IMAGING OF OCULAR MALIGN TUMORS IN CHILDREN

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SUMMARY: Retinoblastoma is the most common intraocular malignant tumor at children. Purpose. The aims of this study is to presenting the imaging methods used to diagnosis retinoblastoma and to establish the optimal method of pre and post surgery imaging exploration. Materials and methods. We have performed a retrospective five-year study, which included 15 cases (7 girls and 8 boys), between the ages 1-7 years. The patients were clinically examined and imaging by oculo-orbital ultrasound, unenhanced and enhanced CT and MRI examination. All cases have been confirmed anatomo-pathologically. Results and discussions. We studied 14 cases of unilateral retinoblastoma and 1 case of retinoblastoma with bilateral localization, associated with pineal gland tumor (“trilateral retinoblastoma”). We established various clinic correlations (according to age, sex, localization), as well as imaging correlations (according to the presence of calcifications, contrast enhancement, tumoral extension and metastasis). We found 6 cases of post surgery relapse, of which 1 case of gigantic relapse, 3 weeks after the surgery. There have been presented imaging method such as, the typical imaging findings, the contribution to the diagnosis, the advantages and the limitations of the method for each case. The differential diagnosis revealed by practice are also discussed. Conclusions. Retinoblastoma represents the most frequent intraocular malignant tumor of childhood. The oculo-orbital ultrasound allows for a supposed diagnosis. The correct diagnosis takes superior imaging methods (CT and MRI). CT represent the first-intention method, with a major role in evidence of calcifications. The CT aspect is a typical one: calcified mass starting from retina, projected in the vitreous. The MRI is superior, serving for the early detection of retro-ocular and intracranial tumoral extension. The MRI is preferred for the post surgery examination. Key Words: retinoblastoma, trilateral retinoblastoma, leukocoria, CT

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CLINICAL OVERVIEW:

Retinoblastoma—graphic illustration of the axial orbit delineates a typical retinoblastoma in the posterior globe. A calcified mass in a globe of an infant or young child is a retinoblastoma until proven otherwise.

Definition: Retinoblastoma is a malignant primary neoplasm of the retina.

Retinoblastoma arises from a cell of neuroepithelial origin, usually in the posterior retina. It is the most common malignant eye tumor of childhood and is responsible for 1% of all deaths from cancer in the age group of newborns to 15 years. Retinoblastoma can be multifocal and bilateral (30%); it undergoes spontaneous regression more frequently than other tumors; has a high incidence of second primary tumors and occurs as a congenital tumor.

Retinoblastoma affects all racial groups. It occurs in approximately 1:15 000 to 1:34 000 live births. The majority of cases are diagnosed before the age of four and the incidence declines with age. It affects males and females with equal frequency.

Retinoblastoma occurs in both familial (40%) and sporadic (60%) patterns. Familial cases typically develop multiple and bilateral tumors, although they may be unifocal and unilateral. Sporadic cases are always unilateral and unifocal.

The majority (70%) are unilateral. Bilateral retinoblastomas tend to present earlier (at an average of 15 months) than unilateral ones (24 months). Retinoblastoma serves as prototype for a group of cancers caused by recessive loss of suppressor cancer genes. These include osteosarcoma, Wilm’s tumor, hepatoblastoma, rhabdomyosarcoma, uveal melanoma, bladder cell carcinoma, acoustic neurinoma, and menigioma. Retinoblastoma and osteosarcoma arise after the loss of the same genetic locus (13q14 band).

Tumor cells may disseminate through the choroidal vascularisation or may spread beyond the eye through the optic nerve or subarachnoid space. In advance cases, the tumor may penetrate through the sclera and grow in the orbit. Metastases to the preauricular and cervical lymph nodes commonly follow over extraocular extension. The most common sites of distant metastases are the CNS, skull, distant bones, and lymph nodes. Spontaneous regression occurs in 1% of cases.

