Exophthalmic syndrome in children

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Abstract
This article reviews the exophthalmic syndrome in children which can be determined by the multiple pathological lesions.

We have to emphasise that in children, benign orbital tumors are more frequent than the malignant tumors.

Keywords: tumoral exophthalmic syndrome, optic nerve glioma, children.

Introduction
The exophthalmic syndrome in children can be determined by the multiple pathological lesions. The systematization of these tumors is necessary in order to ease the work of the team formed by the imagist, the neurosurgeon, the ophthalmologist, and the oncologist.

1. Congenital and developmental anomaly of the orbits
   1.1. True Congenital anophthalmia
   1.2. Microphthalmia
   1.3. Craniofacial deformities
      - Craniofacial syndrome diagnosis
       - Mandibulofacial dysostosis
       - Franceschetti syndrome
       - Oculo-auroculo-vertebral syndrome
       - Goldenhar syndrome
   - Cleidocranial dysostosis
   1.3.2. Craniosynostosis syndrome
      - Oxycephaly
      - Craniofacial dysostosis (Crouzon syndrome)
      - Apert syndrome
      1.4. Meningocele and cranio orbital meningoencephaloscele

2. Infectious orbital diseases
   2.1. Infectious diseases of the orbital wall
      - Severe osteoperiostitis
      - Chronic osteoperiostitis
   2.2. Infectious diseases of the orbital adipose tissue
      - Orbital abscess
      - Orbital phlegmon

3. Orbital inflammatory diseases
   3.1. Orbital cellulitis
   3.2. Inflammatory processes of the orbital venous system
      - Orbital venous thrombophlebitis
      - Cavernous sinus thrombophlebitis
   3.3. Inflammatory processes of Tenon’s capsule

4. Primary orbital tumors
   4.1. Benign orbital tumors
      4.1.1. Choristomas
      - Dermoid cyst
      - Epidermoid cyst
      - Cholesteatoma
      - Lipodermoid
      - Congenital serous cyst
      - Optic nerve sheath cysts
      - Retention cyst
      - Teratoma
   4.1.2. Vascular benign tumors
      - Capillary hemangioma
4.1.3. Benign nerve tumors
4.1.3.1. Optic nerve tumors
- Glioma
- Meningioma
4.1.3.2. Neurofibromas
4.1.3.3. Benign tumors, bone
dysplasias and cartilaginous dysplasias
- Osteoma
- Aneurysmal bone cyst
- Fibrous dysplasia – Jaffe Lichtenstein syndrome
- Ossifying fibroma
- Orbital chondroma
4.1.3.4. Fibroid tumors
- Fibroma
- Myxoma and fibromyxoma
- Fibrous histiocytoma
4.1.3.5. Lacrimal gland benign tumors
- Cystic tumors
- Adenomas: - pleomorphic adenoma
- monomorphic adenoma

4.2. Malignant orbital tumors
4.2.1. Malignant ocular tumors
- Retinoblastoma
4.2.2. Malignant nerve tumors
- Schwannoma (neurofibrosarcoma)
- Malignant meningioma
4.2.3. Malignant vascular tumors
- Hemangioendothelioma
4.2.4. Malignant bone and cartilage
tumors
- Histiocytosis X – localized –
  eosinophilic granuloma
- acute diffuse – B. Lettere – Siwe
- diffuse chronic – B. Hans – Schuller –
  Christian
4.2.5. Malignant fibrous tumors
- Primary fibrosarcoma
- Secondary fibrosarcoma
4.2.6. Malignant lymphomas and
marrow tumors
- non-Hodgkin lymphoma
- Burkitt lymphoma
- Reticulosarcoma
- B. Waldenströmmacroglobulinemia
- Chloroma
4.2.7. Isolated malignant tumors
- Rhabdomyosarcoma

5. Secondary orbital tumors
- Orbital metastases
- Myoblastoma
- Ewing sarcoma
- Acute lymphoblastic leukemia
- Myeloid leukemia

Materials and methods
All these tumoral varieties are met in the pediatric, ophthalmologic and
neurosurgical practice, but the cases of optic nerve gliomas are the most frequent as they
are met in the statistics of the Neurosurgical Clinic in Iasi; out of a total
of 40 observations on orbital tumors, only
13 were registered in childhood and only 7
were gliomas.

