ECRONICON OPEN ACCESS

EC NEUROLOGY
Research Article

Surgical Management of Orbital Malignant Neoplasms, Neoplasms of Cranioorbital, Craniofacial Localization

OI Lystratenko[1]*, AM Kardash[2], ZT Klymov[3], DO Lystratenko[4] and OF Smyrnova[5]

[1]Neurosurgeon of Donetsk Clinical Medical Association (DCMA), Ukraine

[2]Doctor of Medical Sciences, Professor, Head of the Department of Neurosurgery, Donetsk National Medical University (DNMU), Ukraine

[3]Candidate of Medical Sciences, Associate Professor of the Department of Otolaryngology, State University of Higher Professional Education, Donetsk National Medical University Named After M. Gorky, Ukraine

[4]Neurosurgeon of DCMA, Donetsk, Ukraine

[5]Doctor of Medical Sciences, Professor, Ophthalmological Clinic, Donetsk, Ukraine

***Corresponding Author:** OI Lystratenko, Neurosurgeon of Donetsk Clinical Medical Association (DCMA), Ukraine.

Received: April 08, 2021; **Published:** April 28, 2022

Abstract

The aim of the work was to highlight clinical manifestations and symptoms, histology and diagnostic methods, treatment of patients with MNO (malignant neoplasms of the orbit) and PS (paranasal sinuses) with growth into the orbit. Determination of the optimal surgical approach that allows radical removal of malignant neoplasms of orbital, cranio-orbital localization with maximum preservation of the functions of the visual analyzer. Materials and methods: the article considers and analyzes the results of treatment of 23 patients with malignant craniobital, orbital tumors and 35 patients with PS tumors that spread into the orbit. All patients, aged 2 to 85 years, were operated on in the clinic of neurosurgery and ENT diseases DCMA in Donetsk in the period from 2015 to October 2020. The conclusions were drawn: FOZ (frontal-orbito-zygomatic approach) is shown to remove orbital and cranio-orbital tumors of gigantic sizes, causing a sharp compression of the structures of the orbit, brain and tumors involving bone structures in the process. This type of operative approach allows performing organ-preserving operations with maximum radicalism, in relation to malignant neoplasms, with minimal complications, and maximum preservation of visual function.

An external (facial or sublabial) approach is optimal for the removal of the MN (malignant neoplasm) of the anterior PS group (frontal, maxillary, anterior ethmoid cells) that extend into the orbit.

In case of MN of posterior group (sphenoid sinus and posterior ethmoid labyrinth cells), as a rule, an endonasal endoscopic approach is used, which allows a good overview of this anatomical region. The previously proposed scheme for determining the choice of surgical approach to orbital neoplasms has successfully justified itself in practice [7,8], which is reflected in the results of treatment of patients with malignant tumors presented in the article.

Treatment of malignant neoplasms of the indicated location is always complex. Surgical treatment as the first stage. The second stage, depending on the histological response, is chemotherapy or radiation therapy.

Keywords: *Orbital Malignant Tumor; Ptosis; FOZ; Sub; Transconjunctival Approach; Canthotomy; Orbital Exenteration; RMS*

Abbreviation

FOZ: Frontal-Orbito-Zygomatic Approach; MN: Malignant Neoplasm; OMN: Orbital Malignant Neoplasm; PNS: Paranasal Sinus; MS: maxillary sinus; RMS: rhabdomyosarcoma; NHL: Non-Hodgkin's lymphoma; UA: Ultrasonic Aspirator; CT: Computer Tomography; MRI: Magnetic Resonance Imaging; USE: Ultrasound Examination; CXR: Chest X-Ray; TB: Trepan Biopsy; OND: Optic Nerve Disc; ON: Optic Nerve; RT: Radiation Therapy; PCT: Polychemotherapy; DOCTMO: Donetsk Regional Clinical Territorial Association

Introduction

Malignant neurosurgical tumors of cranioorbital, craniofacial localization is a heterogeneous group of neoplasms developing from various histogenetic tissue types may be accompanied by loss of visual functions and patient's disability [5]. They can be primary, secondary (growing into the orbit from adjacent anatomical structures, most often from the paranasal sinuses) and metastatic [3]. The most common primary malignant neoplasms are sarcomas, lacrimal cancer and lymphomas. [4] The number of patients with malignant neoplasms, which make up 1-3% of all malignant neoplasms, 23-25% of all neoplasms of patient's vision organ [6]. Currently, there is a tendency to expand the indications for osteoplastic orbitotomy as 20 years ago. Not only we, but also our foreign colleagues [16] carried out a retrospective analysis of osteoplastic orbitotomy complications. Microsurgical techniques, electro-, cranial drill**,** craniotomic ultrasound, cold plasma happened to minimize the number of functional complications and cosmetic defects. FOZ approach and its configurations used by us fully meet the requirements of cancer surgery and cranio-orbital localization.

