RESULTS OF 15 YEARS OF COLLABORATION BETWEEN THE DEPARTMENTS OF OPHTHALMOLOGY AND STOMATOLOGY IN ONCOLOGICAL SURGERY OF THE ORBIT: A DIAGNOSTIC AND THERAPEUTIC APPROACH

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Dedicated to the long-term colleague prof. MUDr. Jiří Mazánek, Dr.Sc. in memorian

SUMMARY

Aim: To report an overview of the most frequent tumors of the orbit, suggest diagnostic approach and possible solution according to experience with own cohort of patients.

Material and Methods: From patients' files from the Department of Ophthalmology and Department of Stomatology, First Medical Faculty, Charles University, and General Faculty Hospital in Prague, Czech Republic, there were selected patients, who underwent the surgery due to the suspicion of malignant development in the orbit during the period 2005 – 2019. From the surgical records we found information about 497 cases. At the Department of Stomatology, there were 282 surgeries under general anesthesia performed, and at the Department of Ophthalmology, there were 215 surgeries, mostly under local anesthesia performed.

Results: The number of surgeries in men and women was equal; patients of all ages were present. The median of patients' age operated on at the Department of Stomatology was 53 years, and at the Department of Ophthalmology 63 years. The most common primary benign tumor was the cavernous hemangioma (9 %), the most common non-tumorous expansion was the dermoid cyst (7 %); the most common malignant tumor was the lymphoma (17.5 %). The last mentioned tumor was the most common diagnosis in the whole cohort as well.

Conclusion: Our cohort of patients is comparable with large cohorts published in the literature concerning age and gender distributions. Differences in frequencies of some lesions may be explained by that our cohort includes patients after the surgery only. The malignant lymphoma is the most common diagnosis indicated to surgical procedure, mostly biopsy. Comparing the two cohorts from our departments 20 years apart, the malignant lymphoma remains the most common indication for surgery, but the incidence of adenomas and adenocarcinomas of the lacrimal gland decreased. It is not the goal of this paper to evaluate all possible orbital affections. Suggested surgical approaches are just recommendations according to years of experience; however, in some situations, to choose an individual surgical approach is necessary.

Key words: orbital tumors, diagnostic approach, MRI studies of the orbit, surgery of the orbit

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INTRODUCTION

Orbital tumours are a relatively rare pathology. Primary benign and malignant tumours appear in the orbit, in addition to secondary tumours from cavities, bones, eyelids or the eyeball, as well as metastatic tumours. The diagnostic approach is derived from the symptoms and clinical picture, most frequently the patient visits a doctor due to exophthalmos, diplopia, malfunctions of vision or pain, less frequently the impulse for examination is deformation of the ocular aperture, swelling of the eyelids, reddening of the eyelids or conjunctivas, congestion on the optic nerve papilla or changes on the retina, or potentially a palpable lesion on the orbital entrance With the availability of imaging methods, we now identify orbital tumours more often than previously by chance, upon examination for other indications (incidentaloma). In differential diagnostics we may distinguish between "false tumours" (cysts, mucoceles) and inflammatory and non-inflammatory pathologies, above all endocrine orbitopathy (EO) and inflammatory pseudotumour of the orbit (PST). The examining and therapeutic procedure in most cases is multi-disciplinary. While examination should always be co-ordinated by an ophthalmologist, treatment is within the jurisdiction of the relevant specialist or team of experts with various specialisations. The precise incidence and prevalence of orbital pathologies can only be estimated, because the method of processing different cohorts is not uniform. The individual centres differ both in the catchment area and in their focus on a particular problem. However, it ensues from large cohorts that approximately one half of patients referred to orbital centres have EO, 20 % are patients with tumours (including cysts and mucoceles), 14 % have structural changes of the bone orbit (traumas and their consequences or congenital changes) and 9 % inflammations (acute infectious or chronic processes – PST) [1]. Approximately 2/3 of orbital tumours are benign and 1/3 malignant, but over the age of 60 years the percentage of malignant tumours increases. The most common malignancy in adulthood is non-Hodgkin's malignant lymphoma (ML), in children rhabdomyosarcoma, of benign tumours the most common are cavernous haemangiomas and dermoid cysts.

Examining procedure

Orbital pathologies threaten visual function and motility of the eye, even in the case of slow growth. A risk is posed also by surgical and non-surgical treatment. Careful examination is therefore necessary in order to ensure the correct choice of therapeutic approach. The basis of the examining procedure is recording of the patient's medical history. In the general anamnesis we focus on non-specific symptoms (fatigue, subfebrile temperature, repeated infections, fluctuations of body weight, enlargement of nodes...) and we also conduct a detailed traumatological, endocrinological and oncological **anamnesis**. In the ocular anamnesis we focus attention on changes in the face, we determine the main and secondary symptoms, the period of their duration and dynamic over the course of time, dependency on external influences, head position etc. In the case of conspicuous changes of physiognomy and minimal subjective complaints, the orbital pathology probably increases in size very gradually. By contrast, when accompanied by pains and obtrusive symptoms, the growth of the lesion is rapid. This can be verified by a comparison with older photographs. We target our inquiries precisely in order to ensure that the patient understands the formulation of our questions.

There follows a **complete ocular examination**. Upon a stated deterioration of vision it is necessary to exclude hypermetropisation of the eyeball (in the case of pressure on the posterior pole of the eye) or the induction of astigmatism (in the case of lateral pressure, e.g. by a tumour of the lachrymal gland). By means of examination of the visual field we exclude compression of the optic nerve, especially in the case of lesions in the apex of the orbit, or in the channel of the optic nerve. An analysis of binocular functions will show the scope of any applicable malfunctions of motility and sensorimotor adaptation to change of position of the bulb in the orbit. Upon examination of the face, we observe symmetry of the cheeks, ocular apertures, eyelids and temple areas (in women bulging may be masked by hairstyle). We measure the size of the protrusion, as well as any applicable shift in the frontal plane, thus dislocation (Fig. 1, 2), which must be differentiated from deviation of the eyeball in the case of malfunction of motility. In motility malfunction, in the field of action of one muscle it is difficult to deter-

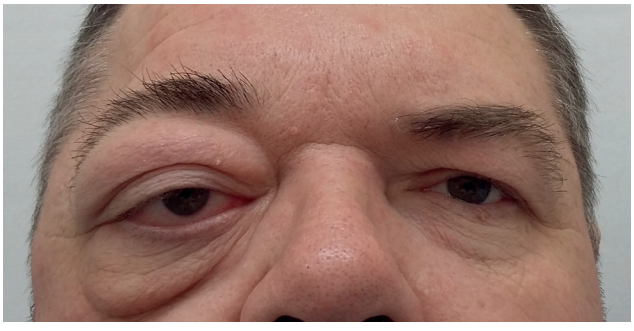

Fig. 1. Axial exophthalmos, baggy swellings of the eyelids and bulging of the temple area in the case of pterional meningeoma in right eye

