

ORBITAL TUMORS - CLINICAL CASES PRESENTATION

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ABSTRACT

Introduction: Tumors of the orbit are rare diseases in ophthalmic pathology – 3.5 - 4 %. They are a great challenge for the ophthalmologist. **Purposes:** Presentation of epidemiology, clinical signs and symptoms, histology, methods for diagnosis and treatment and follow up of patients with orbital tumors for the period 2001-2005. **Materials and methods:** A retrospective study was carried out of 28 patients with orbital tumors for the period 2001-2005. **Results:** The study includes 28 patients with diagnosis Orbital tumor - 17 males and 11 females. The greater number being in the sixth and the seventh decade -17 patients (61 %). **Conclusion:** Tumors of the orbit are always a great diagnostic problem. In orbital processes all diagnostic methods have to be used and the results from the treatment should be followed regularly.

Key words: orbital tumor, neoplasm, eye, proptosis

INTRODUCTION

Tumors of the orbit are rare diseases in ophthalmic pathology – 3.5 - 4 % [1, 2]. They are a great challenge for the ophthalmologist since they present with a variety of signs and symptoms and they are often difficult to diagnose at the initial stages. All anatomic structures of the orbit can give rise to neoplasm. Primary orbital tumors includes à several benign and malignant neoplasms. The direct extension from contiguous anatomical structures, lymphoproliferative disorders, and hematogenous metastasis result in secondary orbital invasion [3]. Top 3 pediatric tumors are dermoid cysts, capillary hemangiomas, and rhabdomyosarcoma [4, 5]. Top 3 adult tumors are lymphoid tumors, cavernous hemangiomas, and meningiomas [2, 3]. The major presenting symptom is proptosis, resulting from the mass effect. Changes in visual acuity or field, diplopia, extraocular motility disturbances, or pupillary abnormalities can result from invasion or compression of intraorbital contents secondary to solid tumor. Lid dysfunction and lagophthalmos or lacrimal gland dysfunction can result in exposure keratopathy, keratitis, and thinning of the cornea [2].

CLASSIFICATIONS of orbital tumors:

BY HISTOLOGY:

A. - **Benign**

- **Malignant**

B. - **Epithelial:** Tu mixtus, cylindrinoma, adenocarcinoma, carcinoma (secondary)

- **Mesenchymal:** fibroma, lipoma, rhabdomyoma, osteoma, sarcoma

- **Vascular:** hemangioma, lymphangioma

- **Neurogenic:** glioma, schwannoma, neurofibroma, meningioma

- **Lymphatic and hematogenic:** lymphoma, lymphosarcoma, lymphoid hamartoma, plasmacytoma

- **Developmental:** teratoma, dermoid cyst, choristoma

- **Pigment:** melanoblastoma

BY ORIGIN:

- **Primary** - lesions originating from the orbital tissues

- **Secondary** - lesions originating from the neighboring cavities and tissues

- **Metastatic** - lesions reach the orbit via hematogenous or lymphatic spread

PURPOSES

Presentation of epidemiology, clinical signs and symptoms, histology, methods for diagnosis and treatment and follow up of patients with orbital tumors for the period 2001 - 2005.

MATERIALS AND METHODS

A retrospective study was carried out of 28 patients with orbital tumors for the period 2001-2005. Documentation from the archive of Eye clinic, UMBAL - Pleven was used: Clinical history, Operative register, histological results.

