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Benign Tumors of The Orbit

Marieta Dumitrache[1]* and Ciobotă M[2]

[1]*University of Medicine and Pharmacy Bucharest, Romania.*

[2]*Doctoral School of Biology, Faculty of Biology, University of Bucharest, 91-95 Splaiul Independenței, 050095 Bucharest, Romania.*

***Correspondence:**
Marieta Dumitrache, University of Medicine and Pharmacy Bucharest, Romania.

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ABSTRACT

Primary benign orbital tumors are more common in adults, some of them may have malignant potential after 60 years. Histopathologically, benign tumors can be: congenital, fibroosseous, vascular, from nervous structures, lymphoproliferative, histiocytic, muscular, conjunctival, parasitic. In children, the most common are: dermoid cyst, epidermoid, lymphangioma, hemangioma, glioma NO. Located intra or extraconally, benign orbital tumors are clinically manifested by: irreducible exophthalmos, ophthalmoplegia, fundus changes due to tumor compression of NO. Paraclinical investigations CT, MRI, ultrasound are necessary to highlight the tumor and neighboring structures, orientation in the surgical treatment of tumor excision performed depending on the local evolution, but also on the possible malignancy. Sometimes decompression is necessary for unresectable tumors. Primary benign (and malignant) tumors may develop in the orbit secondary to tumors in the vicinity (meningioma, nasopharyngeal fibroma, mucocele). Mucocele (most commonly frontal) present in adults and the elderly requires complete excision with restoration of normal sinus drainage. The prognosis of benign orbital tumors is generally good and is related to the origin of the tumor and its location.

Keywords

Primary, Secondary benign orbital tumors, Malignant orbital tumors, Hemangioma, Dermoid cyst, Fibroma, Lipoma, Meningocele, Mucocele, Osteoma.

Introduction

- The orbits are two bony cavities located laterally from the midline, pyramidal in shape, with the base directed forward and outward, with the apex directed posteriorly with an anteroposterior axis of 4.5 cm. The orbit is prolonged by the optic canal.
- The orbital cavity contains: the anterior eyeball, NO, extraocular muscles, optic nerve, cranial nerves, blood vessels, components of the lacrimal apparatus behind the eyeball
  - all these components can present**primary orbital tumors**benign or malignant
- The orbit is limited by the bone structures of the neighboring skull, maxilla, ethmoid, sphenoid, frontal, zygomatic bone, palatine bone, lacrimal, and facial sinuses.
- The orbit has openings in its posterior pole through which it communicates with neighboring cavities, which makes it possible for neighboring tumors to extend into the orbit, generating secondary orbital tumors.
  - intraocular tumors spread into the orbit: choroidal melanoma, retinoblastoma
  - soft tissue tumors spread into the orbit – palpebral epithelioma, conjunctival neocarcinoma, mucocele
  - tumors in the middle cerebral fossa – spread to the orbit: meningioma of the lesser wing of the sphenoid
  - nasopharyngeal fibroma [1].
- Orbital tumors can also be local periregional metastases or distant orbital metastases – from primary malignant tumors located in the lung, breast, prostate, melanoma (most common); they constitute metastatic orbital tumors:

Classification of benign orbital tumors

Orbital tumors can be:

- benign orbital tumors, located in the orbit
- malignant tumors of the orbit – neoplastic can invade nearby tissues locally or cause distant metastases

Histopathologically, benign orbital tumors can be:

- congenital tumors: cephalocele, dermoid cyst, epidermoid cyst
- fibroosseous tumors – bone dysplasia, orbital osteoma
- vascular tumors of the orbit – capillary hemangioma, cavernous hemangioma, lymphangioma, hemangiopericytoma
- histiocytic tumors – eosinophilic granuloma, juvenile xanthogranuloma
- tumors of the nervous structures of the orbit – glioma NO, meningioma of the sheath NO
- lymphoproliferative lesions – lymphoid hyperplasia
- muscle tumors – rhabdomyoma
- conjunctival tumors – fibroma, lipoma
- parasitic tumors – hydatid cyst of the orbit [2].

