

# Lymphangioma of the Orbitopalpebral Area

J. Krásný<sup>1</sup>, D. Baráková<sup>1</sup>, Z. Chodounský<sup>2</sup>, J. Šach<sup>3</sup>

<sup>1</sup>Department of Ophthalmology, Královské Vinohrady University Hospital, Prague

Head: prof. MUDr. P. Kuchynka, CSc.

<sup>2</sup>Department of Radiotherapy, Královské Vinohrady University Hospital, Prague

Head: prim. MUDr. Z. Chodounský, CSc. (+)

<sup>3</sup>Institute of Pathology, 3rd Faculty of Medicine, Královské Vinohrady University Hospital, Prague

Head: prof. MUDr. V. Mandys, CSc.

## SUMMARY

**Aim:** The authors refer about five patients with different types of lymphangioma, who were followed up at the Department of Ophthalmology, Faculty Hospital Královské Vinohrady (King's Vineyards), Charles University, Prague, Czech Republic, E.U., during the period 1995 – 2013; the follow-up period lasted from 5 to 17 years. The lymphangioma of the orbitopalpebral area is discussed according to the evaluation of the tumor development, histological verification, treatment, and its results.

**Methods:** In four boys, the first signs of tumor were eyeball protrusion (exophthalmos) and bleeding into the conjunctiva or palpebral skin before the age of 5 years. In all four patients, the histological confirmation of the orbital lymphangioma was performed in the beginning of the disease. In three cases, it was the orbital type, and the fourth one was frontal type with bilateral orbital lymphangiomatosis. In one girl, there were present conjunctival changes only, appearing as one-sided hyperplastic changes. For these changes, she was followed-up since her 13 years of age under the false diagnosis of chronic conjunctivitis. The definite histological confirmation of only conjunctival lymphangioma was done from the diagnostic probatory biopsy not until ten years of symptoms and unsatisfactory treatment.

**Results:** In the girl with superficial conjunctival lymphangioma and in the patient with lymphangiomatosis, the follow-up was recommended only. In two patients with extraconal type of orbital tumor, the total or sub-total resection was performed. In the years of the follow-up, the remission of the disease was observed. In the patient with mostly intraconal type of the tumor, causing decrease of the visual acuity according to the optic nerve neuropathy and macular cystoid edema, the focused actinotherapy by means of linear accelerator treatment with the dose of 30 Gy after previous evacuation of chocolate cysts under ultrasound control. The regression of the tumor and normalized visual functions lasted for 17 years.

**Conclusion:** As method of treatment of extraconal lymphangiomatosis, it seems, it is its resection, and in the intraconal localization of the tumor it is the focused actinotherapy by means of linear accelerator.

**Key words:** orbital lymphangioma, conjunctival lymphangioma, lymphangiomatosis, tumor resection, linear accelerator actinotherapy

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## INTRODUCTION

Lymphangioma occurs on a congenital basis and together with haemangioma is ranked with hamartomas. Their incidence is bound to tissues which are normally present within the orbit, which does not apply in general for lymphatic blood vessels. Due to its cellular structure lymphangioma ranks amongst benign tumours, but from a clinical perspective it does not merit this indication. It does not have a capsule, it grows diffusely and infiltrates the surrounding tissues. Microscopically lymphangioma is formed by a network of lymphatic areas covered with the endothelium and thin stroma with a varying degree of lymphocyte in-

filtration, within the framework of which lymphoid follicles may be formed. The local behaviour in the orbit is thus similar to that of malignant tumours, but unlike these it does not metastasise (39). Three fundamental clinical forms are distinguished. Superficial lymphangioma afflicts only the conjunctiva, manifested as cystoid edema, after which there is the separate orbital type which afflicts the orbit postseptally. The combined form is manifested pre and postseptally in the oral cavity and in the cheek (13). It grows slowly and persistently without a tendency towards spontaneous involution, thankfully in adulthood growth usually stabilises. It is generally unilateral and presents both cosmetic problems and functional disorders such as strabismus

and amblyopia on the same side. A basic symptom includes asymmetry of the face and eye apertures, namely fuller cheeks and bulging of the eyelids on the afflicted side. The conjunctiva is afflicted in any part initially in the form of vitreous chemosis, which passes into a cystic character with lymphangiectasia. The clinical picture is completed with enlargement of the bone part of the eye socket. Protrusion and dislocation of the bulb is variable depending on sudden massive haemorrhage into the tumour, which may represent a severe complication endangering the eyeball itself. It is manifested on the conjunctiva through repeated suffusions. Vision is then endangered by the protraction of the optics or compression of its nutritive blood vessels (39).

**First author:**

**MUDr. Jan Krásný**

Oční klinika FN Královské Vinohrady  
Šrobárova 50  
10 034 Praha 10  
jan.krasny@fnkv.cz