Malignant neoplasms and cranio-orbital areas, PNS surgery in some aspects differs from the surgical ablasty principles of the other anatomical areas.

PNS tumors comprise 8 - 10% of all head and neck tumors (Jatin Shah, 2003). MN are most often localized in MS and make up 60 - 65% in the structure of PNS tumors (A.M. Sdvizhanov, 1998). Considering the anatomical peculiarities and the nature of the lymph outflow, PNS tumors rarely metastasize to the surrounding tissues and lymph nodes. More often this is continuation of the process with germination into the cranial cavity, orbit, pterygopalatine fossa. Treatment of patients with cancer, cranio-orbital localization and PNS should be combined. At the first stage surgical treatment is carried out with oncological ablation or palliative surgery principles, since MN PNS always occurs with purulent sinusitis due to sinus drainage function disturbance leading to purulent-inflammatory complications and impossibility or interruption of RT, PCT.

Aim of the work

description of clinical manifestations, diagnostic methods and treatment of patients with malignant neoplasms, cranio-orbital, craniofacial localization (NPS with growth into the orbit). Determination of the optimal surgical approach, allowing the removal of malignant neoplasms, of cranio-orbital localization with maximum preservation of the visual analyzer functions.

Materials and Methods

60 patients with malignant neoplasms of various localization and pathological and genetic structure operated on in the clinic of neurosurgery and ENT diseases DOCTMO in Donetsk were analyzed retrospectively from 2014 to November 2020.

Among them three groups were identified. The first group (I) included 20 (33%) patients with primary tumors located only within the orbit. The second group (II) comprised 5 (9%) patients with tumors of cranio-orbital localization with destruction of the skull bones. The third group (III) consisted of 35 (58%) patients with MN NPS with spread into the orbit (craniofacial localization).

Regarding 20 patients with primary orbital tumors: 8 (40%) were men and 12 (60%) were women aged from 2 to 79 years. 4 (13%) of them were children under the age of 10 - 2 boys and 2 girls. The number of patients with tumors of cranio-orbital localization with destruction of the skull bones was 5 men, 3 of them were 50 - 59 years, 2 of them - 70-79 years.

Among 35 patients with MN PNS group: 4 patients were diagnosed with the tumor with predominant growth in the area of the hard palate and the alveolar arch of the maxillary sinus; 5 people had the tumor in the frontoetmoidal area; 14 people had MN localized in the maxillary ethmoid area; 12 patients had MN in the posterior cells of the ethmoid labyrinth and the sphenoid sinus. According to gender difference there were 19 men (54.3%), 16 women (45.7%). All patients were aged from 50 to 70 years.

OMN - 20 (33%)

MN Cranioorbital - 5 (9%)

MN PNS -35 (58%).

Group	Age 0-19 Years		20-49 Years		50-69 Years		70-89 Years	
	M	W	M	W	M	W	M	W
Group I (20)	2	2			5	3	2	6
Group II (5)					3		2	
Group III MN PNS (35)								
1) Palate, alveolar arch, maxillary sinus (4)	1		1		2			
2)Maxillary- ethmoidal (14)					9	5		
3)Fronto-ethmoidal (5)					5			
4)Posterior ethmoidal (12)					9	3		

Table 1

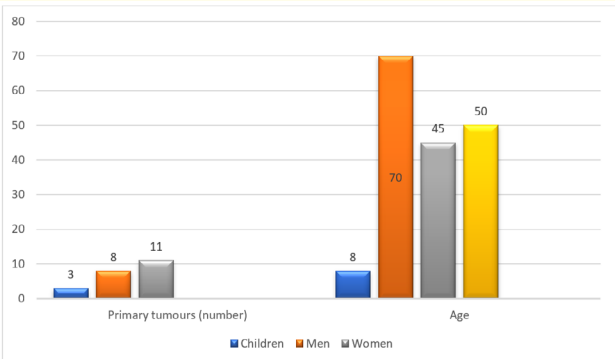


Diagram 1

All patients were examined according to our proposed algorithm: examination by an ophthalmologist, ophthalmoscopy, ultrasound of the eye, as a screening study, examination by an ENT doctor, neurologist. CT with intravenous contrast, MRI, to clarify the process extent; TB under ultrasound navigation or Brainlab navigation system, All body CT with contrast medium should be done to exclude metastasis.