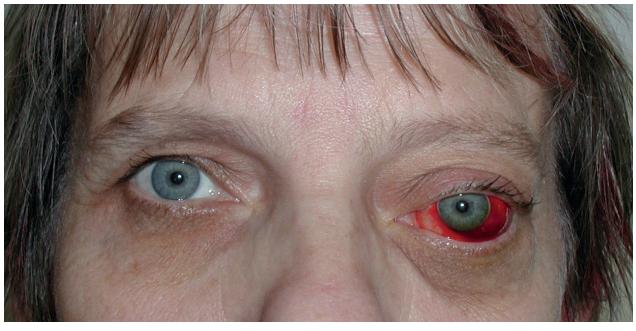

Fig. 2. Paraaxial exophthalmos with downward shift of eyeball in lymphangioma in the upper intraconal space in the left eye. Subconjunctival suffusion following spontaneous haemorrhage

mine whether the cause is neurogenic or myogenic, or mechanical. Upon restriction of motility of the eyeball in more than one direction, the cause is more often neurogenic, and according to combination with the affliction of other than the oculomotor nerves (n. II, n. V/1,2) it is possible to attribute it to localisation of the lesion (superior orbital fissure syndrome, orbital apex syndrome or cavernous sinus syndrome) [2]. Anisocoria is a manifestation of a malfunction of the efferent part of the pupillomotor reflex, relative afferent pupillary defect (RAPD) attests to compression of the optics. By examination of the conjunctiva we exclude changes in the region of the lachrymal gland and subconjunctival spread of the tumour, most often malignant lymphoma [3,4]. We conclude the external examination with palpation of the orbital entrance or eyelids in order to exclude resistance to accessible biopsy from anterior orbitotomy. We verify any disputed finding by means of pressure on the orbital entrance (or eyeball) outside the location of the envisaged tumour. By increasing the pressure in the orbit we displace the resistance against the finger of the other hand. On the ocular fundus we examine for possible changes of the papilla (from hyperaemia, via congestion to atrophy, in rare cases we identify growth of the tumour into the eye), or we search for cilioretinal vessel and arteries (pathognomic for meningioma of the optic sheaths). Pressure on the wall of the eye is the cause of chorioretinal folds, most commonly on the posterior pole, less frequently in other localisations. Increased intraocular pressure may be caused by worsened venous swelling upon overpressure in the orbit. The outcome of the ocular examination is then a working diagnosis and indication for other paraclinical examinations (laboratory and imaging). Sometimes in parallel we also indicate an inter-disciplinary medical consultation, most often endocrinological. With regard to urgency, it is possible to resolve the majority of ocular tumours within the space of weeks, at other times we merely observe a known tumour over the long term. Ruptures of cysts (spontaneously or following trauma) require an acute solution, in which the content of the cyst generates an acute inflammatory reaction. It is necessary to differentiate these conditions from orbital cellulitis infectious etiology. The cause of acute enlargement of a protrusion (or simultaneous subconjunctival suffusion and haematoma of the eyelids) may be haemorrhage into a lymphangioma, neuroblastoma, from vascular malformations or in the case of haematological pathologies. The most common secondary complications are changes on the cornea caused by exposure in the case of lagophthalmos and/or chemosis of the conjunctiva. Rhabdomyosarcoma and sometimes also PST may have an acute course in children, and in adults malignant melanoma may be acute. It is necessary to differentiate EO and carotid-cavernous fistula from acute deterioration of the condition generated by a tumour.

Upon selection of an **imaging examination** we start out from the envisaged cause of the symptoms. **Sonography** is the most accessible, fastest and in the hands of an experienced specialist also the most reliable method for examination of the anterior 2/3 of the orbit. A disadvantage is its inferior reproducibility and only referential information concerning the relationship to the other structures of the orbit.

Computer tomography (CT) is suitable for referential

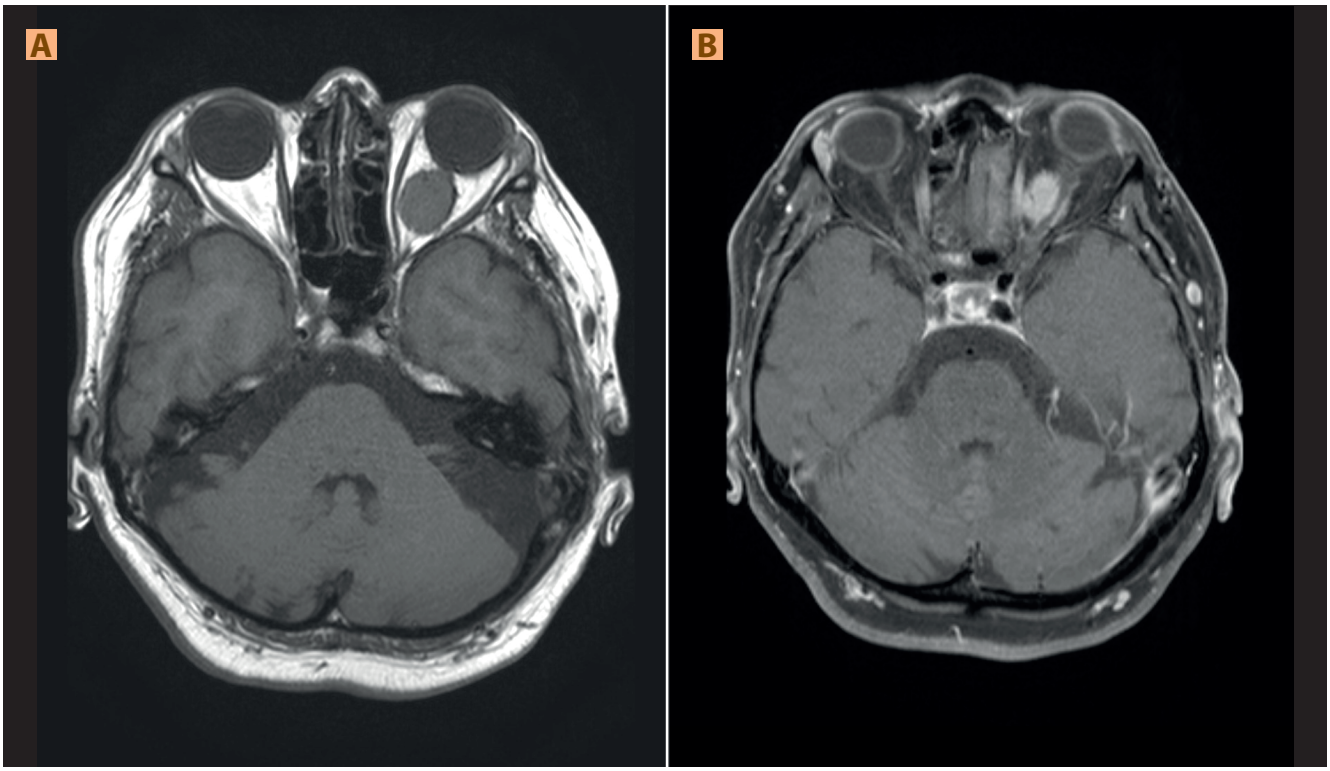

Fig. 3. Meningeoma of optic nerve sheath: sharply bordered formation encasing optic nerve, (A) isosignal in T1WI and (B) post-contrast saturated in T1SPIR

assessment of the content of the orbit, it is essential for a precise evaluation of the condition of the bones, and demonstrates any applicable calcification. Upon use of a contrast substance (CS) we obtain information about the filling of the affected area with blood. CT examination is also irreplaceable for patients who have contraindication for magnetic resonance imaging (MRI).

