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Craniofacial malignant tumors, their extensions, and surgical strategy for their treatment

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Purpose: *To identify extension routes of craniofacial malignancies and formulate a surgical treatment plan based thereupon.*
Material and Methods: *We retrospectively reviewed the medical records of 253 patients with craniofacial malignancies who underwent surgical treatment at the Romodanov Neurosurgery Institute from 2002 through 2022. Of the 253 patients, 112 had a primary tumor, and 141, a secondary tumor. Preoperative Karnofsky performance scores ranged from 50 to 70 points. Patients underwent neurological and ophthalmological status assessment, as per routine protocols.*
Results: *Epithelial malignancies were the most common (53.7%), whereas anaplastic meningioma and embryonal malignancies were rather uncommon (1.2% and 0.4%, respectively) craniofacial malignancies. The presence of certain clinical symptoms was associated primarily with tumor origin and extension. A high rate of general brain and rhinological symptoms in our study sample was caused by a high percentage of intracranial and paranasal sinus tumors. Craniofacial malignancies most commonly originate from the midline (particularly, anterior midline skull base). Ethmoidal labyrinth was the most common site of origin (45.0%), followed by a sphenoid sinus (12.2%), pterygopalatine and infratemporal fossae (9.9%), whereas the cavernous sinus and olfactory fossa were the least common sites of origin (0.4% and 1.2%, respectively). Craniofacial tumors extended most commonly intracranially (transdurally, epidurally, via adhesion to the dura mater, and/or cavernous sinus growth) or intraorbitally. Anterior craniofacial resection (bifrontal craniotomy with combined with either lateral rhinotomy or supraorbital advancement; or a subcranial approach) was the most common surgical treatment. Postoperative cerebrospinal fluid rhinorrhea and infectious complications (meningitis and meningoencephalitis) were the most frequent complications. The overall postoperative mortality rate was 2.0%.*
Conclusion: *First, compared to the transcranial and facial approaches, the craniofacial resection is advantageous in terms of the radicality of tumor excision. Second, the subcranial approach is preferable to the bifrontal approach in the presence of marked extracranial tumor component, whereas the transbasal Derome approach is effective in the presence of marked extracranial and/or intracranial tumor components. Finally, both the orbitozygomatic and infratemporal approaches allow for the radicality of excision of lateral skull base malignancies, but the latter approach is associated with a lower rate of complications.*

Keywords:

malignant, craniofacial tumors, subcranial approach, orbitozygomatic approach, infratemporal approach

Introduction

Craniofacial malignancies mostly include epithelial tumors, bone and cartilage tumors, sympathetic ganglion neoplasms, peripheral nerve neoplasms and muscle neoplasms. They are uncommon head and neck tumors, accounting for 3% of all cases, with ethmoidal tumors comprising as much as 30% of all craniofacial malignancies. Intraorbital and intracranial extensions of ethmoid tumors are rather common [1-5]. Opinions vary with regard to formulating the plan of the most adequate surgical treatment on the basis of histological and biological features and extension routes of craniofacial neoplasms [6-9]. Radicality of excision is important and depends on the degree of tumor extension within paranasal sinuses and cranial base and the presence of intracerebral and intraorbital extension and extension into the cavernous sinus [4, 10-12].

The purpose of the study was to identify extension routes of craniofacial malignancies and formulate a surgical treatment plan based thereupon.

Material and Methods

We retrospectively reviewed the medical records of 253 patients with craniofacial malignancies who underwent surgical treatment at the Romodanov Neurosurgery Institute from 2002 through 2022.

The inclusion criterion was the presence of intracranial or extracranial extension of a craniofacial malignancy. Of these patients, 104 (41.1%) were women and 149 (58.9%) were men. Patient age ranged from 15 to 88 years, with a mean age of 43.4 years.

Of the 253 patients, 112 (44.3%) had a primary tumor, and 141 (55.7%), a secondary tumor. In addition, of the 253 patients, 69 were referred from outside institutions for their recurrence after craniofacial resection followed by radiotherapy and chemotherapy; 44, for poor outcome of non-adjuvant radiotherapy and chemotherapy; and 28, for continued tumor growth after ear, nose and throat (ENT) surgery.