Tools used: Computed tomography (Philips Brilliance 64), magnetic resonance imaging (Philips Ingenia 1.5T), ultrasound diagnostic machine (LOGIQ P5).

In the ophthalmic-neurological status of patients of the first (I) and second (II) groups, rapidly developing symptoms prevailed: unilateral ptosis, exophthalmos, more often with axial displacement, diplopia, progressive decrease in vision in one eye, severe pain in the eye with hemicranial spread.

All these symptoms were clear since a tumor growth in the orbit lead to compression and infiltration of intraorbital structures - all the nerves of the orbit, oculomotor muscles, vessels, which determined the clinical manifestations of the disease. Considering cancer and the cranio-orbital area of various pathological and genetic structure, we see no reason to differentiate the symptoms that are common for any type of cancer. Symptoms depended on the location of the MN and not on the pathological and genetic structure

Symptoms of MN germinating from PNS were most often manifested by the following triad: nasal congestion, bloody discharge from the nasal passage, purulent rhinitis (typical for MN of the anterior PNS group - frontal, maxilla sinus and anterior cells of the ethmoid labyrinth). It could be sometimes characterized by tetrad. Conductive hearing loss could be added on the side of the lesion typical for the posterior group of PNS -posterior lattices, sphenoid sinus). The characteristic symptoms of orbital lesion described above were noted with the rapid growth of the tumor.

Surgery was planned carefully taking into account the depth of the orbit [1], the size, location of the tumor, involvement of bone structures in the process, and the patient's ophthalmic-neurological status. Priority was given to organ-preserving surgery. Orbit exentration was condired to be as operation of choice. All patients in the postoperative period received PCT or RT depending on the histological response.

25 patients with malignant neoplasm, cranio-orbital localization were operated: by FOZ- 16 (40%) patients; transcutaneous subperiosteal- 5 (20%) people; transconjunctivally with canthotomy- 2 (8%) people; orbital exenteration - 2 (8%) patients.

Indications for FOZ

1. Sizes over 2.0cm;
2. Localization - the optic nerve canal, upper medial, upper lateral surgical angles of the orbit, intermuscular funnel, orbital tumors of a diffuse nature of large sizes.
3. Osteosarcomas of the orbit walls, malignant meningiomas spreading intracranially.

Indications for the use of transcutaneous subperiosteal, sub-, transconjunctival approach, by canthotomy:

To remove encapsulated MN up to 2.0 cm in diameter, open biopsy of diffuse tumor of the lower medial, lower lateral surgical angles. This approach is ineffecient when the orbital tumors are located in the intermuscular funnel area.

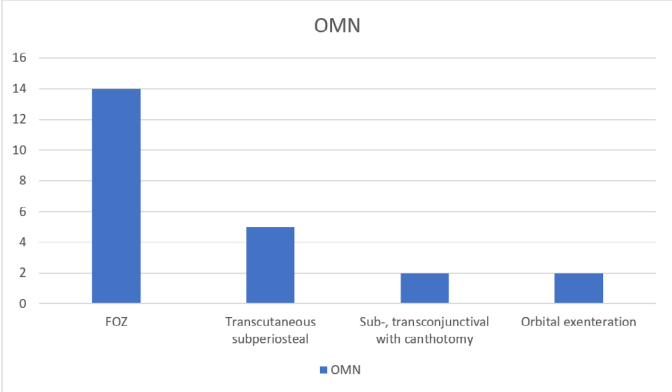


Diagram 2

35 patients with MN of the ENT group underwent the following surgical internventions: orbital exenteration, removal of the upper jaw and combined sinusotomy in 4 (11.4%) people; in 5 (14.3%) people, MN was located in the frontoetmoidal area - external frontoetmoidotomy; in 14 people, MN was localized in the maxillary ethmoidal region - of them in 9 (25.7%) people - external approach (Denkir, Moore, Subelau), in 4 (11.4%) cases - sublabial approach (sublabial lateronasal approach, Deglovin), in 1 (2.9%) cases, the maxillary sinus tumor was removed with an endonasal endoscopic pre-acrimal approach; in 12 (34.3%) patients, MN was located in the posterior cells of the ethmoid labyrinth and in the sphenoid sinus (endonasal endoscopic transnasal approach).