Magnetic resonance in imaging of orbital tumours

MRI is a suitable method for displaying soft-tissue expansions of the orbit, it has highly sensitive resolution of intraorbital structures, and is therefore ideal for evaluation of the localisation, scope and tissue characteristics of pathological tumorous mass. With the use of MRI we are also able to assess any applicable intracranial spread of the tumour, in which it is possible at the same time to detect also other potential intracranial pathologies. In addition to generally known contraindications for MRI, from an ophthalmologist's perspective it is necessary to avoid examination by this method in patients with intra or periorbital metallic bodies. Problems with quality of imaging are sometimes caused by cosmetics, whose pigments often contain metallic oxides, and some then manifest a magnetic moment which may lead to deformity of the MR image of the orbital structures. Artefacts from tattoos may be generated upon a similar background. It is also recommended to remove contact lenses, jewellery and piercings before the patient is brought into the magnetic field.

The standard protocol for MRI of the orbit is composed of 3 mm axial and coronary cross-sections, mostly accompanied by cross-sections on a sagittal plane (which often tilt in parallel with the course of the optic nerve and are therefore performed separately for each orbit). T1 and T2 sequences (T1 and T2WI) are routinely produced, always with suppression of the fat signal (according to the type of used instrument indicated e.g. as SPIR/STIR or FATSAT). The protocol also frequently involves the application of a contrast substance (indication of sequence T1GDSTIR/SPIR or T1GDFATSAT). In indicated cases a dynamic study is performed for the purposes of a more detailed assessment of vascularisation of the tumorous mass, the principle of which consists in repeated acquisition of data in several cross-sections leading through the largest diameter of the tumorous mass, enabling the observation of speed and intensity (or character) of saturation with the contrast substance in the observed place, or to construct a curve from this data, expressing their mutual relationship. A component of the protocol is also imaging of intracranial structures (usually in T2WI natively and T1WI natively and post-contrast). Upon suspicion of vascular malformation or aneurysm, MR angiography (MRA) is a suitable choice, either natively by the TOF method or as post-contrast MRA.

Fundamental MRI characteristics of the most common orbital lesions in relation to differential diagnostics:

Optic glioma is displayed as a spindle-shaped extension of the optic nerve, isosignal in T1WI, with slight post--contrast saturation, slight hypersignal in T2WI [5].

Meningeoma from the optic nerve sheath on axial cross--sections typically encases the optic nerve in a rail-like formation, in coronary cross-sections it surrounds it like a disc, with post-contrast saturation. With the use of MRI it is possible also

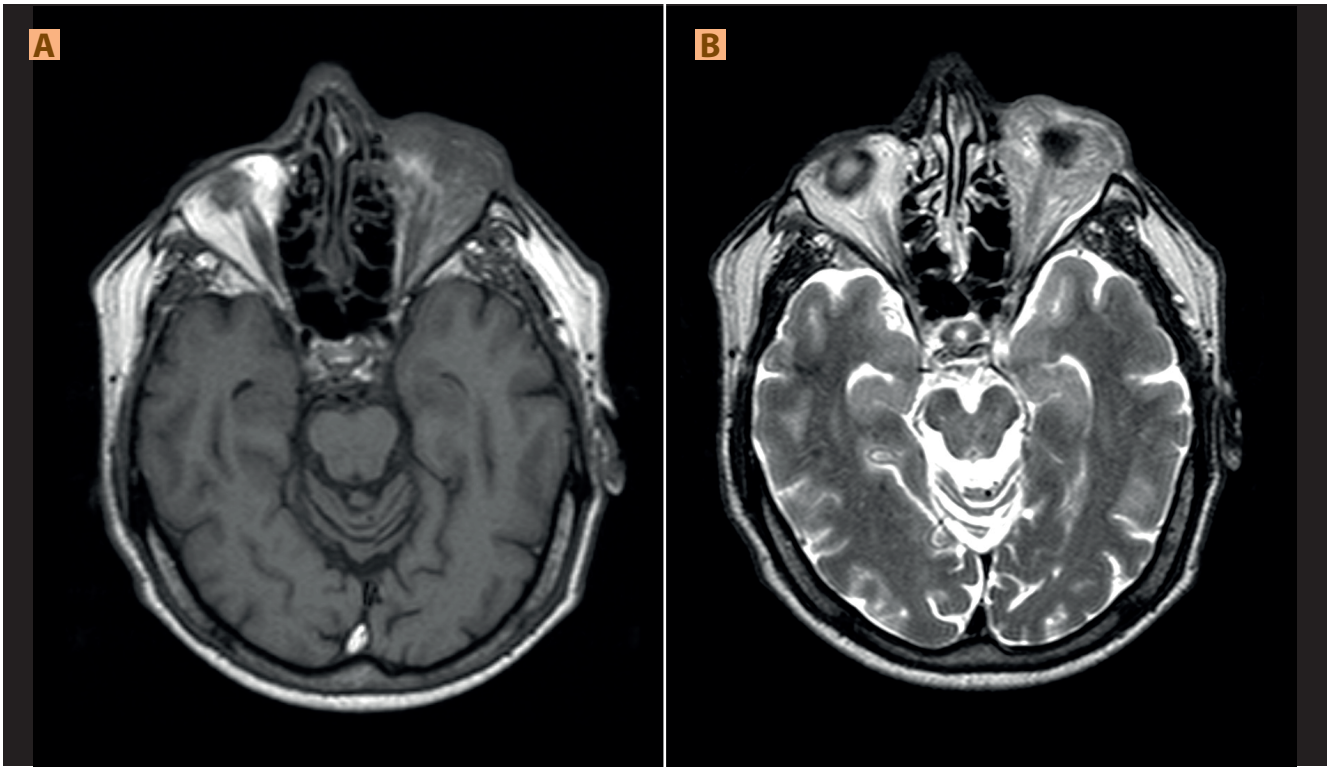

Fig. 4. Pseudotumour (A) non-homogeneous hyposignal infiltration of left orbit in T1WI and (B) non-homogeneous hypersignal in T2WI, protrusion of left eyeball

to identify any propagation of the meningeoma via the orbital channel to the intracranial meninges, and in rare cases also to the meninges of the contralateral optic nerve (Fig. 3).

The extraocular muscles are most frequently afflicted within the framework of endocrine orbitopathy (Graves-Basedow disease). We usually observe changes on the musculus rectus medialis and inferior or musculus rectus superior, in rarer cases on the musculus obliqus superior and musculus rectus lateralis [6]. Exophthalmos is conditioned by hypertrophy of these muscles and enlargement of the volume of retrobulbar fat. The muscles have a spindle shape, the tendons are normal. In MRI the areas of fat tissue in the muscles are hypersignal in T1 and T2WI and hyposignal in sequences with suppression of the fat signal (SPIR/FATSAT), whereas the inflammatory component is hypersignal in T2WI with and without fat suppression. The fibrous portion is hyposignal in T1 and especially in T2WI.