## OWN OBSERVATION

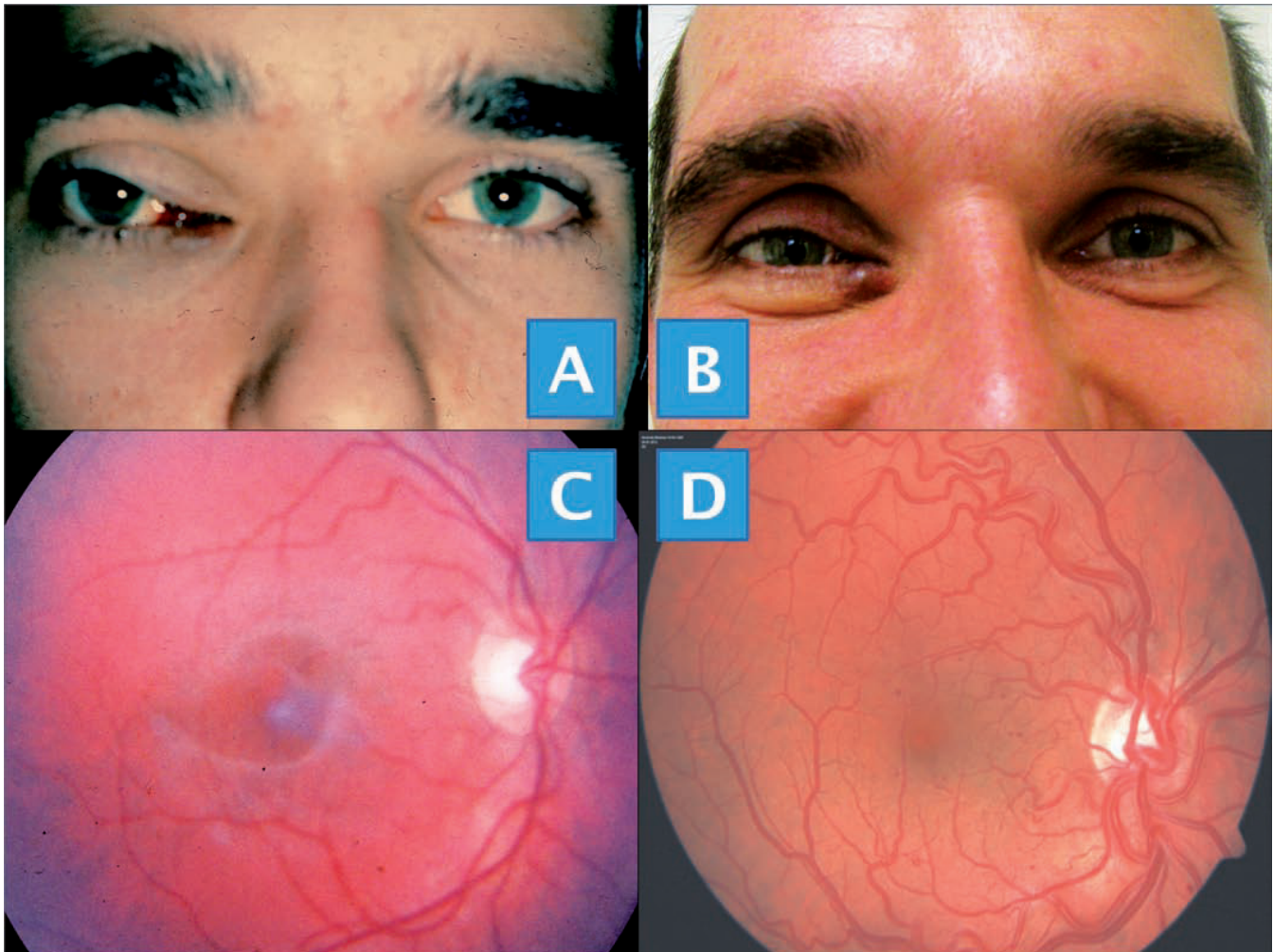
Lymphangioma was not included in the previous ten-year study (28) on pathologies of the orbit in adulthood at the Department of Ophthalmology at the Královské Vinohrady University Hospital in the years 1998-2007, since the occurrence of this disorder in all the five patients stated at the time was in childhood. As a result, we decided to recall the issue of lymphangioma from a clinical, histological and therapeutic perspective within the framework of long-term observation from 1996-2013, i.e. over the course of 17 years.

### Patient no. 1

The first observation of our cohort was the only patient to be observed, even if irregularly, throughout the entire period of our study. In 2013 the patient was aged 32 years. The beginning of

his symptoms date from the age of five (1985), when there was protrusion of the eyeball on the right side, accompanied by haemorrhage into the conjunctiva. A complex examination was conducted at a regional centre outside of Prague. Probatory excision was performed, which detected a diagnosis of lymphangioma. After three years there followed actinotherapy of the entire orbital cavity in a dose of 16.7 Gy/focus. The condition did not progress further for several years, but haemorrhage into the conjunctiva was repeated. During the course of 1995 progressive protrusion again gradually appeared. In September 1995 CT demonstrated an irregular formation with a diameter of approximately 17 mm with a density of 50 HU. intraorbitally nasally, which was only slightly pronounced postcontrast. The retrobulbar space in the direction of the apex of the orbit was also filled with non-homogeneous masses of the same density as the formation nasa-

lly. The condition continued to progress, and as a result the patient was admitted to the Department of Ophthalmology of the KVUH in September 1996 for a decision on the further procedure. The right eyeball was in 5 mm protrusion and deviated outwardly by 10 mm, with limited motility in adduction. A perfused tumour, which was reductible in the direction towards the orbit was evaginated beneath the conjunctiva nasally in the internal corner (fig. 1a). The other finding on the anterior segment was physiological. Cystoid macular edema (CME) was perceptible on the ocular fundus, as well as slightly constricted blood vessels, and the papilla of the optic nerve was bordered (fig. 1c). VOR (= visus of the right eye) 0.33 with + 1.0, J. no. 6 best correction. (In left eye extraocular and intraocular finding was physiological, VOL (visus of the left eye) 1.0 nat., J. no. 1 nat.). A transocular B scan of the eye socket showed a tumour of a cystic character