General condition of patients was assessed by Karnofsky performance scores. Preoperative Karnofsky performance scores ranged from 50 to 70 points. Patients underwent neurological and ophthalmological status assessment, as per routine protocols.

Brain magnetic resonance imaging (MRI) (T1, T1Gd+, T2, and fluid-attenuated inversion recovery (FLAIR)) and multidetector computed tomography (MDCT) (including those with intravenous contrast enhancement) were used for pre- and postoperative tumor assessment.

Statistica 6 (StatSoft, Tulsa, OK) software was used for statistical analysis. Conventional parametric statistic methods were used for quantitative variables, whereas non-parametric statistic methods, for qualitative variables (expressed mostly in percentages).

Results

Major preoperative clinical symptoms were divided into 4 categories (non-focal and focal neurological symptoms; rhinological symptoms; ophthalmological symptoms; and otological symptoms), with their frequencies presented in Fig. 1. The presence of certain clinical symptoms was associated primarily with tumor origin and extension. A high rate of general brain and rhinological symptoms in our study sample was caused by a high percentage of intracranial and paranasal sinus tumors (Tables 2 and 3).

Epithelial malignancies (squamous cell carcinoma, transitional cell carcinoma, low-differentiated carcinoma, basal cell carcinoma, adenocarcinoma, and adeno-cystic carcinoma) were the most common craniofacial malignancies (53.7%), whereas anaplastic meningioma and embryonal malignancies were rather uncommon (1.2% and 0.4%, respectively).

Craniofacial malignancies most commonly originate from the midline (particularly, anterior midline skull base).

Table 2 shows the frequencies of various sites of origin of craniofacial malignancies.

Ethmoidal labyrinth was the most common site of origin (45.0%), followed by a sphenoid sinus (12.2%), pterygopalatine and infratemporal fossae (9.9%), whereas the cavernous sinus and olfactory fossa were the least common sites of origin (0.4% and 1.2%, respectively) (Table 2).

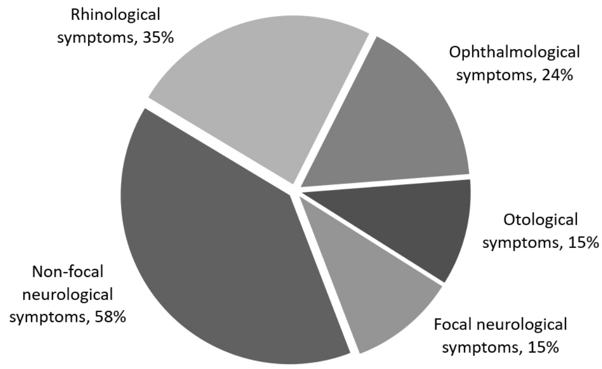

Fig. 1. Frequencies of categories of preoperative clinical symptoms in patients of the study

Malignant epithelial and vascular neoplasms originated from the paranasal sinus area. Sympathetic ganglion, paraganglion and muscle neoplasms arose from the pterygopalatine or infratemporal fossae. Malignant peripheral nerve tumors originated mostly from the ethmoid bone, nasal cavity or the floor of the anterior cranial fossa. The chordomas and chondrosarcomas arose from the clivus. Craniofacial meningiomas originating from the olfactory fossa were seen extending extracranially to involve the nasal cavity and/or paranasal sinuses or to the infratemporal and pterygopalatine fossae.

Table 3 shows routes of extension of craniofacial tumors.

Craniofacial tumors extended most commonly (188/253 cases; Table 3) intracranially (transdurally, epidurally, via adhesion to the dura mater, and/or cavernous sinus growth).

Table 4 shows resection types used for treatment of malignant craniofacial tumors, with anterior craniofacial resection being the most common.

The analysis of radicality of excision (Table 5) and complications (Tablr 6) depending on the surgical approach was performed to assess the efficacy of treatment for malignant craniofacial tumors. The percentages attributed to total tumor excision were 92.4% for bifrontal craniotomy with supraorbital advancement (the transbasal Derome approach) and 92.0% for transfrontal craniotomy (through the frontal sinus) (Table 5).