According to the histological analysis, we distributed clinical cases in table 2:

Malignant Tumors Of The Orbit, Craniofacial Localization (NPS With Growth In The Orbit)	Number
Neuroblastoma	2
Chondrosarcoma	2
Papilloma with malignancy	3
Osteosarcoma	1
Lymphoma	9
Rhabdomyosarcoma	2
Plasmacytoma	2
Adenocarcinoma	26
Melanoblastoma	2
Malignant meningioma	1
Anaplastic cancer	3
Hemangioperiocytoma	1
Undifferentiated Squamous Cell Carcinoma	4
Total	58

Table 2

The most common orbital tumors characteristics [2]

Sarcoma

According to the tissue there are angiosarcoma, fibrosarcoma, liposarcoma, myosarcoma, reticulosarcoma, neurosarcoma, rhabdomyosarcoma, chondrosarcoma, osteosarcoma. The most malignant is rhabdomyosarcoma, a tumor that develops from striated muscles. RMS is a highly malignant tumor that is more common in pediatric patients, although it can also develop in adults. It is more often localized in the upper inner quadrant of the orbit, infiltrates the levator and extraocular muscles early, leading to ptosis, displacement of the eye, and limitation of its mobility. In children, clinical symptoms increase within a few weeks, in adults - within a few months. Most often tumor growth is accompanied by severe pain syndrome. Hematogenous metastasis is possible.

An example of successful treatment of RMS in a 9-year-old child (Figs. 1,2,3,4 a,b) [17]. Microsurgical removal of the tumor by FOZ using ultrasound. Subsequent PCT, RT. The figures show the state before the operation, in 30 days after the operation, in 18 months, respectively.

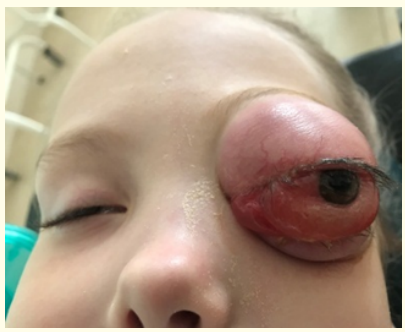


Figure 1: *Before surgery.*

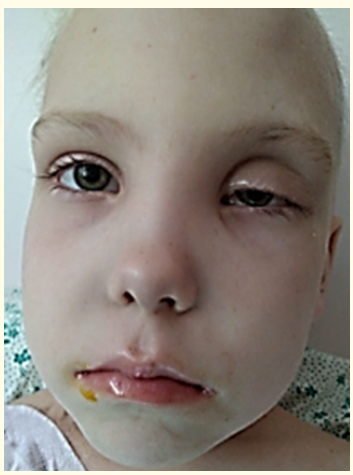


Figure 2: *30 Days after surgery.*

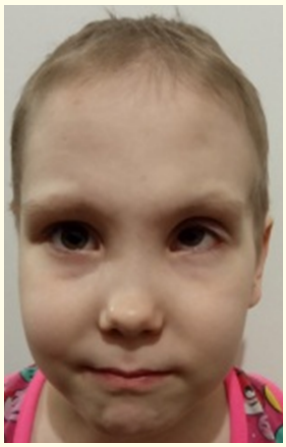


Figure 3: *18 Months after surgery.*

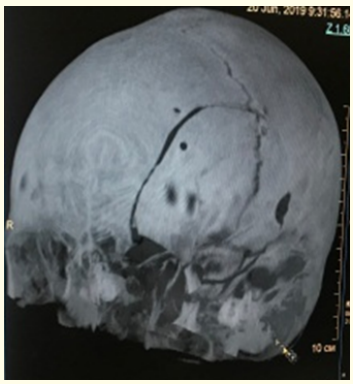


Figure 4A: *(FOZ). CT after surgery.*

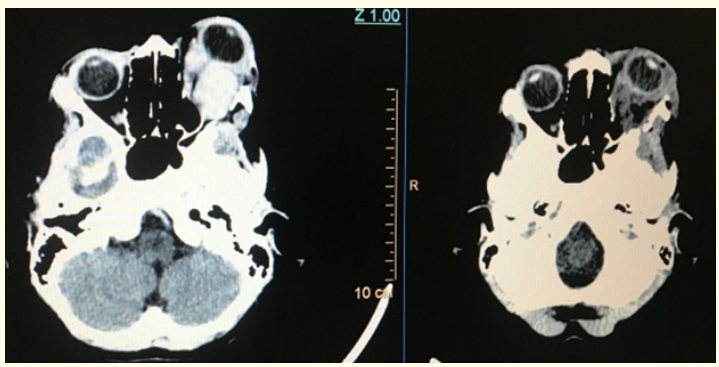


Figure 4B: *CT left before-, right- after surgery.*

Lacrimal gland cancer

Adenocarcinoma is differentiated morphologically in pleomorphic adenoma and adenocystic and mucoepidermoid adenocarcinomas that occurs spontaneously. As a rule, the tumor appears in adulthood, more often in women. Cancer in the pleomorphic adenoma of the lacrimal gland grows slowly over the years. Adenocystic cancer develops at a younger age, rarely in children.

