage; 2 - a burr hole is formed in the area of tuberculum sellae, dissection of the dura mater in the projection of the tumor; 3 - endcapsular removal of the tumor; 4-5 - stages of dissection of the tumor capsule, detection of adhesion between internal carotid artery and tumor capsule (indicated by a blue arrow); right ON - right optic nerve; planum - site of the main bone; tuberculum sellae - Turkish saddle; CS - cavernous sinus; PG - pituitary gland; clivus - slope; paracilval ICA - paracilval internal carotid artery; tumor - growth; tumor capsule - tumor sac; adhesions with ICA - adhesion to the internal carotid artery; ICA - internal carotid artery; stalk - the stem of the pituitary gland
Fig. 5. Automated perimetry, which shows tunnel vision.

Fig. 6. Histologic specimen of the tumor. A - General histoarchitecture of tissue: epidermis, sebaceous glands. Hematoxylin and eosin staining. × 400; B - mesenchymal component of teratoma - expression of CD 45. × 800; B, G - proliferative activity, index KI-67. Immunopositive nuclei are colored brown. Reaction with KI-67 antibodies, hematoxylin staining. × 400
Discussion
The choice of surgical approach in removing teratomas remains controversial. MRI and MSCT of the brain suggested that the patient had cholesteatoma (epidermal cyst), which should be differentiated from suprasellar craniopharyngioma. Considering current trends in skull base surgery, the extended transnasal binocular transsphenoidal transsellar transtubercular endoscopic approach was chosen, which is optimal for the surgery of these tumors of suprasellar localization. Detection of yellow-pink glandular tissue intraoperatively gave reason to suggest that the histological structure of the tissue may not correspond to the epidermal cyst. Tight adhesion to the supraclinoid region of the right internal carotid artery did not allow to completely remove the mass due to the high risk of hemorrhagic complications in case of arterial damage. The assumption of the presence of teratoma when planning the surgical treatment would determine the choice of transcranial fronto-temporal basal approach, which provides a wider surgical corridor and the possibility of safer visualization and dissection of neurovascular structures from the tumor. According to current trends and the standardized GCT treatment protocol, the gold standard for the treatment of mature teratoma is the definitive surgical resection of the tumor without adjuvant therapy. Analysis of clinical cases indi-
icates the use of transcranial approaches as the safest and those that provide the maximum safe surgical resec-
tion. The use of transnasal endoscopic approaches has
become widespread over the past decade and represents
the gold standard for the treatment of such tumors of the
skull base as craniopharyngiomas, pituitary adenomas.
They are also widely used in the surgical treatment of
meningiomas of the anterior and middle cranial fossae
and tumors of the upper third of the slope. However, the
analysis of experience of domestic and foreign clinics
indicates that transnasal endoscopic approaches do not
provide an adequate volume of surgery for intracranial
teratomas of the skull base and their use is not the
method of choice. A thorough analysis of MR and CT
characteristics of the tumor and determination of tera-
Toma from its differential range allows to make a choice
in favor of transcranial surgical approach.

Conclusions

This case of surgical treatment of mature teratoma,
in our opinion, is interesting since the confidence in
the diagnosis of «epidermal cyst» actually led to the
choice of suboptimal surgical approach and treatment
strategy in general. MRI and MSCT data in such cases
are not the only criterion for diagnosis and planning
of surgical treatment. In case of suspicion of having
teratoma in a patient, it is advisable to use transcranial
basal approaches, which provide free manipulations
around the neurovascular structures of the skull base,
allow dissection of the tumor from them and total
removal with less risk of intraoperative complications.

Immunohistochemistry assay is important since the
differential diagnosis of a type of teratoma allows you
to make the right decision about adjuvant therapy,
including the absence of the need for it. Further follow-up
of the patient for 2 years (control MRI at 3, 6, 12, and
14 months) revealed no evidence of continued tumor
growth.

Disclosure

Conflict of interest

The authors declare no conflict of interest. Ethical approval

All procedures performed on the patient during the
study meet the ethical standards of the Institutional and
National Ethics Committees and Helsinki Declaration of
1964 and its later amendments or similar ethical
standards.

Informed consent

Informed consent was obtained from the patient.

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