Orbital tumors in children:

- benign: dermoid cyst (choristoma), epidermoid cyst, lipodermoid, capillary hemangioma, lymphangioma, optic nerve glioma [3].

Orbital tumors by location are:

- intraconal – cavernous hemangioma, vascular malformations, lymphoma
- extraconal - capillary hemangioma, lymphoma, rhabdomyoma.

- The evaluation of an orbital condition requires differentiating an orbital lesion from a periorbital condition, or from an intraocular lesion.
- To support the diagnosis and apply appropriate treatment, a detailed history, a thorough clinical examination, as well as laboratory tests specific to an orbital lesion are necessary.

Clinical signs

Exophthalmia represents an abnormal protrusion of the eyeball, due to an increase in the volume of the retroocular structures (the orbit is a cavity with inextensible walls). Exophthalmos is measured with an exophthalmometer and is mild (21-23 mm), moderate (24-27 mm), severe (over 28 mm).

Exophthalmos can be:

- axial - lesions are located inside the muscular cone (cavernous hemangioma or optic nerve tumors)
- lateral, eccentric (dystopia) - lesions are extraconical (maxillary sinus tumors, dermoid cyst, lacrimal gland tumors, mucocele, osteoma)
- it may have a rapid or slow development (in correlation with the etiology);
- reducible or irreducible:
- **pseudo-exophthalmos**, impression of protrusion of the globe
  - **Tumor exophthalmos is irreducible, axial or lateral, accompanied by diplopia and ophthalmoplegia.**
  - **ophthalmoplegic**includes – ocular motility disorders due to orbital lesions of firm consistency; or optic nerve compression (meningioma).

Changes in the fundus of the eye

- Retinal changes:
  - papillary edema followed by atrophy NO
  - dilation and tortuosity of the veins (arteriovenous communication);
  - vascular occlusions (optic nerve tumors)
  - choroidal folds (tumors, mucocele)

Imaging investigations

Complementary examinations are necessary to establish a positive diagnosis of orbital tumor: lateral skull radiography (Blondeau incidence checks the orbital walls and neighboring sinuses), CT, MRI, B-ultrasound, and vascular explorations if necessary.

- CT: is useful for highlighting areas of bone lysis, fractures, and evaluating optic nerve tumors;
- MRI: highlights soft tissues more clearly, thus demyelinating, vascular or hemorrhagic lesions can be revealed; MRI images can be useful for studying the structures of the orbital apex, periorbital spaces, orbitocranial tumors.
- Ultrasound: has a lower diagnostic value than CT, especially for lesions of the posterior orbit, sinuses, or those of the intracranial space.
- Radiography: currently has a reduced diagnostic value, after the use of CT and MRI. There are two views that can more accurately highlight orbital lesions: Caldwell (nose-forehead-plate), Waters (occipitomental, with the chin slightly raised) [4].

Positive Diagnosis of an Orbital Tumor

Diagnosing an orbital tumor is not easy, and definitive diagnosis is sometimes impossible..

Some signs that can guide the diagnosis: [1,5]

- Age
  - child: congenital malformations - dermoid cyst, angioma, lymphangioma, meningocele; retinal glioma spread into the orbit;
  - child under 10 years: rhabdomyoma; meningioma
  - adolescents: angioma, dermoid cyst, optic nerve tumors
  - adult – all benign, malignant tumors may be possible
  - elderly: malignant tumors spread to the orbit; metastatic tumors
- Previous history – breast and prostate cancer draws attention to possible metastasis
- Mode of occurrence, evolution
  - rapidly evolving exophthalmos – malignant tumor;
  - pain accompanying exophthalmos – malignant tumor, pseudotumor;
  - visual disturbances – optic nerve tumor
- Character of exophthalmos
  - axial – optic nerve tumor
  - lateral – orbital wall tumor
  - intermittent – vascular tumor
  - inflammatory with eyelid edema – pseudotumor
- Impaired ocular motility
  - Metastatic malignant tumor
  - pseudotumour
  - intracranial carotid aneurysm