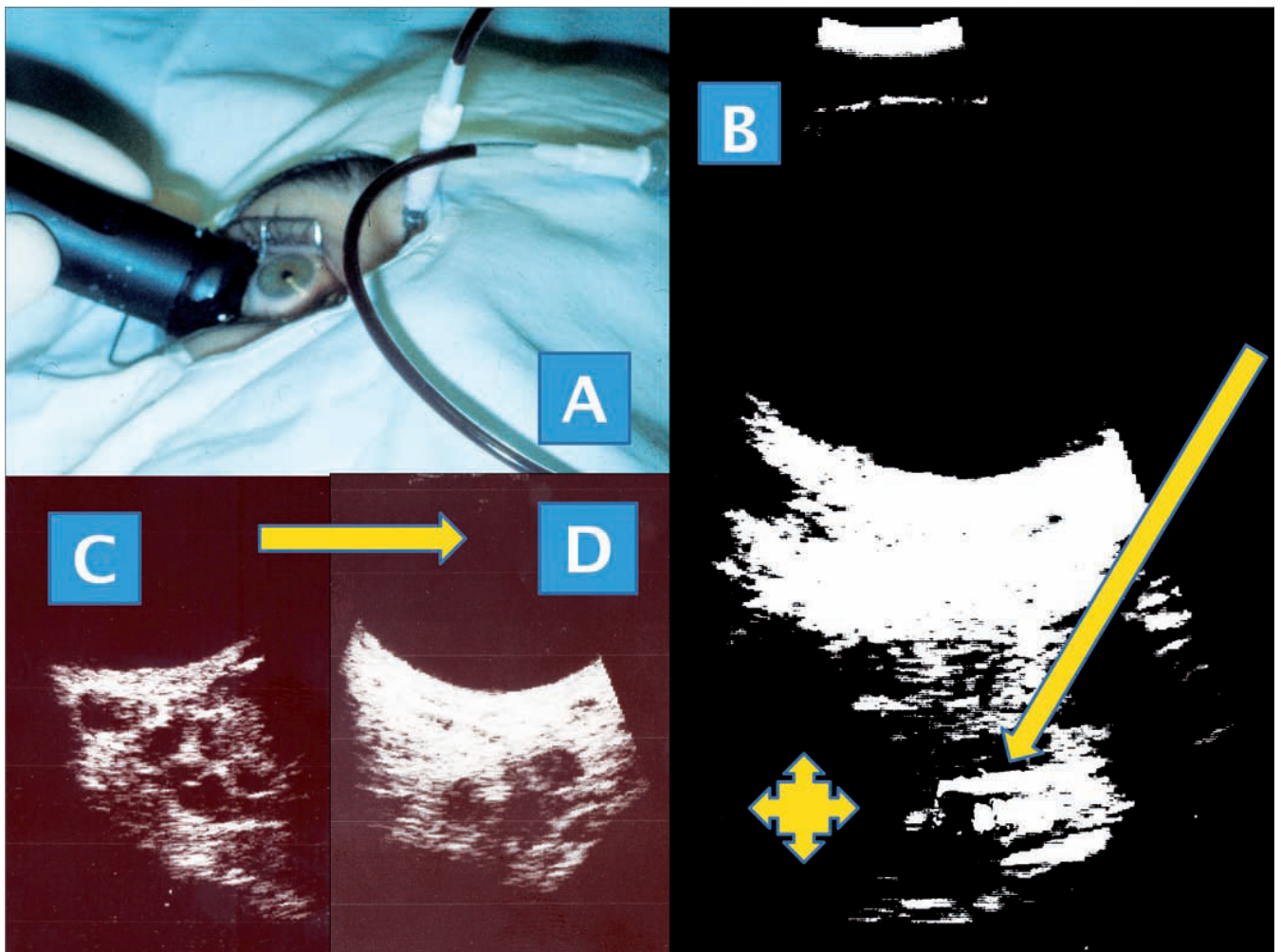


**Fig. 1** Patient no. 1  
a – clinical picture before aspiration of chocolate cysts and actinotherapy.  
b – clinical picture following treatment after 17 years.  
c – finding on fundus of right eye with CME before treatment.  
d – finding on fundus of right eye after 17 years.

with echographic features typical of lymphangioma: irregular internal structure, high reflectivity of the septa and low reflectivity of the interseptal space filled by the stroma. One of these larger cysts manifested higher internal reflectivity in comparison with the others. This concerned a "chocolate cyst", which was filled with what was probably colliquated blood (fig. 2c). After consultation with the radiotherapists we decided upon repeat actinotherapy. Due to its higher efficacy, it was first of all necessary to drain the content of the cyst, with the aim of relieving the pressure on the optic nerve. We performed partial aspiration of the content of the cyst under general anaesthesia under the control of ultrasound (fig. 2a, b). The subsequent regression of the protrusion and outward deviation by several mm was accompanied also by a change of the image on the transocular B scan, where a reduction of the diameter of the cyst was visible (fig. 2d). Sub-

sequent actinotherapy was performed in April 1996 in fifteen sessions of 2 Gy/lfocus (total dose 30 Gy/focus). A linear accelerator was used, and the planning scheme covered the retrobulbar space up to the apex of the eye socket in the form of a "bowl base". Three months after the end of irradiation treatment the protrusion had been reduced to 1 mm and the outward deviation had decreased to a mere 3 mm. Motility of the bulb was now entirely free. The finding on the anterior segment of the eye was physiological, on the ocular fundus the finding in the area of the macular region had been normalised, VOR 0.66 nat., J. no. 2. Three years after actinotherapy of the right orbit, normophthalmos was demonstrated with only a suggestion of outward deviation. The finding on the anterior segment was physiological up to the incipient manifestation of a post-radiation cataract in the posterior cortex in the form of a disciform 2-3 mm cataract. The