An example of successful treatment of malignant pleomorphic adenoma in a patient aged 84 is presented. Microsurgical removal of the tumor was performed by FOZ. Binocular vision function restoration, excellent cosmetic effect is determined figure a,b,c.

Figure 5: *Before surgery.*

Figure 6: *14 Days after surgery.*

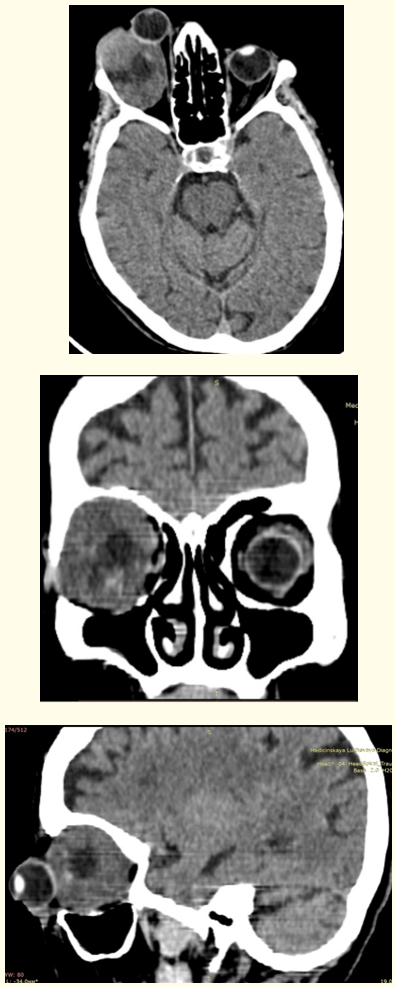
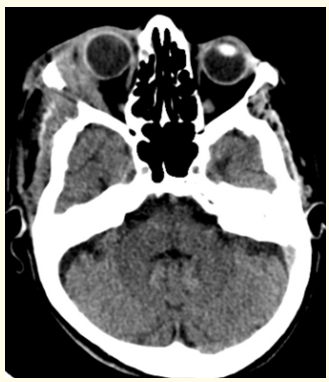


Figure 7A: *CT before surgery (axial, frontal, sagittal).*



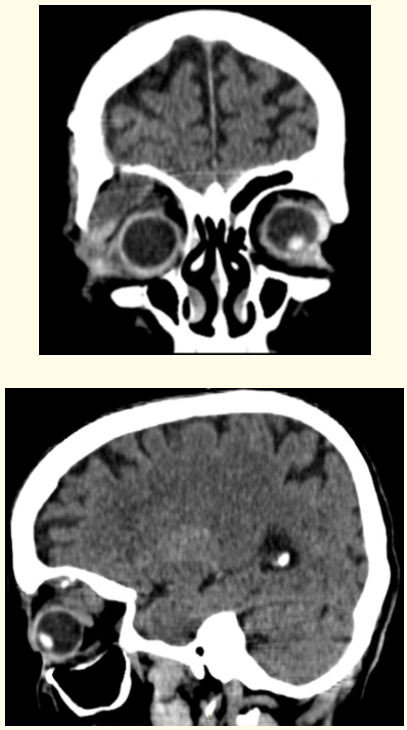


Figure 7B: *CT after surgery (axial, frontal, sagittal).*

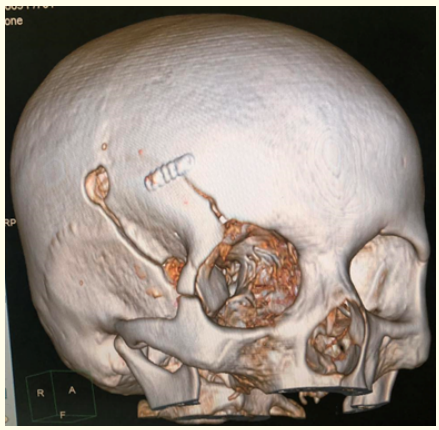


Figure 7C: *FOZ, CT after surgery*

NHL of the orbit is a heterogeneous group of malignant diseases of the lymphoid system,, they originate from the lymphoid tissue associated with the mucous membranes. Cases of primary localization of lymphoma in the orbit have become more frequent. The tumor affects one orbit more often. In 25% of cases, both orbits are affected with the high histological class of the tumor figure. 8 (a,в,c).