RESULTS

For the period 2001 - 2005: 28 patients with orbital tumors

Sex ratio: 17 males and 11 females

Age ratio:

• 2 children - at the age of 11 and 12

• 26 adults - at the age of 34 to 81 (middle age 64)

The greater number being in the sixth and the seventh

decade -17 patients (61 %)

Malignant tumors - 14 (56 %)

Benign tumors - 11 (44 %)

Primary tumors - 14 (56 %)

Secondary tumors - 9 (36 %)

Metastatic tumors - 2 (8 %)

Epithelial tumors – 10 (42 %)	
Basal cell carcinoma	4
Spinocellular carcinoma	2
Mucoepidermoid carcinoma	1
Adenocarcinoma	3
Mesenchymal tumors – 2 (8%)	
Angiolipoma	1
Angiofibroma	1
Lymphatic tumors - 4 (16 %)	
Lymphoid chamartoma	1
Plasmocytoma	1
Langerhans Cell Histiocytosis	1
B-cell lymphocytic lymphoma	1
Vascular tumors – 1 (4 %)	
Cavernous hemangioma	1
Developmental tumors – 3 (12.5 %)	
Dermoid cyst	3
Pigment tumors – 1 (4 %)	
Malignant melanoma	1
Pseudotumors – 3 (12.5 %)	

Surgical procedures: Biopsy - 6 patients, Excision - 13 patients, Enucleation - 3 patients, Exaneration - 3 patients

Recurrence of the tumor formation was found in 3 cases from 1 to 2.5 years postoperatively.

A case of EOSINOPHYLLIC GRANULOMA (fig.1)

K.M.S. - an 11-year-old boy with a two month history of oedema and redness of the upper eyelid of the right eye. A soft tumor formation is felt under the skin, engaging it, but progressing towards the orbit. The eye has normal motility and function. The CT shows a 20 mm in diameter soft tissue lesion causing bone destruction of the upper lateral orbital wall. After the excision of the tumor the bone loss of the orbit is found and the tumor is seen not distinctly limited from the surrounding tissues with expansion into them. Histological diagnosis: Eosinophyllic granuloma - a very rare tumor of the haemopoetic and lymphoid tissue 5/1.000.000, most often found in children [6]. Postoperatively a few courses of chemotherapy are followed. One year after the excision: a light depression of the skin. The control CT shows no signs of recurrence.



Fig. 1.

A case of EMBRYONAL RHABDOMYOSARCOMA

(fig.2)

M.K.B. - a 9-year-old boy with a small tumor of the upper eyelid of the right eye. The tumor enlarges in a few months, dislocates the eyeball and causes a drop in the visual acuity. The tumor is removed after CT. Histological diagnosis: Embryonal rhabdomyosarcoma a very malignant tumor, seen in children [7, 8]. A few courses of chemotherapy are followed. The tumor recurs in one year and is excised by a neurosurgeon. Due to a new recurrence of the tumor, which showed rapid progression, the patient is admitted to the Eye clinic in Pleven for orbital exaneration. The tumor and the orbital content protrude in the eyelid fissure. The MRI shows multiple metastases in the internal organs and regional lymph nodes. Two weeks after the operation a lethal outcome occurs.



Fig. 2.

A case of MELANOTIC SCHWANNOMA (fig.3)

K.P.E. - a 68-year-old woman complains of pain and dislocation of her right eye. Following CT an excision of the orbital tumor is carried out, which tumor has the appearance of haemangioma. Histological result: Melanotic schwannoma with malignant potential an exceptionally rare tumor, derived from some of the oculomotor nerves [9]. Postoperatively the patient has 3 courses of chemotherapy. Four months after the excision she is seen with a recurrence of the tumor with temporal dislocation of the eyeball and the tumor prolapses under the upper eyelid. CT scan: retrobulbar lesion 30/50 mm, infiltrating the orbital tissues. She is admitted to the Plevn Eye clinic for exaneration of the orbit. Pulmonary metastases are found on X-ray examination. After the exaneration she is referred for chemotherapy.



Fig. 3.

A case of CARCINOMA PLANOCELLULARE (secondary orbital tumor) (fig.4)

B.J.I. - an 80-year-old woman with a tumor growing between the eyelids of the left eye. The tumor is solid in consistency with irregular surface. The process engages the eyeball. Enucleation follows. Histological results: Low-differentiated planocellular carcinoma. Postoperatively radiotherapy is performed. 1 year after the enucleation a recurrence of the tumor is diagnosed with secondary engagement of the eyelids and the orbital contents. Exaneration of the orbit with excision of the eyelids is performed. On follow-up 2 months later a granulomatous tissue develops in the orbit, no signs of recurrence.