finding on the ocular fundus was physiological. VOR 0.66 with +2.5/80 and J. no. 2 obt. with +2.5/80. The patient was recommended regular eye checks in his place of residence, which he however did not undergo, upon his own decision. He did not report to our clinic again until January 2013, upon the request of a doctor from a psychiatric institution, where the patient had been monitored due to a schizophrenic disorder. During his residence at the institution, repeated haemorrhage into the patient's conjunctiva occurred repeatedly in the internal corner on the afflicted side. Upon an examination at our clinic, the configuration and position of the bulb in the right eye were entirely physiological, motility free and spatial vision was also within the norm (stereoscope, Randot). Suffusion of the tumescent conjunctiva was perceptible in the internal corner (fig. 1b), in a further finding on the anterior segment only irregular opacities were perceptible in the



**Fig. 2** Patient no. 1

- a – aspiration of chocolate cysts under ultrasound control.
- b – ultrasound image of aspiration: needle with acoustic shadow by chocolate cyst (arrow).
- c – orbital cavity with chocolate cysts.
- d – regression of finding after aspiration.

lens beneath the posterior capsule. On the fundus the papilla of the optic nerve was overall paler than on the left side, the macular region now had physiological contours, the blood vessels were of a normal calibre and slightly more coiled (fig. 1 d). VOR 0.8 – 0.9 with +0.5/50 and J. no. 2 with + 0.5/50. Contrast sensitivity was on the borderline of the norm at 3, 6 and 12 c/deg, and only slightly diminished at 18 c/deg (CSV 1000). This change of reduction was confirmed by normal contrast sensitivity in the left eye, where the finding was entirely physiological VOL 1.0 nat., J. no. 1.

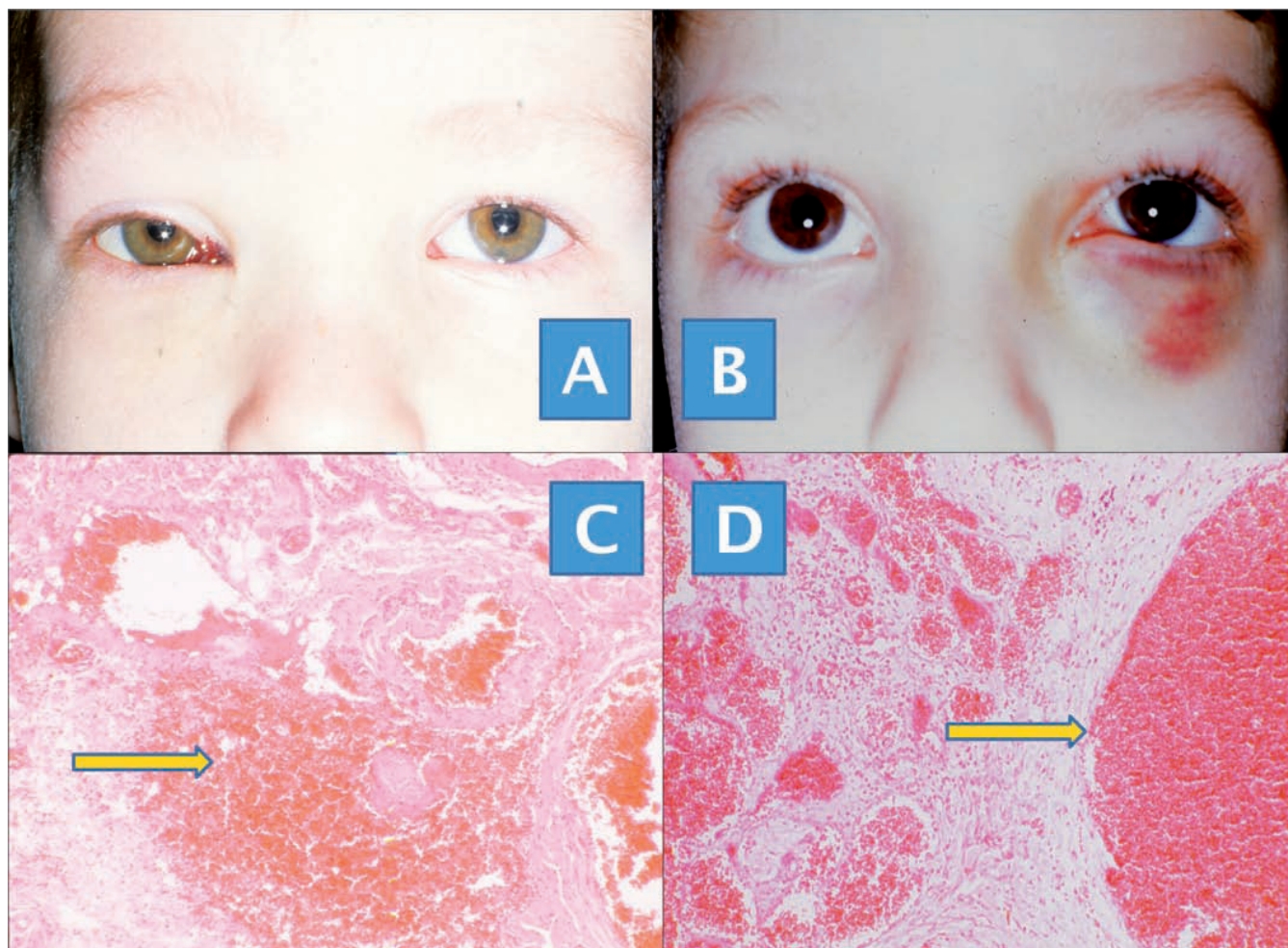
Overall the condition following actinotherapy was evaluated as highly satisfactory. The finding on the lens following irradiation did not demonstrate progression, since the initial changes had receded, together with practical normalisation of refraction. The slight atrophication of the optic nerve could be the result of a side effect of irradiation, but also an echo of