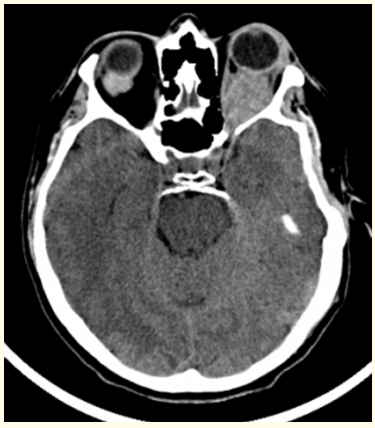


Figure 8A

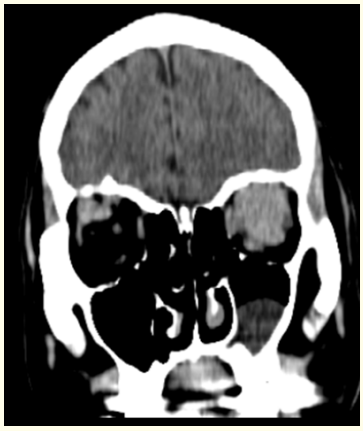


Figure 8B

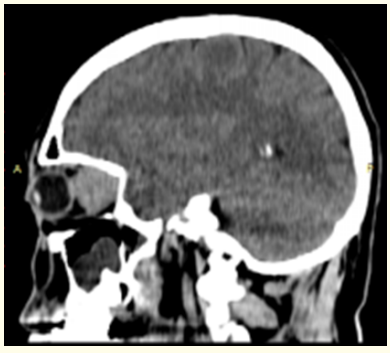


Figure 8C

There are diffuse infiltrative forms of growth with spread to muscles and optic nerves. If there is a local lesion of the orbit surgical, radiation or combined is administered after exploratory or cytoreductive orbitotomy. Immunohistochemical study of the removed neoplasm is mandatory (according to the clinical guidelines of "Association of Ophthalmologists", Russian Federation 2015).

NHL in the orbital area is most often diagnosed by TB with following PCT. The rapid growth of NHL causes a manifestated exophthalmos, accompanied by chemosis, pain syndrome, decreased visual acuity, initial stagnation of the OND in the fundus, simple atrophy of the ON. In this case surgical treatment is indicated to save the organ of vision - intraorbital decompression, decrease in the critical mass of the tumor, followed by PCT.

Metastatic orbital tumors - secondary damage to the orbital tissues associated with the primary localization of the tumor in other organs (mammary gland, lungs, etc.). Painless exophthalmos with displacement of the eye develops predominantly with metastatic lesions of the orbital tissues. Ophthalmoplegia and diplopia appear early.

Tumors growing into the orbit from adjacent anatomical structures - the cranial cavity (malignant meningiomas), from the paranasal sinuses.

The indications for FOZ were huge tumor size (in all 16 cases), cranio-orbital growth, bone structures damage (4 patients). Carcinoma was verified histologically in two patients with lesions of bone structures. Despite the complex treatment (PCT + RT on a linear accelerator), in 18 months, continued tumor growth with the orbital structure involvement, including bones, was diagnosed. In both cases the orbit exenteration was performed.

Squamous cell carcinoma was diagnosed in 4 patients of the ENT group with predominant growth in the area of the hard palate and the alveolar arch of the maxilla with expansion into the orbit. Orbital exenteration, maxilla removal and combined sinusotomy were performed. Subsequently, the course of LT, PCT was indicated. In all other cases, no relapses were noted during the observation period.

Minimally invasive approaches (transcutaneous subperiosteal, sub-, transconjunctival with canthotomy) in removal of OMN were more often used by us when the tumor was located along the inferior, inferior medial, and upper lateral edges of the orbit. It should be noted that canthotomy with this approach gives an excellent view of the lateral angles of the orbit, which increases the degree of radicality of removal of OMN. Mostly these were tumors of metastatic origin, NHL figure 9 a). figure 9.b).