In case of significant intra- and extracranial extension of the tumor, anterior craniofacial resection was performed for advancement of intracranial tumor component, and transdural and intracerebral tumor extension and a portion of the floor of the anterior cranial fossa were resected, and extracranial tumor extension was excised.

Bifrontal craniotomy was performed in 36 cases. The transbasal Derome approach included bifrontal cranioto-

Table 1. Histological types of craniofacial tumors in patients of the study

Histological types of craniofacial tumors	Number of cases, n	Percentage from the total number of cases
Epithelial malignancies (squamous cell carcinoma, transitional cell carcinoma, low-differentiated carcinoma, basal cell carcinoma, adenocarcinoma, and adeno-cystic carcinoma)	136	53.7
Cartilaginous or osseous tumors (osteoblastoma, osteosarcoma, chordoma, and chondrosarcoma)	51	20.1
Peripheral nerve tumors (neurofibrosarcoma and esthesioneuroblastoma)	24	9.5
Sympathetic ganglia and paraganglia tumors (neuroblastoma, ganglioneuroblastoma, malignant chemodectoma, malignant paraganglioma)	19	7.5
Muscular tissue tumors (angioleiomyoma, leiomyosarcoma, and rhabdomyosarcoma)	10	4.0
Vascular and lymphoid tumors (hemangiopericytoma, hemangiosarcoma, and reticulosarcoma)	9	3.6
Anaplastic meningioma	3	1.2
Embryonal malignancies	1	0.4
Total	253	100

Table 2. Points of origin of craniofacial malignancies

Points of origin of craniofacial malignancies	Number of cases, n	Percentage from the total number of cases
Ethmoid labyrinth	114	45.0
Sphenoid sinus	31	12.2
pterygopalatine and infratemporal fossae	25	9.9
Maxillary sinus	20	7.9
Clivus	17	6.7
Floor of the middle cranial fossa	12	4.7
Orbit	10	4.0
Nasal cavity	7	2.8
External ear	7	2.8
Frontal sinus	6	2.4
Olfactory fossa	3	1.2
Cavernous sinus	1	0.4
Total	253	100

Table 3. Routes of extension of craniofacial tumors

Routes of extension of craniofacial tumors	Number of cases (percentage from the total number of cases)
Transdural (intracerebral growth)	49 (19.4)
Adhesion to the dura mater	61 (24.1)
Epidural	57 (22.5)
Periorbital adhesion and ingrowth with intraorbital extension	47 (18.6)
Destruction of the medial orbital wall	4 (1.6)
Growth into the cavernous sinus	21 (8.3)
Pterygopalatine and infratemporal fossae	21 (8.3)
Nasal cavity	52 (20.5)
Maxillary sinus	23 (9)
Sphenoid sinus	42 (16.3)
Frontal sinus	7 (2.8)

my with supraorbital advancement and was utilized in 66 cases. Endoscopic endonasal inspection (particularly, that utilizing wide-angle optics) of the wound for the presence of a residual tumor component at the paranasal sinus area was additionally performed in 32 cases. The subcranial approach with a trephination of the anterior frontal-sinus wall and removal of the posterior frontal-sinus wall was utilized in 37 patients. The endoscopic endonasal approach (40 patients) was utilized in cases with significant extension to the paranasal sinus area (the nasal cavity, maxillary, frontal and sphenoid sinuses, and ethmoid labyrinth) and medial extension to the pterygopalatine fossa. The transoral approach was utilized only in cases with middle skull base pathology, medial clival processes, and further caudal extension of the tumor at the level of the O, C1 and C2 vertebrae. The combined endoscopic endonasal and transoral approach was utilized in 19 cases with tumors of nasal or nasopharyngeal origin.