In differential diagnostics, in addition to EO, inflammatory pseudotumour of the orbit (Fig. 4) or non-specific myositis most often come into consideration. The latter typically affects the musculus rectus medialis and lateralis, in rarer cases also the lachrymal gland, uvea and sclera. Inflammatory infiltration is hypersignal in T2SPIR/T2 FATSAT, conspicuous post-contrast saturation then stands out in T1SPIR/T1 FATSAT sequences. If acute inflammatory infiltration passes into the chronic phase, the hypersignal in T2WI is generally replaced by hyposignal, which corresponds to fibrous metamorphosis.

Differential diagnostics of infiltrative tumorous processes afflicting the extraocular muscles, lacrhymal gland and other structures cover haematological malignancies, especially lymphomas (Fig. 5). In addition to hypersignal in T2WI and post-contrast saturation in T1WI, in MR image they are typified by restriction of diffusion (hypersignal on DWI and hyposignal on ADC – apparent diffusion coefficient – map).

Cavernous haemangioma (Fig. 6) is generally considered the most common benign orbital tumour, displayed as a sharply bordered, usually ovoid or rounded lesion in T1WI isosignal. Upon reading of native results of MRI, it may thus be confused with other lesions of a similar appearance. As a result it is appropriate to focus on the T2WI image, in which it is mostly possible to observe coincidence of hyperintensity with homogeneity, or it is possible to examine for internal septation of the tumour or a hypointensive border, which corresponds to a fibrous pseudocapsule. This sometimes differentiates cavernous haemangioma from other tumours (e.g. schwannomas). However, it is more reliable to distinguish cavernous haemangioma in a dynamic study (DCE MRI), since in the T1WI image it has a specific character of saturation, ensuing from a single point [7,8].

Lymphangioma (Fig. 7) is a tumour occurring in childhood age, complicated by frequent haemorrhage, in MRI imaging a multilocular septated tumour, hypersignal in T2WI, often with blood breakdown products, therefore hypo and hypersignal in T1WI. Hypersignal in T1WI may be conditioned also by the content of proteins, and upon haemorrhage the liquid content in the lacunas may form various signal intensities.

In the case of a positive oncological medical history (most often breast or lung cancer), it is necessary to consider the metastatic origin of the pathological process

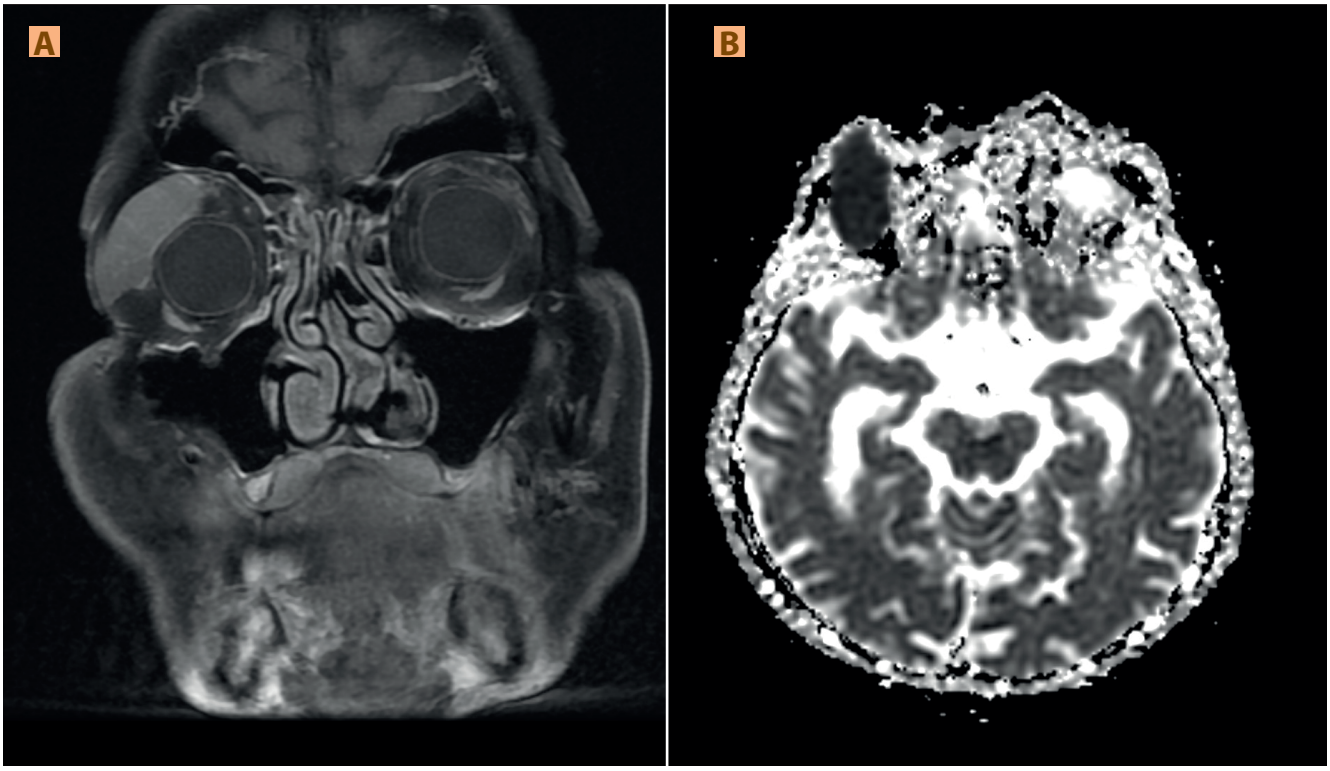

Fig. 5. Lymphoma affecting the lachrymal gland: (A) post-contrast hypersignal in T1FS, (B) hyposignal in ADC map

of the orbit. Metastasis in MRI image is generally iso to hyposignal in T1WI, with post-contrast saturation, and hypersignal in T2WI.

Rhabdomyosarcoma is a mesenchymal tumour occurring in children, manifesting itself in a rapid onset of exophthalmos. In the MRI image isosignal in T1WI, hypersignal in T2WI, with post-contrast saturation and possible content of the haemorrhaged areas.