- without motility impairment – benign tumor
- Palpation of the orbit
  - in the supero-internal angle – meningocele
  - balloon consistency – frontal sinus mucocele
  - boney, bony consistency – frontal sinus osteoma, ethmoidal
  - visual impairment, decreased AV – optic nerve tumor
  - CV involvement – lesser sphenoid wing meningioma spread into the orbit

Orbital tumors, depending on their location, can cause displacement of the eyeball in:
- downward displacement of the eyeball in: fibrous dysplasia, frontal mucocele, lymphoma,
- neurofibroma, schwannoma
- upward displacement of the eyeball in: lacrimal sac tumors, lymphoma
- lateral displacement of the eyeball in ethmoidal mucocele.

Infantile exophthalmos: capillary hemangioma, dermoid cyst, encephalocele, histiocytosis
Exophthalmos in children: capillary hemangioma, dermoid cyst, inflammatory pseudotumors, lymphangioma, optic nerve glioma, meningioma, neurofibromatosis, sphenoorbital encephalocele, plexiform neuroma
Exophthalmos in adults: hemangioma, orbital tumors, orbital pseudotumors
Enlargement of extraocular muscles on CT scan: lymphoma, histiocytoma, fibroma
Unilateral periorbital inflammations: ruptured dermoid cyst, orbital pseudotumors, eosinophilic granuloma
Painless exophthalmos: cavernous hemangioma, optic nerve tumors, neurofibroma, histiocytoma, lymphoma, osteoma
Painful exophthalmos: orbital pseudotumors, lymphangioma, hemangioma, meningioma, optic nerve glioma.

PRIMARY BENIGN ORBITAL TUMORS

Congenital Tumors - Orbital Cystic Lesions

- Cephalocele are congenital anomalies that consist of the herniation of the brain into the orbit - encephalocele, the meninges - meningocele, or both structures - meningoencephalocele.

This information can be located:
- *Previous*by frontomaxillary coalescence defect in the superointernal angle of the orbit with anterior and temporal globe displacement, becomes clinically evident after a few months, years (young age), manifests itself by pulsatile exophthalmos (or not), which may increase with the Valsalva maneuver, may be accompanied by craniofacial or ocular anomalies (hypertelorism, cleft palate, neurofibrosis, microphthalmia, coloboma, morning-glory syndrome).
- *back*– exceptionally by sphenoid dysplasia, located on the superointernal wall of the orbit, it may be accompanied by ocular malformations.

Dermoid cyst

It is a congenital teratoma (choristoma) tumor that occurs by the displacement of the ectoderm to the subcutaneous areas, along the embryonic fissures, and contains hairs, sebaceous and sweat glands, keratin and, rarely, even teeth, contained in a fibrous capsule.
- *Superficial dermoid*it appears in the first years of life, has the appearance of a round, smooth tumor, of firm consistency, painless, approximately 1-2cm in diameter, mobile under the skin, with an easily palpable posterior edge, located in the supero-temporal portion of the orbit (less often supero-nasal), with slow development;
- *Deep dermoid*occurs in adolescents or adults, presenting clinically: progressive exophthalmos, displacement of the eyeball, the presence of a firm tumor with imprecise posterior margins, ocular motility disorders and even decreased vision; it can extend beyond the orbit to the frontal sinus, temporal fossa or skull, it is a well-defined lesion, with increased internal reflectivity [6,7].

Treatment

- Surgical
  - Complete excision, along with the capsule, avoiding its rupture as much as possible
  - Incomplete excision can lead to recurrences, and the loss of contents into the surrounding tissues can generate painful, granulomatous inflammation, followed by fibrosis.
- Deep dermoid cysts, extended to the sinuses or intracranial, require a team approach with an ENT specialist or neurosurgeon. **Epidermoid cyst (choristoma)** It is derived from epidermal elements, contains keratin; its rupture can cause an acute inflammatory process.

FIBROUS TUMORS

It is a tumor of connective tissue, cartilage, and bone that has a progressive evolution and affects children and young adults.
There are 2 types:
- *monostatic*– located in the orbit where it can cause unilateral exophthalmos, diplopia, decreased AV due to optic canal compression, decreased hearing
- *poliostatic*– Albright syndrome, short stature, premature closure of the epiphyses, precocious puberty, hyperpigmented macules [8].