Table 4. Types of surgery in patients with craniofacial malignancies

Surgical approach	Number of cases, n	Percentage from the total number of cases, %
Anterior craniofacial resection: - bifrontal craniotomy with lateral rhinotomy - bifrontal craniotomy with supraorbital advancement (the transbasal Derome approach) - subcranial approaches: - transfrontal craniotomy (through the frontal sinus) - endoscopic endonasal approach - endoscopic endonasal approach combined with transoral approach - transoral approach	204 36 66 37 40 19 6	80.6 14.2 26.1 14.6 15.8 7.5 2.4
Lateral craniofacial resection: orbitozygomatic approach infratemporal approach	48 18 30	19 7.1 11.9
Petrosectomy	1	0.4
Total	253	100

Table 5. Radicality of resection for different surgical approaches to craniofacial tumors

Surgical approach	Number of cases, n	Gross total resection (GTR), n (%)	Subtotal resection (>90%), n (%)	Partial resection (50-90%), n (%)
Bifrontal craniotomy	36	19 (52.7)	9 (25.1)	8 (22.2)
Bifrontal craniotomy with supraorbital advancement (the transbasal Derome approach)	66	61 (92.4)	4 (6.1)	1 (1.5)
Rransfrontal craniotomy (through the frontal sinus)	37	34 (92.0)	2 (5.4)	1 (2.6)
Orbitozygomatic approach	18	14 (77.8)	3 (16.6)	1 (5.6)
Infratemporal approach	30	19 (63.3)	6 (20)	5 (16.7)
Endoscopic endonasal approach	40	25 (62.5)	5 (12.5)	10 (25.0)
Endoscopic endonasal approach combined with transoral approach	19	11 (57.9)	2 (10.5)	6 (31.6)
Transoral approach	6	3 (50.0)	3 (50.0)	-
Petrosectomy	1	1 (100.0)	-	-
Total	253	187	34	32

Table 6. Complication types and rates for different approaches to craniofacial resection

Complication type	Surgical approach						Total, n (%)
	1	2	3	4	5	6	
Cerebrospinal fluid (CSF) rhinorrhea, n	7	5	2	1	-	-	15 (6.0)
Cerebral infectious complications (meningoencephalitis), n	8	6	2	1	1	-	18 (7.1)
Brain prolapse in the craniobasal defect in the floor of the anterior cranial fossa, n	2	-	-	-	-	-	2 (0.8)
Intraorbital complications (oculomotor disorders)	2	-	-	2	-	1	5 (2.0)
Intraoperative hemorrhage and cerebral ischemic abnormalities, n	1	1	-	2	2	1	7 (2.8)
Total	20	12	4	6	3	2	47 (100.0)

Note: 1, bifrontal approach; 2, transbasal approach; 3, transfrontal approach; 4, endoscopic endonasal approach; 5, orbitozygomatic approach; 6, infratemporal approach

After resection of craniofacial tumor, the average transverse and anterior-posterior dimensions of a bony defect in the floor of the anterior cranial fossa (the lamina cribrosa (projection), planum sphenoidale, posterior frontal-sinus wall, and defect in the medial floor of the anterior cranial fossa) were of 4 cm and 6 cm, respectively. Plastic surgery for closure of the defect in the floor of the anterior cranial fossa was performed mostly with a pericranial periosteal flap from the frontal region of the head (115 cases). In addition, the defect in the floor of the anterior cranial fossa was closed with a flap from the temporal muscle and fat tissue in 5 cases each, and with a free fascia lata flap in 10 cases. Prolene (Ethicon, Somerville, NJ) mesh was placed on the defect in the floor of the anterior cranial fossa between the pericranial periosteal flap and the dura mater to improve the mechanical characteristics of the floor in 6 cases. The basal dural defect was repaired with a piece of fascia lata and periosteum in 34 and 27 cases, respectively.

Lateral craniofacial resection (48 cases) was performed when the tumor originated in the pterygopalatine and infratemporal fossae or nasal portion of the throat with extension to the middle cranial fossa and projections of the cavernous sinus, maxillary sinus and nasal cavity. In the series of cases reported here, the extent of surgical resection for the orbitozygomatic approach was higher than for the infratemporal approach (Table 5), although the difference in the extent of surgical resection was not significant ($p > 0.05$).

Complication types and rates for different approaches to craniofacial resection are presented in Table 6. Postoperative cerebrospinal fluid (CSF) rhinorrhea and infectious complications (meningitis and meningoencephalitis) were the most frequent complications (6.0% and 7.1%, respectively). The complication rate was higher for the bifrontal approach (accounting for half of total complications) than for the transbasal and other subcranial approaches. The complication rate for the orbitozygomatic approach was comparable to that for the infratemporal approach, with fewer complications seen in tumors treated by the latter approach.