The cause of rapidly appearing exophthalmos may also be intraorbital haematoma, in which the signal characteristics change according to its age, depending on the degradation of haemoglobin breakdown products. In the acute stage, the intensity of the blood signal is increased in T1WI, reduced in T2WI (intracellular methemoglobin), after seven days

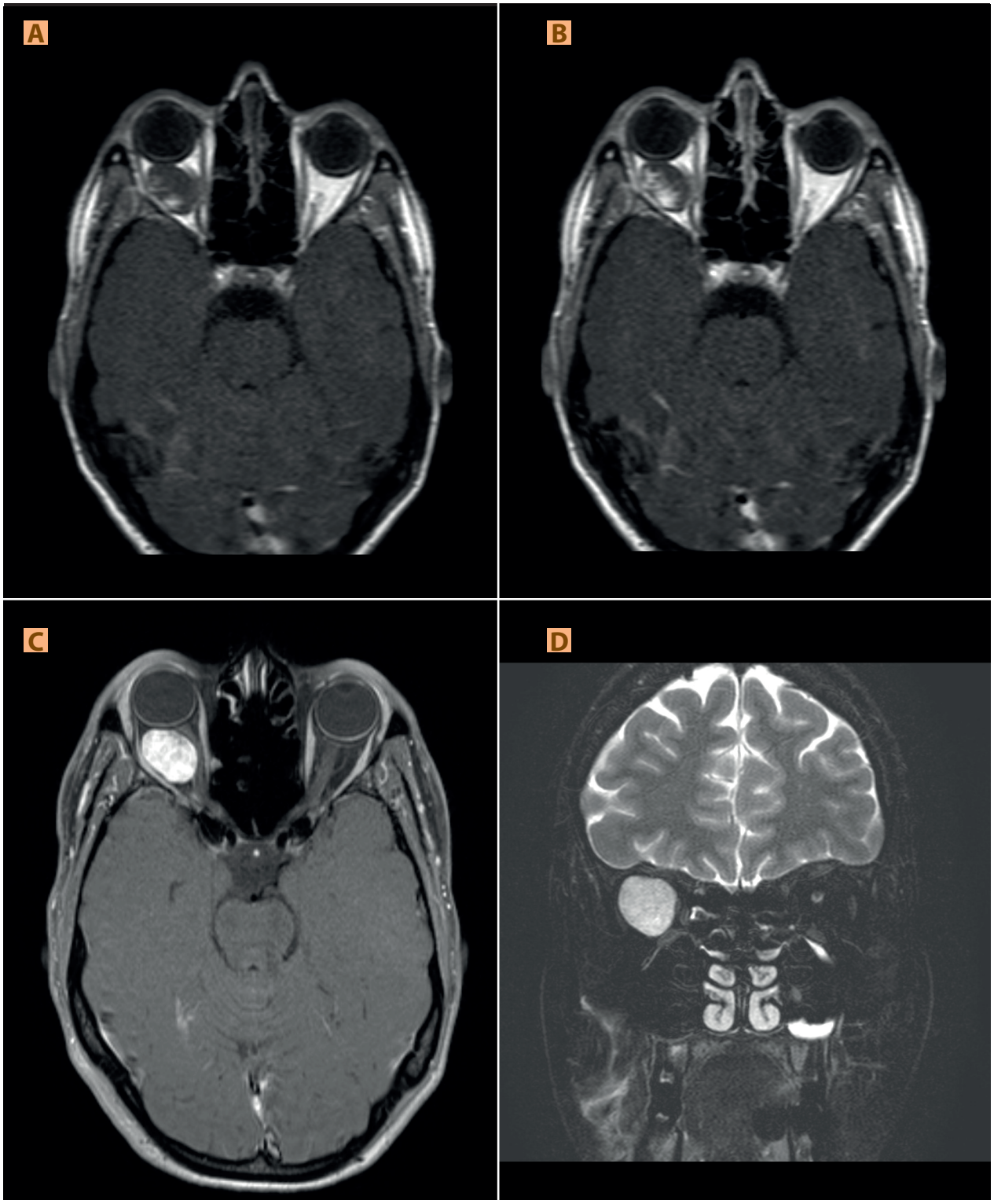

Fig. 6. Cavernous haemangioma: (A, B) progressive saturation in dynamic study, (C) saturated tumour in T1STIR, (D) hypersignal well bordered intraconal formation in T2STIR

extracellular methemoglobin appears, which is hypersignal in both T1WI and T2WI. Old blood breakdown products such as haemosiderin or ferritin are hyposignal in both T1WI and T2WI. Upon the absence of trauma in the medical history, the most common cause of orbital haematoma is vascular malformation or orbital varix, demonstration of which is more arduous in the acute phase of haemorrhage (haemorrhage may cause destruction of the malformation). An extended and coiled vena ophthalmica superior may imitate a tumour. Varix of the upper ophthalmic vein is a tubular formation, hyposignal in T1WI and T2WI (outage of signal on a background of flow-void phenomenon). If thrombosis of the varix occurs, the signal is altered according to the breakdown stages of haemoglobin. Another cause of intraorbital haemorrhage may be haemorrhage into a true tumor.

The lachrymal gland may be affected by a tumorous pro-

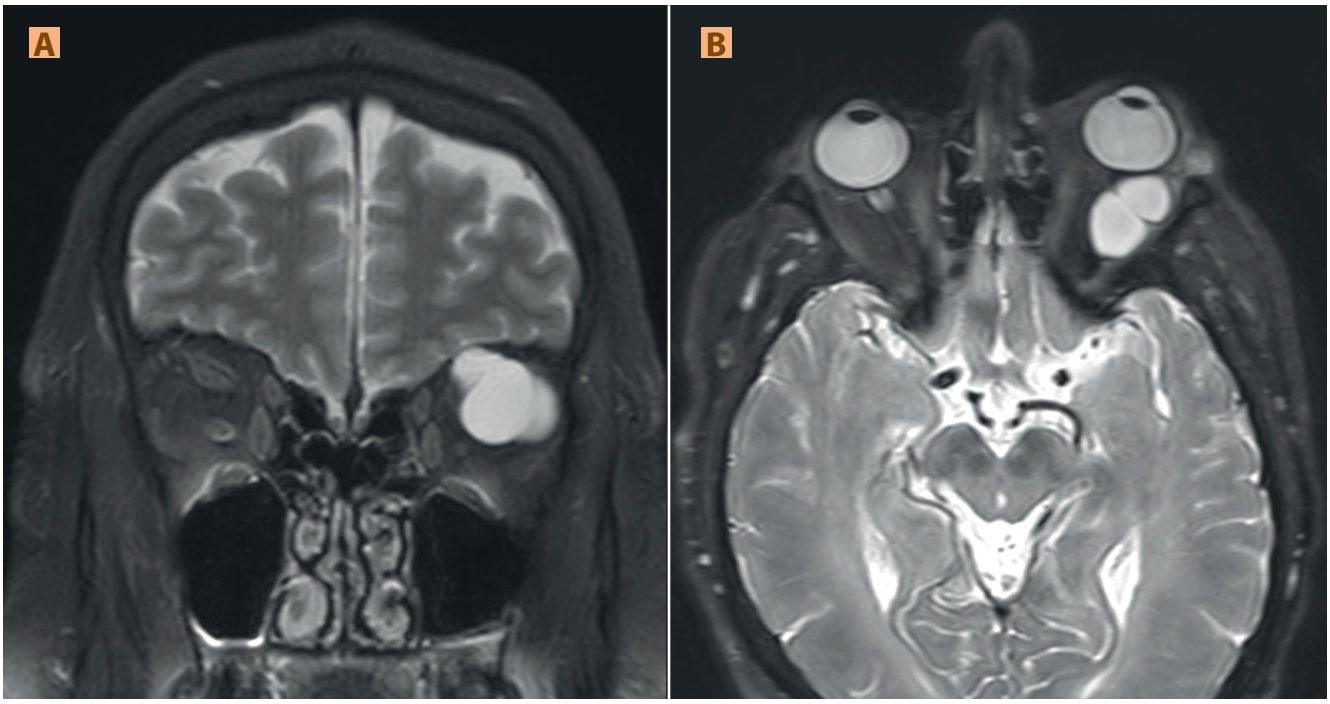

Fig. 7. Lymphangioma: (A) bordered multilocular intraconal formation in left orbit hypersignal in T2WI and (B) T2STIR