Orbital osteoma

It is a benign tumor that occurs in young males with slow evolution, being an orbital bone wall tumor originating in the paranasal sinuses, frontal, ethmoidal, rarely maxillary, sphenoidal or periorbital sinuses, clinically presenting sinus signs (nasal obstruction, nasal catarrh, epistaxis) and irreducible lateral exophthalmos [9].

Vascular Tumors of the Orbit

Vascular tumors are the most common benign tumors of the orbit.

Capillary hemangioma

It is the most common benign tumor in children with spontaneous involution in the following years. Clinical manifestation is present in the first weeks, months after birth, with a slight preponderance in girls 3/2, they enlarge at 6-12 months, can stabilize and regress after 5-8 years in 80% of children [10].

Superficial Hemangioma

The tumor is frequently located in the superonasal quadrant of the orbit with imprecise boundaries and possible extension towards the eyelid. Clinically, the tumor is purplish red (possible change to blue) and increases in size during sucking or crying.

Deep Hemangioma

Deep orbital tumor, frequently located in the antero-superior portion of the orbit, without skin color changes, associated with variable unilateral exophthalmos and, sometimes, with displacement of the eyeball from the axial position. Large capillary hemangiomas may be associated with systemic manifestations – Kasabach-Merrin syndrome (thrombocytopenia, anemia, decreased coagulation factors, heart failure). Treatment of capillary hemangioma is related to the evolution of the tumor, with possible spontaneous resorption.

Treatment is indicated when [11]:

- signs of optic nerve compression, exposure keratopathy
- amblyopia secondary to visual axis occlusion or associated astigmatism
- alteration of the aesthetic appearance of the face.

In superficial tumors, fast- or slow-acting corticosteroids (dexamethasone, triamcinolone) are administered.

- If necessary, systemic steroids can be administered for 3-6 weeks with a slow dose reduction.
- In the case of deep, well-demarcated tumors located in the anterior portion of the orbit, tumor excision can be performed (preferably at the stage of stopping the growth of the formation). Complete resection of the tumor is difficult to achieve. In the treatment of deep hemangioma, the following may be indicated: cryotherapy, low-dose radiotherapy, interferon alpha 2a, 2b for several months. Preoperative embolization to reduce the size of the tumor may sometimes remain the only treatment.

Cavernous hemangioma

Cavernous hemangioma is the most common benign orbital tumor in adults, 3-9%, which predominates in women 40-50 years old (pregnancy accelerates the progression) with progressive intraconical development behind the eyeball. It is an encapsulated tumor of soft consistency, formed by dilated vascular channels in which blood stagnates and where thrombosis is possible [12].

Clinically, the tumor manifests itself through:

- axial exophthalmos (intraconal location) with progressive evolution after 40 years with possible compression of the NO
- limitation of ocular motility
- small (moderate) decrease in VA, increase in IOP
- squint
- FO: papillary edema, choroidal folds due to NO compression.

Paraclinical investigations

- ultrasound – intraconical lesion, well circumscribed, with increased internal reflectivity
- CT, MRI – well-demarcated oval lesion, intraconical, with discrete, moderate contrast enhancement [13].

The patient requires clinical surveillance related to the progressive growth of the tumor, when complete surgical excision is necessary, the tumor being encapsulated. Recurrence is possible after incomplete resections.

Hemangiopericytoma

Hemangiopericytoma is a rare orbital tumor 0.1-0.3% that occurs in women aged 2-=70 years. It is a well-demarcated, encapsulated, red tumor located in the upper portion of the orbit. Clinically, it manifests itself by progressive unilateral exophthalmos, with slow impairment of vision and limitation of eyelid motility. By malignancy, possible local CNS invasion and even distant metastases in: lung, liver, bone.

Treatment

- Complete surgical resection would be indicated, but it is difficult in the case of large tumors (many tumors are bulky because the tumor's evolution can be asymptomatic for a long time).
- In case of recurrence or incomplete resection, 50cgy radiotherapy and exenteration are recommended.