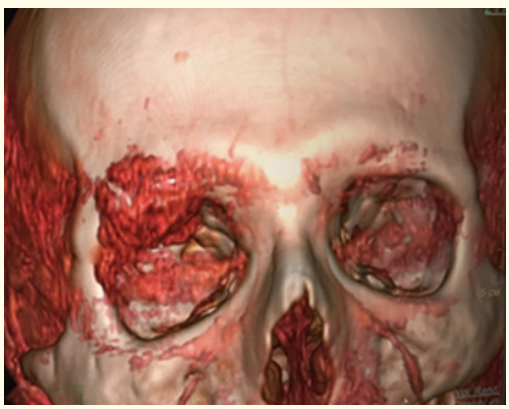


Figure 9A: *CT. Metastasis of glandular cancer.*

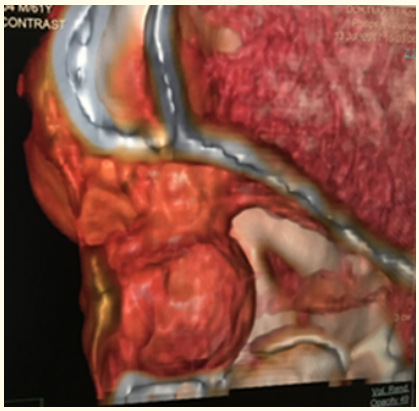


Figure 9B: *CT metastasis of glandular cancer.*

Absolute indications for orbit exenteration: 1. verified OMN, germinating orbital tissues throughout and leading to the death of the eye due to melting of the cornea; 2. intracranial spread of the process into the epidural space accompanied by severe pain in the hemifacial territory due to damage to the nerves of the orbit.

Here is one of the clinical cases of a rare type of melanoma. Uveal melanoma was diagnosed by the ophthalmologist with ophthalmoscopy, ultrasound. After enucleation melanoma was confirmed histologically. In 21 days a rapid tumor growth with severe pain syndrome and pain in the hemifacial territory was noted. After examination by the neurooncologist the orbit was exenterated in compliance with all ablation techniques to save the patient's life (Fig. 10 a, b). After discharge from the hospital, the patient was referred for treatment to an oncologist.

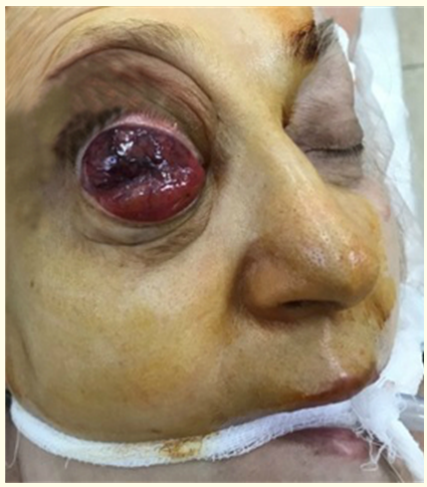


Figure 10A: *Before surgery.*

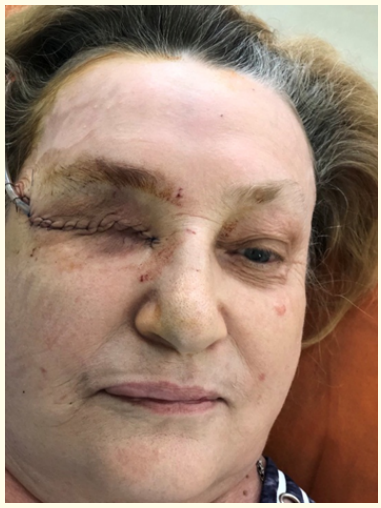


Figure 10B: *After surgery.*

Taking into account the worked-out surgical approaches that allowed removing OMN with the maximum degree of radicalism, we preferred organ-preserving treatment. OMN treatment is combined: operation + RT or complex (operation + RT + PCT). After complete removal of even a small malignant tumor from the orbit, RT, PCT was performed in the postoperative period, especially in children with RMS.

With OMN (more than 2 cm in diameter), most often, we gave preference to FOZ approach. The individual anatomical and topographic peculiarities of the orbit, the size of the tumor, its type, the degree of invasion into the orbital tissues, and the state of the bone walls were also taken into account. Despite the relative complexity of this approach we have a good overview of the orbital structures. To remove a tumor with a high degree of radicality and ablation a microsurgical technique, ultrasound, cold plasma were used. At the same time the most careful attitude to the structures of the orbit, their minimal traction, traumatization was preserved that was reflected in the final result of treatment. The most radical MN was removed (up to a "clean" surgical margin), the lower the frequency of local recurrence [18].

Conclusions

1. The results of treatment of patients with OMN depended directly on the degree of radicality of their removal, which is associated with the choice of surgical approach, the use of PCT, RT in the postoperative period. In our opinion, FOZ approach and its modifications, in combination with the use of ultrasound, cold plasma "Soring", bipolar coagulation for removal OMN and neoplasms in cranio-orbital localization provided an organ-preserving operation with maximum preservation of visual function, minimization of oculomotor disorders, patient's disability combined with a good cosmetic effect.