the nutritional disorder of the head of the optic nerve due to a suppression of the nutritive blood vessels by the tumour itself. Perfusion in the internal corner sketched in an image of possible residue of the tumour, since this area had been only minimally irradiated, and this had been within the framework of the first actinotherapy in 1988. The subjective complaints of dryness of the right eye improving following the application of lubrication were linked to KCS following irradiation.

#### Patients no. 2 and 3

The next two patients were four year old boys in whom there was identical haemorrhage in the conjunctival orbital region due to medical care, without anamnesis of trauma. The complete laboratory examination in both boys, including sedimentation of erythrocytes, was negative. In patient no. 2 the parents initially noticed haemorrhage beneath the con-

junctiva in the internal corner in the right eye, which gradually transformed into an enlarging formation which displaced the eye outwardly. At the first examination at our centre in 1998, there was protrusion of the right eyeball of 2 mm with outward deviation by 5 mm (fig. 3a) after a three week anamnesis. Motility of the bulb was free without diplopia. The further finding on the anterior segment of the eye and the intraocular finding was physiological bilaterally, VOR = VOL 1.0 nat. An ultrasound examination demonstrated a pre-equatorially, irregularly bordered, lobed formation 6 x 5 x 6 mm in the internal part of the orbit, localised retroseptally. With regard to the character of the clinical picture and the speed of onset of the symptoms, we proceeded to a probatory excision for the purpose of histological verification of the nature of the tumour. Due to the minimal haemorrhaging during the course of the operation we performed partial resection of the formation.



**Fig. 3** Patient no. 2 in right eye and patient 3 in left eye  
a – clinical picture of lymphangioma in right eye.  
b – clinical picture of lymphangioma in left eye.  
c – histological verification of right-sided lymphangioma of orbit (blood staining of tumour – arrow).  
d - histological verification of left-sided lymphangioma of orbit (haemorrhagic dilated cavity, future chocolate cyst – arrow).

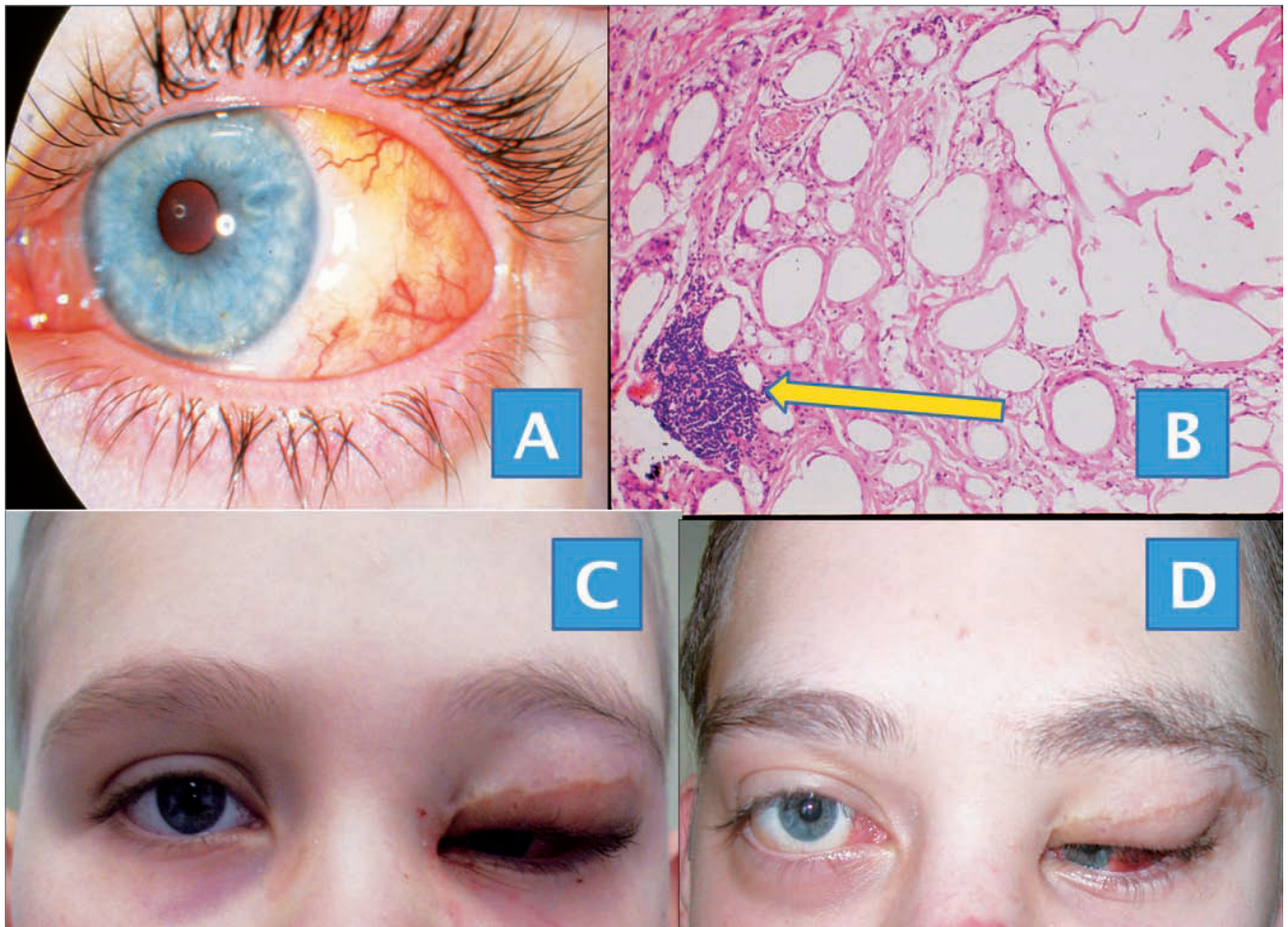
In addition to the haemorrhagic structures of lymphangioma, scarry regression and fibrous conversions of certain parts of the tumour with its microscopic residues were determined in the histological image (fig. 3c). After six months the configuration and position of the right eyeball had been normalised, and no relapse of the tumour was manifested over the further course of several year observation. The reason for hospitalisation of the third patient in our group, also in 1998 was haematoma persisting for ten days in the area of the lower eyelid of the left eye, with dislocation of the bulb upwards and outwards by 2 mm (fig. 3b). Its motility was free, without diplopia. The further finding on the anterior segment of the eye and the intraocular finding was physiological bilaterally, VOR = VOL 1.0 nat. Ultrasound examination in the internal part of the orbit demonstrated a relatively well bordered oval formation of an irregular internal structure and size of 5 x 4 x 3 mm lying retroseptally and