Lymphangioma

Lymphangioma is a rare, benign congenital tumor with onset under 10 years of age that may remain asymptomatic until adulthood, when it may occasionally be evidenced after trauma or intratumoral bleeding. The tumor has a slow evolution, and can affect the conjunctiva, eyelids, deep orbit. It is accompanied by pain, slowly progressive exophthalmos, palpebral ecchymosis, subconjunctival hemorrhage, decreased vision, "chocolate cyst" hemorrhage.

Superficial tumors are more common and have a better prognosis for AV.

CT and MRI highlight the multicompartmental structure of lymphovenous malformations.

Clinical forms

- superficial anterior lesions of the eyelid or conjunctiva with cystic spaces
- deep lesions with exophthalmos, pain, "chocolate cyst" hemorrhage, decreased vision.
- Surgical treatment of lymphangioma is difficult due to the infiltrative nature of the tumor and requires clinical monitoring:
- surgical excision is subtotal because the tumor is friable without an external capsule
- after surgical treatment, a remnant is kept so as not to affect orbital circulation
- there is no risk of malignancy
- large cysts can be aspirated and the vessels electrocoagulated
- In the case of a bleeding “chocolate cyst,” surgical drainage or carbon dioxide laser resection is indicated [14].

Tumors of the Nervous Structures of the Orbit

Optic Nerve Glioma

NO glioma is a tumor developed from astrocytes, relatively common in children, 20-25% in the first decade of life, predominates in females. The tumor can be solitary or can be associated with Recklinghausen neurofibromatosis 2.5-50%. NO glioma is a benign, slow-growing tumor, associated with morbidity and possible mortality when the glioma extends to the optic chiasm, hypothalamus, third ventricle.

Clinical signs and symptoms [15].

- early progressive decrease in vision without correlation with exophthalmos
- unilateral axial proptosis with late onset after decreased vision, with possible impairment of ocular motility + strabismus
- papilledema progressing to optic atrophy
- intracranial extension can cause headache and facial pain
- chiasmatic extension can cause pituitary and hypothalamic dysfunction
- nystagmus due to compression of the third ventricle

Most commonly, glioma is associated with extension to the optic chiasm (70%), sometimes with extension to the hypothalamus [15,16].

Paraclinical Investigations

CT shows fusiform dilation of the NO that is dense to the brain

MRI highlights intracranial extension.

Treatment

- isolated tumor of the NO, with normal VA, moderate exophthalmos, requires clinical monitoring, MRI of the patient
- Patients with severe exophthalmos, extensive pain, chiasmatic tumor require surgical excision of the tumor with preservation of the eyeball.
- If the tumor has spread to the chiasm, surgical excision is NOT indicated. These cases require chemotherapy and radiotherapy that precede neurosurgical intervention.
- The vital prognosis is favorable in cases with tumors limited to the NO, but is severe in adults with intracranial tumor extension, with death within 6-12 months.

Optic Nerve Sheath Meningioma

The meningioma of the NO sheath is a rare benign tumor (after the NO glioma in frequency), which develops from the meningothelial cells of the meninges (arachnoid) along the NO inside the orbit or optic canal. Unilateral tumor, present at middle age (40 years) more frequently in women (1.5/1) may be associated with neurofibromatosis II. Meningioma may infiltrate the NO, in the sheaths of nerve fascicles, vessels or extend bilaterally and intracranially favored by: young age, intracanalicular tumor location or/and the presence of neurofibromatosis II [17].

Orbital meningioma can be:

Primary Orbital Meningioma

- ***primary intraorbital meningioma*– NO sheath meningioma**which causes: early vision loss, limitation of eye movements, papilledema/optic atrophy, slowly progressive unilateral exophthalmos
- ***Meningioma with intradural extension clinically cannot be differentiated from glioma of NO***.