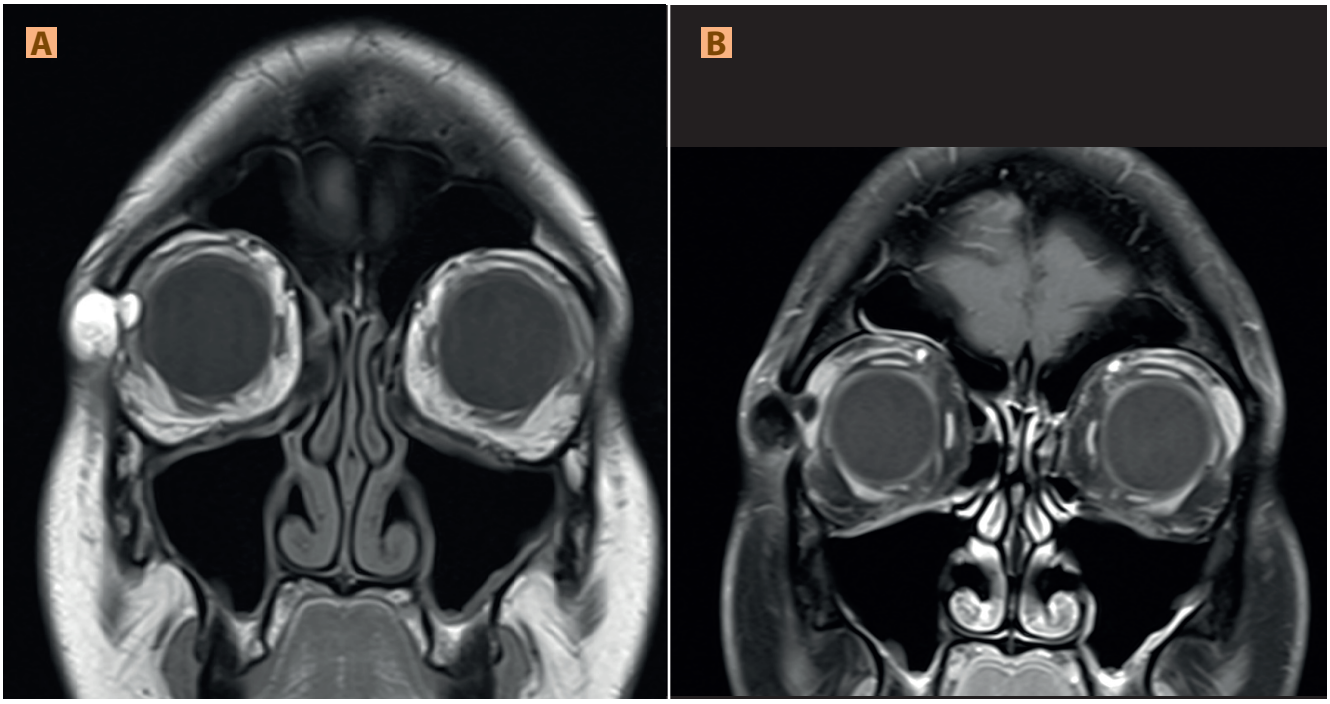

Fig. 8. Dermoid cyst: (A) bordered septated formation ensuing from frontozygomatic suture, larger portion subcutaneously, smaller infringing intraorbitally, hypersignal in T1WI, (B) hyposignal T1FS (lipid-rich content)

cess, its most common primary tumour is a pleomorphic adenoma. In MRI imaging it is well bordered, rounded or oval, in T1WI iso to hyposignal in relation to the extraocular muscles, in T2WI iso to hypersignal, with post-contrast saturation, if it is of a larger size it is generally heterogeneous, with necroses or cystic degeneration [9]. The most common malignant tumours of the lachrymal gland include adenoid cystic carcinoma and adenocarcinoma. In MRI image usually evident as an infiltrative mass affecting the lachrymal gland, in T1WI iso to hyposignal in relation to the extraocular muscles, in T2WI non-homogeneously hypersignal, with post-contrast saturation, and unlike benign tumours it may be accompanied by destruction of bone. In differential diagnostics we always consider also other clinical units (inflammatory pseudotumour, lymphoma), see above.

Dermoid cyst (Fig. 8) is displayed mostly as a smoothly bordered extraconal, post-contrast non-saturating mass, containing cystic and/or solid components. In children it presents itself shortly after birth as resistance in the hypodermis. The signal characteristic depends on the representation of the liquid component (hypersignal in T2WI, hyposignal in T1WI), and on the portion of the liquid content (hypersignal in both T1WI and T2WI with suppression of fat signal in SPIR or FATSAT sequences).

We always indicate laboratory examination according to the envisaged cause of the pathology, which applies also in the diagnosis of orbital tumours. We extend the basic laboratory examination (biochemical and blood count) by examination of the parameters of thyroid function, including antibodies upon suspicion of EO. We also exclude the most acute haematological pathologies, and thereby accelerate the diagnostic process.

TREATMENT

The further procedure depends on the conclusion of the performed tests, but above all on the result of the imaging examinations. It is necessary to send an acutely ongoing orbital pathology immediately to a centre which is capable of providing inter-disciplinary care, e.g. orbital cellulitis requires an immediate procedure or permanent monitoring of the condition. The largest proportion of patients from the group with observed exophthalmos are designated for conservative therapy (EO, PST, infantile haemangiomas, gliomas etc.) [10,11]. In the case of bordered lesions of the orbit, we decide on the performance of an early or deferred operation with regard to the patient's subjective complaints, expected further development of the pathology and the risk ensuing from leaving the lesion, or from potential iatrogenic damage. A part of these tumours are indicated for radiotherapy [12]. In the case of non-bordered orbital lesions, we always attempt to obtain a sample for histological verification [13], which is essential for the choice of subsequent therapy, especially in the case of suspicion of ML. We proceed to anterior orbitotomy in the case of lesions localised in the entrance to the orbit, which are mostly palpable. We indicate total extirpation for cysts and unilateral lesions of the lachrymal gland, in which there is safer

Table 2. Incidence of operated tumours in individual age categories of patients (at Departments of Stomatology and Ophthalmology, General University Hospital in the period of 2005-2019)