pre-equatorially. From the nasal orbitotomy we performed total resection of the tumour, in which small cystic formations with brownish fluid were perceptible perioperatively, which were evaluated as "chocolate cysts". Lymphangioma was verified histologically, in which especially perfusion of the areas of the tumour predominated amongst the secondary changes, leading in places to their thrombotic content, which in their secondary changes caused the formation of "chocolate cysts" (fig. 3d). The postoperative period of actual healing was accompanied only by epiphora. After six months the orbit was without clinically manifest pathological resistance. As soon as in 1998, monocular dacryocystorhinostomy was subsequently performed on the left eye following interception of the lacrimal pathways within the framework of prior resection of lymphangioma. A control histological sample of scarry tissue was taken, which demonstrated a scarred residue of the angiomatous tumour formed

by very dense fibrous tissue. Despite two repeat operations during the further course, attempts to ensure through flow of the lacrimal pathways did not succeed in a fully satisfactory manner up to 2003. The orbital finding did not change. Ultrasound examination did not demonstrate any manifest pathological resistance. In 2006 MRI was indicated for the purpose of repeat verification of the therapeutic result of resection of the tumour, which excluded lymphangiomatic infiltration. The boy continues to be monitored in his place of residence.

#### Patient no. 4

The fourth case included in our study cohort concerned a twenty three year old woman who had been sent from the district in 1996 due to only unilateral chronic hyperplastic conjunctivitis with recurrences of saturation and perfusion. Anamnestically the complaints had persisted for more than ten years. Subconjunctive tumescence was of a gelatinous charac-



**Fig. 4** Patient no. 4 above and patient no. 5 below  
a – separate lymphangioma of bulbar conjunctiva.  
b - histological verification of conjunctival lymphangioma (lymphatic tissue – arrow).  
c – left-sided lymphangioma of orbitopalpebral area in 2006.  
d – lymphangiomatosis now afflicting both orbits in 2011.

ter with dilation of the capillaries, and reached from the limbus to the depth of both fornices (fig. 4a). We decided in favour of probatory excision of the subconjunctival tumorous tissue. Histological verification demonstrated chronic inflammatory infiltration, as well as structures characteristic of lymphangioma (fig. 4b), namely a system of interconnected lymphatic cavities, focally in their edges with islands of lymphatic tissue. We did not indicate a radical surgical procedure and recommended solution of the condition at a time of pronounced secondary conjunctival injection with local application of collyrium, namely a combination of antihistamines with adstringent effect and fluorometholone.

#### Patient no. 5

The last patient in our cohort, sent for consultation in 2006, was a ten year old boy (fig. 4c). The first symptoms of tumorous pathology of the left orbit appeared at the age of three years. Subsequent partial resection of the tumour in the area of the upper eyelid in an attempt to alleviate pseudoptosis at a non-ophthalmological centre did not have the envisaged cosmetic effect. A histological examination verified lymphangioma. This represented a combined type of lymphangioma of the orbit, which also infiltrated the cheek on the afflicted side and reached into the oral cavity. With regard to the fact that it later spread also to the forehead and the other side of the face, the finding was concluded to be a combined venous lymphatic vascular malformation of the frontal area and both orbits. The scope of affliction was confirmed by an MRI examination in 2011, which demonstrated an analogous cystic formation also in the right orbit. On the left there was infiltrative tumescence of the upper eyelid, and the dorsonasal formation in the orbit was stationary. The examination did not detect any further intracranial changes. The actual clinical finding was manifested in an indication of axial protrusion on the right side and on the conjunctiva in the internal corner by gelatinous tumescence of a salmon pink colour. The actual orbit was without palpable resistance, the eyeball was reducible and freely mobile. On the left side the pronounced tumescence of the upper eyelid restricted its motility and constricted the eye aperture, the eyeball was deviated downwards to 20 degrees of convergence (fig. 4d). The finding on the anterior segment and the intraocular finding were physiological bilaterally. Amblyopia from secondary droop of the upper eyelid due to the tumour contributed to a deterioration of vision in the left

eye (VOL 0.3 with correction – 0.75 = +2.0/100, VOR 1.0 naturally). We did not recommend a further intervention with regard to the above diagnosis, and the patient remains under our observation.