- Clinically, primary meningioma is accompanied by: the clinical triad which appears in order:
  - transient blurring of vision initially
  - decreased vision
  - signs of optic neuropathy due to tumor compression of the optic nerve with papilledema or optic atrophy; papilledema and VCR turgor precede the appearance of shunts by 1-2 years
  - Optociliary vascular shunts (connections between the VCR and the choroidal circulation, post VCR occlusion) provide an alternative for the removal of retinal venous blood from around the area of vascular obstruction caused by tumor compression. Optociliary shunts are present in sheath meningioma NO.
  - ocular motility deficit (frequently when looking up) due to stiffening of the NO sheath, rarely deficits are paretic due to oculomotor compression
  - exophthalmos that occurs after vision loss [18-23].

Paraclinical investigations

CT scan

- tubular or fusiform enlargement of the NO
- reactive hyperostosis if the tumor is adjacent to the bone
- calcifications NO 20-55%

MRI – differential diagnostic investigation

- "tram track" sign
- presence of calcifications
- absence of NO twisting
- absence of cystic degeneration within the tumor

Secondary Orbital Meningioma

It is an intracranial meningioma, which secondarily invades the orbit and originates from the sphenoid. The orbital invasion is through the anterior cranial fossa, superior orbital fissure, optic canal.

Clinical

- significant exophthalmos
- severe vision loss (greater in primary meningioma)
- palpebral edema on the ipsilateral side of the face (more pronounced if the tumor originates from the lateral side of the sphenoid wing).

Treatment for meningioma NO requires:

- continuous clinical follow-up and imaging (vision loss is slow, in elderly people with good VA the meningioma may remain confined to the orbit
- radiotherapy 5000-5500 to improve vision
- Surgical excision indicated in: aggressive tumors, large AV decrease, chiasmatic extension [19].

Since tumor-related mortality is zero even in intracranial extension, the indication for surgical excision is to prevent tumor extension to the congeneric eye.

Favorable vital prognosis.

Schwanom – Neurinem

Schwannoma is a rare benign tumor of the peripheral or cranial nerves (inside the orbit). Clinically, the tumor is associated with: exophthalmos, painless diplopia, decreased ocular motility, rarely ophthalmoplegia, decreased vision with associated pupillary defect. Treatment is surgical with complete excision of the tumor with a favorable postoperative prognosis.

CONJUNCTIVAL TUMORS

Fibroma– dense, encapsulated tumor, non-adherent to the eyeball, located in the superointernal angle of the orbit near the orbital roof; accompanied by progressive tumor exophthalmos. Requires surgical treatment [20,22].

Lipoma– structure similar to adipose tissue, developing in the vicinity of a muscle or nerve.

Lymphoproliferative Orbital Lesions

Lymphoid hyperplasiabenign reactive

It is a rare lymphoid proliferative lesion located in the anterosuperior portion of the orbit with possible involvement of the lacrimal gland, and possible evolution to malignant lymphoma in 20%. Clinically it is accompanied by: painless exophthalmos, displaced eyeball, ocular motility disorders, firm "rubbery" pink-orange formation, with a "salmon" appearance located palpebrally at the edge of the orbit. Treatment requires clinical monitoring, chemotherapy and radiotherapy [20,21].

A Typical Lymphoid Hyperplasia

It is an intermediate tumor between benign hyperplasia and lymphoma, with the appearance of atypical hyperplasia associated with localizations in the lymphatic system. Local radiotherapy is used in the treatment of lymphatic system involvement and chemotherapy [22].

MESENCHYMATOUS – MUSCLE TUMORS– Rhabdomyoma – benign, clearly demarcated, encapsulated tumor adhering to an extrinsic muscle.

Histiocyt Tumors are a group of disorders resulting from the abnormal proliferation of histiocytes (Langerhans cells) that can affect the orbit.

Eosinophilic granuloma– it is a benign, isolated tumor, located supertemporally in the orbit, present in children, adolescents, CT and radiography show delimited osteolytic lesions.

PARASITIC TUMORS

Hydatid cyst of the orbit is a primitive echinococcosis, present in children, adolescents with insidious onset with orbital or retroorbital pain that increases, becomes permanent, paroxysmal, presents progressive exophthalmos after 3-5 months. Surgical treatment with cyst excision after formalinization by prior puncture with possible anaphylactic reactions.