Obs. 1 C.C. (f) 1 year and 11 months
Left irreducible axial exophthalmym. Left
frontal flap with the orbital roof resected.
The periosteum is cut and between the
external and the superior right, the ocular
globe is penetrated retrobulbarly. An
encapsulated slightly multiloculated tumor
is highlighted with the cortical veins
attached to the optic nerve. The retenent
tumor the size of a little plum is “gray
colored” (1.5 – 1.8 cm) and after the
electrocoagulation of the vessels a
incision is made in the capsule and a part of it
is removed with a curette. The tumor capsule
and the visible remains are
electrocoagulated. The hemostasis and
plane sutures are made.
The anatomopathological examination:
the tumor has a loose structure, consisting
of stellate little cells with round nuclei. The
optic nerve fibers are disparate in
the majority of the fragments and under the
form of fragments in the peripheral area.
The vessels are well represented. Their big
axe is oriented parallel to the nerve’s big axe. In some areas, the vessels have thickened hyaline walls surrounded by hyaline unstructured masses. The nerve sheath is infiltrated with tumoral tissue.

**Diagnosis:** left optic nerve glioma.

**Obs. 2, N.A. (f) – 3 years old.**

Moderate exophthalmos, partially reducible. Right fronto-temporal flap with the resection of the ceiling.

It is highlighted a large tumor the size of an olive that is connected to the optic nerve. The optic nerve is free and the neurosurgeon cuts behind the ocular globe.

Anatomopathological examination: around the nerve it is found a well organized sheath thickened outwards (perineuritis) which comprised thrombosed small vessels with perivasculitis, bigger arteries with the lumen disappeared and with established sclera periarthritis. In the nerve trunk, the doctor finds septal sclerosis with vascular wall sclerosis and involutional lesions of the fibers. On another fragment, besides the vast lesions of polynerytis, a vast area of necrosis in the nervous trunk is found. On the cross section the doctor observes a thickening of the septum, but without tumoral characteristics.

**Diagnosis:** right optic nerve glioma; Recklinghausen disease.

**Obs. 3, RA (f) 11 years**

HIC syndrome with decrease in AV AO, right fronto-temporal approach.

The frontal lobe is lifted on the line of the little wing and around the optic nerve it is found a milky gray thick blade which is well individualized by the arachnoid. This arachnoid sleeve is cut to find the optic nerve which normally measures 1-2 mm and now measures 7-8 mm, and the chiasma is also thickened to 10-12 mm. The surface is well vascularized. A biopsy is undertaken from the right lateral side of the chiasma in an area that is both avascular and very prominent. The intracranial pressure is slow. Hemostasis and plane sutures.

Anatomopathological examination: several fragments of nervous tissues with homogenized cells lesion, interstitial hemorrhage and discreetly infiltrated perivascular lymphocytic. Several fibrous astrocytes are found on one of the fragments.

**Diagnosis:** opto-chiasmatic glioma.

**Conclusion**

In neurosurgery, the orbital tumors in children have a series of particularities:
- In children, benign orbital tumors are more frequent than the malignant tumors.
- The most frequent orbital tumors in children are hemangiomas and disembioplastic tumors (epidermoid and dermoid).

Retinoblastomas are tumors typical of infants and children, they have an intraocular starting point and through invasion they occupy the orbit, they arrive at the intracranial portion through the optic nerve and chiasma (bilateralisation).

- Different types of sarcomas represent the malignant orbital tumors in children.
- The frequency of orbital tumors in children is 20% out of the total number of orbital malformations in all age patients. Hemangiomas are the most frequent followed by epidermoid and dermoid tumors, neurofibromatosis, meningioma and sarcoma.

**References**