## DISCUSSION

Since the publication of the monograph by Dr. J. Otradovec (1986) (39), we have not recorded any more extensive work on this congenital hamartoma of the orbit in the Czech and Slovak literature. Nevertheless, the observations here remain a literary source, also due to the quality of writing. In addition, the number of works on the theme of oncological orbitology since the breakup of Czechoslovakia in the founding period of both ophthalmological societies in the subsequent two decades has not reached an average of even one article per year. Only nine studies have been devoted to the issue of paediatric ophthalmology (1, 9, 10, 11, 12, 24, 26, 40, 43), three studies are focused on an adult age clinic (19, 28, 29), four articles state separate operation of the orbit (20, 25, 27, 33), and two works focus on special diagnostics (34, 42). The last diversely extensive overviews of lymphangioma are presented in a Slovak monograph by Professor A. Gerinec (2005) (13), and also in chapters 16 and 17 of the Czech attestation textbook *Ophthalmology* by Professor P. Kuchynka et al. (2007) (30). Lymphangioma ranks amongst tumours of childhood age with manifestations of a finding in adulthood, and is relatively rare, with frequency stated between 1-5% (39). A summary evaluation of the literary citations of the main paediatric ophthalmological works at the end of the 20th century stated its frequency at up to 3% (26). The most comprehensive retrospective study in all age categories over the last thirty years, which assesses 1264 patients with orbital tumours and stimulating conditions, states a 4% frequency of lymphangioma (46). In the Asian population the incidence of vascular tumours in childhood age is higher, a twenty one year study of 244 cases of orbital tumours demonstrated an 11% representation of lymphangioma at the age of up to 9 years (37). One of the first assessments of orbital tumours in children is a cohort of 174 patients from the period 1923-1977, in which lymphangioma was diagnosed in ten children (5.7%) (21). In two Slovak eight year studies, which are to date the only studies on childhood orbital tumours within our region, lymphangioma was diagnosed twice in each study, on 78 paediatric patients

from 1987-1994 (10) and 130 paediatric patients from 1992-1999 (12) respectively. Our observation of paediatric patients with lymphangioma in the orbitopalpebral region links to a previous ten-year configuration of orbital tumours in 87 adult patients, since all five patients were observed for varying periods at the time of this study (28). The low frequency of occurrence of this tumour in the population and the unclear symptomatology from the inception of the pathology must always lead to a differential diagnostic consideration. An acute beginning of manifestation of lymphangioma caused by intratumorous haemorrhage is manifested in sudden exophthalmos, which may belong to a picture of a malignant process. At childhood age mainly malignant rhabdomyosarcoma of the orbit is considered in differential diagnostics. Manifestations of initial perfusion of the skin of the eyelids and conjunctiva may also be for example the first manifestation of metastases of Ewing's sarcoma of the bone and above all neuroblastoma, where increased sedimentation of erythrocytes indicates this possibility (26). The issue of orbital cysts in children is extensive, in which processes primarily without cysts, which may however have a cystic component, are more difficult to distinguish. These included adenoidal cystic carcinoma, as well as rhabdomyosarcoma and also lymphangioma. Examination using CT and MRI helps their differentiation, but is not pathognomonic (49). The presence of "chocolate cysts", which form through the colliquation of blood in haematomas generated through haemorrhage into the tumour (8), helps to differentiate lymphangioma from capillary haemangioma. The paramagnetic property of haemoglobin helps in the detection of haemorrhagic cysts in tumours well supplied with blood vessels, in the phenomenon of "through flow" (4). In the method of MRI with high resolution a surface coil is used, which improves the diagnostic information in vascular tumours (cavernous haemangioma, lymphangioma, varices or AV malformation), whereas differentiation in solid tumours is more arduous and histological verification is essential (32). Ultrasound examination of the orbital cavity ranks amongst the methods of examination which have a fundamental influence from the perspective of differential diagnostics and treatment of orbital afflictions. This represents a non-invasive, quickly available examination which can be performed in outpatient care, in hospitalisation and in the operating theatre. With regard to the undemanding nature

of the examination for the patient and its harmlessness to the human organism, it is possible to conduct this examination repeatedly, not only on adults but also on children. In the case of lymphangioma of the orbit, the display of the echographic image in both B and A is sufficiently characteristic that it mostly does not cause diagnostic difficulties. This represents a soft, compressible tumour without signs of internal vascularisation. With regard to the infiltrative character, the tumour is less well bordered on the B scan, and its surface is generally irregular. Lymphangioma of the eye socket is formed by a network of lymphatic cavities filled with a thin stroma with lymphocyte infiltration. This is corresponded to echographically by the irregular internal structure and reflectivity in the B scan, alternation of higher echoes (septa) and lower echoes (stroma) in the A scan. In the case of haemorrhage into the tumour we record increased reflectivity of the interseptal spaces (2). A modern method is the pulse Doppler technique for the description of intratumorous vascularity, which is of significance for differentiating haemangioma and lymphangioma (7). The result of ultrasound examination in our two four year old boys (patients 2 and 3) was sufficiently conclusive for surgical intervention that the children were operated on on the third day after admittance. The cohort of our five patients included one patient with lymphangioma of the conjunctiva, in which a radial solution was not possible because the lymphangioma had infiltrated the entire bulbar conjunctiva up to both fornices. Isolated observation of lymphangiomatosis, stated as congenital venous and lymphatic malformations in two localities on the skull (6) in our patient primarily afflicted the orbit and upper eyelid on the left side, from which it passed over the frontal region of the skin of the skull into the right orbit. To date no complex solution is known for this pathology. The main complication is described unconnected vascular malformations (23), which thankfully were not confirmed in our patient. A combination of orbital lymphangioma with cranial arteriovenous malformation and thrombocytopenia, indicated as Kasabach-Merritt syndrome, was described (50). In differential diagnostics it is also necessary to consider the etiology of cavernous haemangioma in the case of these lymphatic-venous malformations. It has similar symptoms of ptosis, pathological deterioration of vision and deviation of the eyeball. Unlike the lymphangiomatic etiology of malformation, a well bordered formation is visible on MRI, nevertheless histological verification is necessary for