Age of patient at time of operation		Number of patients of relevant age (total 497):	Proportion of age category in total number of patients:
decade	years		
1st decade	0–10	8	1,5%
2nd decade	11–20	20	4%
3rd decade	21–30	24	5%
4th decade	31–40	65	13%
5th decade	41–50	71	14%
6th decade	51–60	84	17%
7th decade	61–70	111	22,5%
8th decade	70–80	80	16%
9th decade	81 a více	34	7%

Table 1. Numbers of patients operated on for orbital tumour according to gender representation (at Departments of Stomatology and Ophthalmology, General University Hospital, in the period of 2005-2019) and age

Sex:	Centre where tumour operation was performed:	Number of patients:	Age of patients – median:	Age of patients – mean:
men	Department of stomatology	142	52	51
	Department of ophthalmology	107	63	58
	Total stomatology and ophthalmology	**249**	59	55
women	Department of stomatology	140	54	54
	Department of ophthalmology	108	63	58
	Total stomatology and ophthalmology	**248**	59	56
Men and women in total	Department of stomatology	282	53	52
	Department of ophthalmology	215	63	58
	Total stomatology and ophthalmology	**497**	59	56

Table 3. Representation of operated orbital tumours according to their biological nature (at Departments of Stomatology and Ophthalmology, General University Hospital in the period of 2005-2019)

Biological nature of tumours (result of histological examination):	Number of tumours	Proportion of tumours in entire cohort:
Benign tumours:	278	55,9%
Of which "actual tumours"	111	22,3%
Of which "false tumours" and other similar formations	167	33,6%
Malignant tumours:	207	41,7%
Number of procedures in which result of histology was absent or unconvincing:	12	2,4%
Total number of performed procedures at departments of stomatology and ophthalmology:	497	100%

Table 4. Representation of more frequently operated tumours (at Departments of Stomatology and Ophthalmology, General University Hospital in the period of 2005-2019), out of a total number of 67 different histological diagnoses only units in a minimum quantity of 9 are stated (i.e. min. approx. 1.8 %); numbers include recurrences or necessary reoperations

Biological nature of tumours:		Order of representation within total:	Type of tumour:	Number of tumours:	Proportion of tumours in entire cohort:
Actual benign tumours		1	haemangioma	54	10,9%
			Most often cavernous haemangioma	45	
		2	Pleomorphic adenoma of lachrymal gland	17	3,4%
		3	meningeoma	15	3%
		4	Tumours of peripheral nervous system	13	2,6%
			Most often Schwann	12	
False benign tumours		1	Dermoid formations	38	7,6%
			Most often dermoid cysts	36	
		2	Orbital pseudotumour	25	5%
		3	Prolapse of orbital fat	20	4%
		4	Vascular malformations and varices	10	2%
Malignant tumours	**Primary:**	1	Malignant lymphoma	87	17,5%
		2	Malignant tumour of lachrymal gland	13	2,8%
			Most often adenocarcinoma	9	
		3	Extrapleural solitary fibrous tumour	11	2,2%
	Secondary:	1	Basal cell carcinoma	39	7,8%
		2	Spinocellular carcinoma	15	3%
		3	Malignant melanoma	10	2%
	Metastases:	1	total	9	1,8%
			Most often of breasts and liver (each 3x)	3	

access under direct visual control. Upon breach of the capsule of a malignant tumour, the risk of occurrence of metastases increases. There are several possible surgical procedures into the orbit, and the choice thereof depends on several factors, but primarily on the experience and composition of the operating team [14]. Co-operation of ophthalmological and stomatological departments in orbital surgery has been taking place now for almost 50 years. We are capable of extirpating orbital tumours by means of lateral osteoplastic orbitotomy, with the exception of localisation in the orbital apex [15]. We resolve tumours embedded intraconally medially from the optic using the same procedure, but we extirpate the actual tumour from a transconjunctive approach, with temporary tomy of the tendon of the musculus rectus medialis and its subsequent suturing to the original place of the tendon. A medial extraconal space thus remains for an endoscopic transnasal procedure, beyond the orbital apex [16,17]. The orbital apex is relatively most accessible from above, i.e. from a transcranial extradural approach, which is within the jurisdiction of a neurosurgeon, who often also performs decompression of the orbit or optical channel (meningeoma). It is necessary to select individual approaches also upon affliction of the orbital skeleton (diploid dermoid) and unusual findings [18,19]. It is possible to perform anterior orbitotomy in order to take

Table 5. Most frequently operated tumours (at Departments of Stomatology and Ophthalmology, General University Hospital in the period of 2005-2019) according to centre at which the procedure was performed

Centre:	Most common actual benign tumour:		Most common false benign tumour:		Most common malignant tumour:		Most common tumour regardless of biological nature:	
Department of Stomatology	Cavernous haemangioma		Dermoid cyst		Basalioma		Cavernous haemangioma	
Department of Ophthalmology	Capillary haemangioma		Prolapse of orbital fat		Malignant lymphoma		Malignant lymphoma	
Stomatology and Ophthalmology total:	Cavernous haeman--gioma	45 (9% z 497)	Dermoid cyst	36 (7% z 497)	Malignant lymphoma	87 (17,5% z 497)	Malignant lymphoma	87 (17,5% z 497)

Table 6. Surgical approaches selected in operations on orbital tumours (at Departments of Stomatology and Ophthalmology, General University Hospital in the period of 2005-2019)

Centre:	Selected surgical procedure:	Number of procedures:				Ratio:
Department of Stomatology	Lateral orbitotomy (most common)				**137**	49 % of procedures at Dept. of Stomatology
	Anterior orbitotomy				**82**	29 % of procedures at Dept. of Stomatology
	exenteration				**48**	17 % of procedures at Dept. of Stomatology
	Less frequent (temp. transmuscular, bicoronary, transvestibular...)				**15**	5 % of procedures at Dept. of Stomatology
	celkem:				**282**	
Department of Ophthalmology	Anterior orbitotmy	Transcutaneous (most common)	149	69%	**200**	93 % of procedures at Dept. of Ophthalmology
		transconjunctival	49	23%		
		Combination of above	2	1%		
	Puncture biopsy		15	7%	**15**	7 % of procedures at Dept. of Ophthalmology
	Total:				**215**	
Stomatology and Ophthalmology total	Anterior orbitotmy (most common)				**282**	56 % of all procedures (stomatology and ophthalmology)
	Lateral orbitotomy				**137**	28 % of all procedures (stomatology and ophthalmology)
	Other				**78**	16 % of all procedures (stomatology and ophthalmology)
	Total (all procedures):				**497**	

Table 7. Exenterations of orbit (all realised exclusively at the Department of Stomatology, General University Hospital in the period of 2005-2019)

Tumour for which exenteration indicated:	Number of exenterations:	Proportion of tumours in number of all exenterations:
Basal cell carcinoma (most common)	**17**	35%
Malignant melanoma	**9**	19%
Spinocellular carcinoma	**5**	10%
Meningeoma	**4**	8,5%
Adenocarcinoma of lachrymal gland	**4**	8,5%
Other tumours (represented by only 1-2 patients each)	9	19%
celkem	**48** (i.e. 17 % of procedures at Dept. of Stomatology and 10 % of all procedures in entire cohort)	

a sample in most cases under local anaesthesia, we choose general anaesthesia for children, anxious patients and upon removal of larger and/or adversely localised lesions (deeply embedded lesions, nasal upper quadrant). We consider indication for biopsy by puncture needle in the case of more deeply embedded tumours and in patients who are unable to undergo general anaesthesia. However, due to the risk of iatrogenic damage to orbital structures, the indication criteria of this method are limited. In the case of a thin-needle biopsy the probability of injury is smaller, but its yield is also lower. As a rule we indicate exenteration of the orbit, sometimes extended, in the case of malignant tumours, often secondary. Upon exenteration for meningeoma of the optic sheaths it is possible to preserve the conjunctival sac and eyelid.