2. ENT approaches were most often used to remove tumors of the paranasal sinuses with spread into the orbit, requiring collaboration of ENT-doctor and neurosurgeon, since it was necessary to work with the skull base, intracranial spread.

3. Orbit exenteration is not a radical method of OMN and cranioorbital region management, as an independent method of treatment, since it does not affect the patient's survival rate in 5 years and is used in combination with RT, PCT. This type of operation should be considered as operation of choice. In our observations the number of patients with MN who underwent orbital exenteration is not large enough to draw conclusions about the survival rate of such patients within 5 years. Therefore, we used for conclusions the latest data published in the world literature [19].

The Authors Declare that they Have no Conflicts of Interest

Patients' consent to the processing and use of personal data, photographs is obtained, documented, and is in patients' case history.

Bibliography

1. AF Brovkina. Bolezni orbity. Moskva, «Meditsina», (1993): 240.
2. Goldberg SH and Cantore WA. "Tumors of the orbit". *Current Opinion in Ophthalmology* 8.5 (1997): 51-56.
3. Gloor B and Kalman A. "[Neoplasticspace-occupying lesions of the orbit.I. Review; hemangioma, lymphangiomaand embryonal rhabdomyosarcoma]". *Klinische Monatsblatter fur Augenheilkunde* 201.5 (1992): 291-301.
4. Sobin LH., *et al.* "International Union Against Cancer (UICC)". TNM Classification of Malignant Tumours, 7th edition. editions. New York: WileyBlackwell (2009).
5. Mannor GE., *et al.* "Multidisciplinary management of refractory orbital rhabdomyosarcoma". *Ohthalmology* 104 (1997): 1198-1201.
6. Weisman RA., *et al*. "Orbital tumors". *Otolaryngologic Clinics of North America* 34 (2001): 1157-1174.
7. Lystratenko OI., *et al.* "Clinical and anatomical rationale for the use of fronto-orbito-zygomatic (FOZ) approach for the surgical treatment of tumors of the orbit and cranioorbital region". *EC Neurology* 12.8 (2020).
8. Listratenko AI., *et al*. "Diagnostika i khirurgicheskoe lechenie opukholei orbit razlichnoi lokalizatsii i gistostruktury". *Vestnik Neotlozhnoi I Vosstanovitel'noi Khirurgii* 5.1 (2020): 93-108.
9. Shields CL., *et al*. "Comment in". *Ophthalmology* 110 (2001): 877.
10. Chang CW and LC Wang. "Orbitozygomatic approach for excisions of orbital tumors with 1 piece of craniotomy bone flap: 2 case reports". *Surgical Neurology International* 68 (2007): 56-59.
11. Rohde V., *et al.* "The combined pterional and orbitozygomatic approach to extensive tumors of the lateral and laterobasal orbit and orbital apex". *Acta Neurochir* 132 (1995): 127-130.
12. Bejjani GK., *et al.* "A reappraisal of surgery for orbital tumors. Part 1: extraorbital approaches". *Neurosurg Focus* 10.2 (2001): E2.
13. CL Shields., *et al.* "Clinical spectrum ofprimary ophthalmic rhabdomyosarcoma". *Ophthalmology* 108 (2001): 2284-2292.
14. Hassler W., *et al.* "Orbital Tumors: Diagnosis and Surgical Treatment". *Deutsches Ärzteblatt* 104.8 (2007): 496-501.
15. Schick U., *et al.* "Surgical management of meningiomas involving the optic nerve sheath". *Journal of Neurosurgery* 101 (2004): 951-959.
16. Nicola Boari Alfio Spina Lodoviga Giudice. "Fronto-orbitozygomatic approach: functional and cosmetic outcomes in a series of 169 patients". *Journal of Neurosurgery* 128.2 (2018): 466-474.
17. OI Lystratenko., *et al.* "Diagnosis and Surgical Treatment of Orbital Tumors of Various Localizations and Histostructures". *Archives in Neurology and Neuroscience – ANN* (2010): 2641-1911.

18. Mouriaux F., *et al.* "Survival after malignant tumours of the orbit and periorbit treated by exenteration". *Acta Ophthalmology* 77 (1999): 326-330.

19. Arnaud Martel., *et al.* "Orbital exenteration: an updated review with perspectives". *Survey of Ophthalmology* 66.5 (2021): 856-876.

Volume 14 Issue 5 May 2022