Fig. 4.

A case of CAVERNOUS HAEMANGIOMA (fig.5)

P.S.C. - a 67-year-old man complains of double vision at upgaze and rightgaze. The visual functions are preserved. A consistent formation is palpated superior and nasally of the eyeball, which formation is movable and reductable. Light proptosis of 4 mm is measured accompanied by reduced adduction and supraduction, light ptosis of the upper eyelid and nasal oedema of the eyelid. CT shows medially of the left eyeball an oval zone 20/10 mm in diameter with abrupt borders, not connected to the orbital tissues. After orbitotomy a vascular tumor is excised. Histology: Cavernous haemangioma - one of the most frequent orbital benign tumors in adults. Control CT 2.5 years later: No data for recurrence.



Fig. 5.

DISCUSSION

Patients with moderate unilateral exophthalmos, with excluded endocrine cause, should be thoroughly examined

for tumors of the orbit in order to explain the etiology which allows due and adequate treatment.

In the late stages of a malignant tumor the problem comes out of the limits of the eye. This imposes a more aggressive therapy which causes further sufferings to the patient and the outcome is fatal.

The patients have to be controlled with some diagnostic imaging method in order to choose the proper treatment either conservative or surgical.

CONCLUSION

Tumors of the orbit are always a great diagnostic problem.

In orbital processes all diagnostic methods have to be used and the results from the treatment should be followed regularly.

Cooperation with neurologists, neurosurgeons, otorhinolaryngologists, pathologists, hematologist and oncologists is necessary for correct diagnosis and treatment aimed at maximal sparing of the eye and its functions. In many cases the adequate treatment of the orbital tumor is lifesaving.

The surgical treatment of the orbital tumors requires ophthalmologists with high qualification.

REFERENCES

1. Balabanov Ch., Parashkevova B., Murgova S., Iordanov I. Tumors of the eye and adnexa. Bulgarian Review of Ophthalmology. 2005; 49:12-20.
2. Goldberg SH, Cantore WA. Tumors of the orbit, Curr Opin Ophthalmol. Oct 1997; 8(5):51-6.
3. Weisman RA, Kikkawa D, Moe KS, Osguthorpe JD. Orbital tumors. Otolaryngol Clin North Am. 2001;34:1157-74, ix-x. Review.
4. Bouguila H. Malek I. Nacef L. Marrakchi S. Dagfous F. Ayed S. [Intraorbital dermoid cyst. Apropos of a case] J Fr Ophtalmol.1999; 22:438-41.
5. Gloor B, Kalman A. [Neoplastic space-occupying lesions of the orbit. I. Review; hemangioma, lymphangioma and embryonal rhabdomyosarcoma]. Klin Monatsbl Augenheilkd. 1992; 201(5):291-301.
6. Krivoshiiska E., Nedkova B., Balabanov Ch., Iordanov I., Karaivanov M., Konstantinov D. Langerhans cell histiocytosis – a clinical case. In: Tumors of the eye and the eyes adnexa. Pleven, 2006, 147-153.
7. Shields CL, Shields JA, Honavar SG, Demirci H. Comment in: Ophthalmology. 2003; 110:877; author reply 877-8.
8. Mannor GE, Rose GE, Plowman PN, Kingston J, Wright JE, Vardy SJ. Multidisciplinary management of refractory orbital rhabdomyosarcoma. Ophthalmology. 1997; 104:1198-201.
9. Goasguen O, Boucher E, Pouit B, et al. Melanotic schwannoma, a tumor with a unpredictable prognosis: case report and review of the literature. Neurochirurgie 2003; 49:31-38.
10. Clinical spectrum of primary ophthalmic rhabdomyosarcoma. Ophthalmology, 2001; 108:2284-92.

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