diagnosis (16).

The main representation in the group of five tumours was of orbital lymphangiomas in three boys, in which the result of the therapeutic endeavour was well evident within a short period of time following total and also partial resection of the tumour. A role in the favourable response through the reduction of the tumour after its partial removal was played by the scarry regression and fibrous conversions already before the surgical intervention. The finding was confirmed by a histological examination after one year within the framework of dacryocystorhinostomy and control MRI eight years later. The result of actinotherapy was highly effective, furthermore without any side effect on post-radiation changes in the lens and other intraocular tissues even after 17 years. The basis of the good effect was the use of a linear accelerator, which enabled very precise and targeted focusing of the actinotherapy. Both methods are fundamental therapeutic procedures (39), but the results of irradiation were still unconvincing during the last century (22). Repeated excision of small lesions with subsequent cryotherapy was also attempted (44). The fundamental procedure still remains surgical solution. Diffuse orbital lymphangioma can be resolved using a Krönlein surgical technique to achieve a good effect with clinical remission using subtotal resection (3). Extraconal lymphangioma can be resected fully effectively without relapse within a short period. In the case of intraocular location of the tumour the possibility is only of a subtotal effect with the danger of neuropathy of the optic nerve (15). Improvement of the success rate of the surgical solution may be assisted by the application of tissue glue to the base of the fibrin in order to increase the effectiveness of resection and prevent relapse (5, 17). General corticosteroid therapy has been used in a number of children only as a supplement to treatment following surgical intervention in the case of lymphangioma of the orbit (48). By contrast, in the case of capillary haemangioma generally administered corticosteroids are the basic therapy (13, 30), sometimes intratumorous application of a depot corticosteroid is also used (9). On the basis of experiences with sclerotisation of lymphangiomatic lesions in the region of the throat and face (35, 36) its use has begun in isolated cases also for lymphangioma in the orbital area. Two preparations have been administered generally. The first is OK-432 (commercially Picibanil, Chugai Pharmaceuticals Co.): methanol acetone extract from *Streptococcus pyogenes*,

which activates NK cells, macrophages and T-cytotoxic lymphocytes, and regularly serves as an adjuvant to oncological therapy. The second pharmaceutical is the peptide antibiotic Bleomycin (commercially Bleoxane, Bleocin): a product of fungal *Streptomyces verticillus*, which is a cytostatic interfering through DNA induction of fractures and intercorporation of thymidine, applied in the case of various types of lymphomas. In a comparison of the surgical procedure and application of one of the above preparations, an optimal effect was produced in the case of OK-432 (35). A side effect was a local inflammatory reaction, which however did not lead to damage or scarring of the surrounding tissue, but after an interval of 13 years of sclerotherapy of lymphangioma in children was no longer evaluated as fully effective (36). In the case of simple cysts and macrocysts, the effect of OK-432 was better than surgical intervention, which was itself more effective on microcysts and cavernous formations (38). The use of OK-432 in a dose from 0.02 to 0.05 mg/ml for sclerotisation of lymphangioma of the orbit has so far been only isolated (31, 49, 51). Ptosis appeared transitionally as a side effect (51) and the evaluation was not entirely favourable primarily due to local inflammatory reaction (31). Nevertheless, the method of sclerotisation of this tumour, formerly used primarily in the South-East Asian region due to the higher occurrence of lymphangioma, is continuing to develop. Further options for non-surgical invasive therapy of orbital manifestations of lymphangioma or other vascular lesions are still being assessed. 5% sodium morrhuate solution has been used as cytreduction of the tumour without significant side effects (47). Chemoablation using ethanol in combination with intracystic sodium tetradecyl sulphate upon transcutaneous application has been effective as a primary treatment in macrocystic and microcystic lymphatic malformations of orbits, as well as for relapses following surgical interventions generally in twenty patients (18). Injections of pingyngmycine into lesions of orbital vascular malformations in 13 patients have reduced their vascularity and were accompanied only by a mild inflammatory reaction (52). In therapy of vascular tumours generally administered beta blockers have also been used, e.g. in the case of capillary haemangioma of the orbit (14). They have also been used so far in isolated cases in lymphangioma (40), but according to PUBMED outside of the orbital region, which is in accordance with the paediatric ophthalmological textbook (13).

## CONCLUSION

Lymphangioma of the conjunctiva and rare observation of lymphangiomatosis cannot yet be influenced by com-

plex therapy. Extraconal localisation of the tumour was successfully resolved through surgical intervention in two patients through total and subtotal resection respectively. For intraconal lymphangioma with generated neuropathy

accompanied by CME a good choice was actinotherapy by linear accelerator, which enabled targeted focus on the tumour without side effects of radiation on the lens and other intraocular structures.

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