SECONDARY BENIGN ORBITAL TUMORS

Benign Tumors Propagated into the Orbit

Tumors of the middle cerebral fossa

Meningioma of the lesser wing of the sphenoid

It spreads into the orbit by eroding the bone wall and may cause reduced exophthalmos. In evolution it may grow into the orbit simulating a primary tumor. Temporally localized it may be accompanied by exophthalmos and papillary edema. CT shows hyperostosis and calcifications. Surgical intervention is performed via the endocranial route [23].

Nasopharyngeal Fibroma

It is a rare tumor, present in male adolescents, which penetrates the orbit through the sphenomaxillary or ophenoideal fissure after invasion of the skull base. Clinically, exophthalmos is important and is accompanied by palpebral edema, oculomotor paralysis, and blindness [24].

Secondary Soft Tumors Spreading to the Orbit

Oral Mucocele

Mucocele located in the superointernal portion of the orbit represents a collection of secretions in a paranasal sinus due to obstruction of the drainage orifice by an infection, allergy, trauma, tumor, congenital lesion. Evolving by erosion of the walls (the frontal ethmoidal sinus is more frequently affected, less frequently the maxillary sinus. Mucocele is more frequent in adults and the elderly [25]. Clinically, mucocele presents with: headache, progressive exophthalmos with displacement in the external sphere of the eyeball. CT reveals opacification of the frontal or ethmoidal sinus with the absence of the ethmoidal septum and the presence of a bone defect that presents intraorbital extension. Treatment consists of complete excision of the mucocele, restoration of normal sinus drainage or obliteration of the sinus cavity.

Frontal Sinus Mucocele

Starting from the dilated frontal sinus after chronic inflammation of the sinus mucosa, the mucocele bulges into the orbital cavity and is accompanied by significant exophthalmos. Ethmoidal mucocele is rare, and maxillary sinus mucocele is exceptional.

Differential diagnosis

Orbital tumors must be differentiated from orbital diseases that are accompanied by exophthalmos, diplopia, ophthalmoplegia in: thyroid orbitopathy, myositis, orbital metastases. The prognosis of benign orbital tumors is generally good and is related to the origin of the tumor and its location.

Treatment of Orbital Tumors – surgical

- simple orbitotomy – benign tumors, encapsulated or not: diffuse, cavernous angioma; depending on the location of the tumor; the orbitotomy must be wide, with cleavage of the tumor
- extended orbitotomy – Kronlein operation with wide resection of the external orbital wall
- endocranial surgery under the frontal flap with orbital roof resection: optic nerve glioma, meningioma spread into the orbit [6,7].

Conclusion

Orbital tumors can be primary, secondary and metastatic, benign or malignant that can invade nearby tissues locally or cause distant metastases. Primary benign orbital tumors are: dermoid cyst, epidermoid, vascular tumors (capillary hemangioma, cavernous, lymphangioma), fibro-osseous (osteoma), tumors from the nervous structures of the orbit (glioma NO, meningioma), muscular (rhabdomyoma), conjunctival (fibroma, lipoma). The main clinical manifestation in benign orbital tumors is irreducible exophthalmos, axial or lateral, fast/slow, accompanied by diplopia, ophthalmoplegia; in NO compression, FO changes appear with papillary edema followed by optic atrophy, arteriovenous communications, choroidal folds. CT, MRI, ultrasound complete the diagnosis. In the orbit there may be secondary benign tumors propagated from the vicinity – meningioma of the lesser wing of the sphenoid reaches the orbit by eroding the bone wall and may initially cause small exophthalmos, which may grow (simulating a primary tumor). Mucocele (frequently of the frontal sinus) is present in adults and the elderly; requires complete excision with drainage of the sinus.

The treatment of orbital tumors is surgical: wide orbitotomy with tumor cleavage, simple for benign encapsulated tumors, if necessary, wide orbitotomy with resection of the external orbital wall. For NO glioma, meningioma propagated into the orbit, an endocranial approach under the frontal flap is necessary. The prognosis of benign orbital tumors is generally good.

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