The overall postoperative mortality rate was 2.0% (5/253 cases).

Discussion

Craniofacial tumors are difficult to manage, with their histological an biological features causing their infiltrative growth and origin [1, 2, 4, 6, 7]. Due to the oligosymptomatic course of early malignant craniofacial tumors, most cases are diagnosed and hospitalized in the advanced stages of the disease, with the tumors being large and showing marked extra- and intracranial extension. A lesion with both intracranial and extracranial extension requires formulating a plan for the special surgical treatment. A need arises for a team of surgeons to perform a simultaneous resection of intracranial and extracranial tumor components.

Based on the experience by Smith and colleagues [13], Ketcham and colleagues [14] described a bifrontal transcranial approach combined with lateral rhinotomy, which has become a conventional anterior craniofacial resection for patients with malignant craniofacial tumors.

This technique requires the involvement of neurosurgical, ENT surgical and other surgical teams as necessary for simultaneous access to both intracranial and extracranial tumor components [15].

We have started utilizing subcranial approaches in order to optimize surgical treatment of patients with malignant craniofacial tumors.

An advantage of subcranial approaches [16-19] compared with a bifrontal approach to malignant craniofacial tumors is that intracranial and extracranial tumor extensions are simultaneously exposed and a subcranial approach may be performed by a single neurosurgical team. However, a bifrontal transcranial approach should be combined with lateral rhinotomy to make an extractranial tumor component available to the surgeon during surgery. In the current study, subcranial (transbasal and transfrontal) approaches were comparable between each other, and superior to a bifrontal approach, in terms of gross total resection rate (Table 5). In addition, subcranial approaches were associated with decreased complication rates [16, 20-22], compared to a bifrontal approach (Table 6, statistically significant gamma statistics, $p < 0.05$).

Pure endoscopic endonasal approaches have been widely used in managing malignant craniofacial tumors [17-19], partially due to lower rate of surgical trauma compared to transbasal approaches. We believe that the difficulties in utilizing pure endoscopic endonasal approaches are associated with significant intracranial tumor extension (e.g., lateral tumor extension into the orbital roof), vascular involvement in the tumor, and the impossibility of using a periostal flap to cover defects in the anterior cranial fossa [23, 24]. In addition, the transbasal Derome approach [25] is still important in the management of lesions with marked intracranial extension [12], and, in the absence of a significant lateral intracranial extension of the tumor, may be reduced to the trans-fronatal approach (through the frontal sinus) to reduce surgical trauma and shorten surgery time [16, 26].

There have been reports [27-29] on the surgical treatment of patients with malignant pterygopalatine and infratemporal fossa tumors extending to the lateral skull base compartments, with discussions on the treatment of lateral skull base malignancies extending to the orbit, anterior cranial fossa floor, and maxillary sinus or dorsally to the petrous apex and carotid artery. Different authors [27-29] use the infratemporal or orbitozygomatic approach depending on the extension to the pterygopalatine and infratemporal fossae. The infratemporal approach is technically easier to perform than the orbitozygomatic approach given no need for orbitozygomatic advancement and reconstruction, which shortens surgery time. The orbitozygomatic approach is feasible in the presence of marked extracranial tumor extension (particularly to the pterygopalatine and infratemporal fossae), whereas the infratemporal approach, in the presence of intracranial tumor extension [30]. An additional resection of the floor of the middle cranial fossa may be performed for improved visualization of the pterygopalatine and infratemporal fossae.

Conclusion

First, compared to the transcranial and facial approaches, the craniofacial resection is advantageous in terms of the radicality of tumor excision.

Second, the subcranial approach is preferable to the bifrontal approach in the presence of marked extracranial tumor component, whereas the transbasal Derome approach is effective in the presence of marked extracranial and/or intracranial tumor components.

Finally, both the orbitozygomatic and infratemporal approaches allow for the radicality of excision of lateral skull base malignancies, whereas the latter approach is associated with a lower rate of complications.

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