Own cohort

We processed data from the period of 2005-2019, i.e. for 15 years. The cohort consisted of patients in whom an orbital lesion was identified by clinical or imaging examination, in which there was suspicion of a tumorous process, and for this reason the patients underwent surgery at the Department of Ophthalmology or Stomatology at the 1st Faculty of Medicine of Charles University and the General University Hospital in Prague. The distribution according to sex was entirely symmetrical, the median age was higher in patients operated on at the Department of Ophthalmology (Table 1), the frequency of operations in the individual decades of age is illustrated in Table 2. Due to a positive finding in the orbit with an absence of functional or subjective complaints, some of our patients were merely observed for a period of months or up to several years. The cohort does not include patients with a demonstrated orbital lesion who were transferred into the care of radio oncologists for radiation therapy (meningeomas of the optic sheath, metastases), to neurosurgeons (pterional or canalicular meningeomas, haemangiomas of orbital apex) or to ENT specialists (nasal and inferonasal extraconal tumours of the orbit). We obtained complete data on 497 patients, in the case of 19 patients it was not possible to obtain full data. With only a few exceptions (extrapleural fibrous tumour, carcinoma of the lachrymal gland and skin tumour growing into orbit), only one operation was performed on the patients. At the Department of Stomatology a total of 282 procedures were performed under general anaesthesia, in the overwhelming majority of cases with clear or possible need for an osteoplastic approach to the orbit. At the Department of Ophthalmology a total of 215 procedures were performed, more often under local anaesthesia. There were more benign lesions than malignant (Table 3), the representation of the most common lesions in individual groups is illustrated in Table 4. Table 5 presents an overview of the most common lesions operated on at both centres. The selected surgical procedures are illustrated in Table 6. All exenterations of the orbit were performed at the Department of Stomatology, most frequently due to carcinomas of the eyelids growing into the orbit (Table 7).

DISCUSSION

The frequency and absolute numbers of orbital pathologies differ according to the focus and jurisdiction of the specific centre. According to a large retrospective study from the orbital centre of the Department of Ophthalmology in Vancouver, 2985 patients were referred during the period of 1976-1993, of whom 50 % had EO, 20 % tumours, 14 % structural lesions, and 9 % inflammations [1]. In contrast with this, Schields et al. in a cohort of 1264 patients referred over more than 30 years to an orbital oncological centre, stated only 5 % of patients with EO, while the most common primary tumours were vasculogenic lesions (17 %), lymphoid tumours (11 %), inflammatory pseudotumours (11 %), tumours from the meninges and optic nerve (8 %), and cystic lesions (6 %). Secondary tumours were in 11 % of cases and metastatic 7 % [20]. Bonavolontà et al. excluded EO, PST and ocular malformations from the cohort. In their cohort at an orbital centre they evaluate a total of 2480 orbital lesions observed over the course of 35 years. Of these 88 % were primary orbital tumours (24 % vasculogenic, of which 9 % were cavernous haemangiomas, 21 % cystic, of which 14 % dermoid cysts, 13 % malignant lymphomas), 9 % secondary and 3 % metastases [21]. The Japanese authors Ohtsuka et al. present a cohort of 244 patients over the course of 21 years with 89 % primary tumours (24.1 % malignant lymphoma, 18.4 % lymphoid hyperplasia, 8.6 % adenoma of the lachrymal gland, 7.4 % cavernoma, 5.3 % dermoid cyst), 9 % were secondary and 2 % were metastases [22]. In our co-

hort we had 78.5 % primary tumours (17.5 % malignant lymphoma, 11 % vasculogenic tumours, 7.6 % cysts, 6 % tumours of the lachrymal gland), secondary 20.7 % and metastases 1.8 %. The distribution of incidence of primary tumours is closest to the Japanese cohort, evidently also because the majority of patients underwent surgery. The higher percentage of secondary tumours in our cohort is due to the higher proportion of basaliomas and spinaliomas, which formed 10.8 %. These were also the most frequent indication for exenteration. It is interesting to compare the 15-year co-operation of both of our centres during the period of 1970-1985, when 135 operations were performed for orbital tumours, in which the most common were malignant lymphomas (15.5 %), tumours of the lachrymal gland and vasculogenic tumours were equally represented with 10.8 %, inflammatory pseudotumours 8.9 %, and cystic lesions 8.1 %. Exenteration was indicated in 10 % of cases for basaliomas, spinaliomas and sarcomas [23]. It appears that tumours of the lachrmyal gland decreased. So far the trend of increasing incidence of ML within the population has not been manifested in our cohort.

Data on the frequency of tumours from the individual centres differs in terms of the percentage representation of the individual pathologies, but benign lesions are 2/3 and malignant 1/3. According to individual publications 64:36 % [20], 68:32 % [21], in our cohort 55.9:41.6 % (2.4 % of results were unclear). The high percentage of tumours of the eyelids penetrating into the orbit, sometimes following insufficient resections of the primary tumour, sometimes due to negligence on the part of the patient, is a factor requiring consideration.

In our cohort there was equal representation of men and women, in the other cohorts a slight predominance of women (53.6-57 %:46.4-43 %). The cohorts cover all age categories (0-95 years), with a different frequency of tumours in the individual decades of life. In the first decade dermoid cysts predominate, after the age of 30 years cavernous haemangiomas and tumours of the lachrymal gland appear. Malignant lymphomas and skin tumours begin after the age of 50 years [14]. The distribution in our cohort does not differ from other studies, with a peak in the 7th decade (22.5 %). However, we recorded a higher mean age of the entire cohort (56 years in comparison with 43 [21] and 48.7 [22] respectively), and an even higher median age of 59 years. The mean and median age (51 and 52 years respectively) is lower at the time of performance of the procedure in patients at the Department of Stomatology, thus those who are operated on under general anaesthesia. In the case of older patients there is a predominance of procedures under local anaesthesia for the taking of samples, most often due to suspicion of ML.

CONCLUSION

The presented cohort of operated orbital tumours is comparable with the cohorts of other authors, with a dominant representation of malignant lymphomas. An alarming factor is the constantly high number of skin tumours growing into the orbit. The presented examination procedure is the standard, improving imaging methods help facilitate the diagnostic-therapeutic balance. The surgical approaches into the orbit we used proved to be effective, although in certain cases it is necessary to select approaches individually. In diagnosis and treatment, constant inter-disciplinary co-operation is essential.

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