CLINICAL DIAGNOSIS

The diagnosis is usually made before age 2 when a white reflex from the pupil (“cat’s eye”) or leukocoria, strabismus, or eye pain is investigated. Ophthalmoscopy and CT scans are helpful.

Retinoblastoma - Imaging Diagnosis

Fundoscopic examination (Fig. 1)

![Fig. 1: Fundoscopic examination of retinoblastoma: endophytic form (a) and exophytic form (b).](image)
Ultrasound is 80% accurate at diagnosing retinoblastoma (Fig. 2). Typically, the tumor mass is hyperecogenic with calcifications.

**Fig. 2: Ocular ultrasonography:** solid mass with multiple hyperecogenic imagines with posterior attenuation projecting posterior into the vitros.

**CT** (Fig. 3)
- location: commonly at posterolateral wall of the globe
- tend to be solid, smoothly marginated retrolental hyperdense mass (endophytic type)
- most common cause of orbital calcifications (90%)- favorabil prognostic sign
- retinal detachment invariably present
- usually enhances with contrast - poor prognostic sign

CT is very effective at picking up calcifications, an important ability since few other intraocular lesions in patients less than three years old have calcifications.

CT demonstrates a solid retrolental hyperdense mass with associated retinal detachment. Associated findings include extraocular extension, optic nerve enlargement, and partial or complete calcification.

**MRI** (Fig. 4)
- iso- to mildly hyperintense on T1 relative to vitreous
- moderate to marked enhancement
- hypointense on T2
- subretinal fluid usually hyperintense on T1 and T2 (proteinaceous fluid)
- extraocular extension in 25%: optic nerve enlargement, intracranial extension, abnormal soft tissue in orbit

MRI is not as effective at detecting calcifications. However, MRI is better at evaluating intra and extracranial involvement, since it offers better resolution of adjoining structures. Additionally, MR is better at differentiating the different causes of leukocoria.

**Fig. 3: Axial CT** scan showing a retinoblastoma of the posterior part of the left ocular globe, like a solid mass with multiple calcifications inside and with contrast enhancement (a). Completely calcificated right retinoblastoma (b).

**Fig. 4: MRI** demonstrate a mass of intermediate signal, with diffuse areas of hypointense foci corresponding to the calcifications (a) located at the level of left ocular globe. The mass present contrast enhanced. Associated semilunar aria, without contrast enhancing, corresponding to retinal detachment.
Preoperative axial CT examination performed scan show a extensive mass what fill the right ocular globe with posterior extension, retroocular, including the optic nerve, but without intracranial extension. Note the multiple calcification within the intraocular tumor and the contrast enhancement (a).

After enucleation of right ocular globe, anatomo-pathological diagnosis was confirmed. Postoperation, at 3 weeks, we have to difference between postopertor hematoma or an immense recurrent retinoblastoma.

Clinical aspect postoperation (b).

Imaging explorations including: orbital echographie (c), CT with contrast (d) and MRI native and with contrast (e, f) show a immense recurrent mass witch including all the retrobulbar space with contrast enhancement, recurrent tumor is greater then the initial tumor.

Trilateral retinoblastoma (Fig. 6) - bilateral retinoblastoma and pineal gland mass (pynealoma, pynealocytoma, germinoma, teratoma) or suprasellar mass.

Association between a bilateral retinoblastoma(a) - where CT show calcified mass in posterior bilateral globe with pinealoblastoma (b). At the same patient MRI show a lobulated, well defined pineal tumor with intensity lower that of CSF in all sequence and with intense enhancement post Gd-enhanced. The tumor extend into the third ventricle and into the thalami.

Fig. 5: Recidive after retinoblastoma (3-year-old child male with leukocoria and III-th grade right exophthalmus).

Fig. 6: Trilateral retinoblastoma (4 - year- old male patient with bilateral leukocoria, headache, diplopia, strabismus)
MRI of the thoracic and lombar spine was performed and disclosed multiple dissemination metastatic intra and extramedullary (c).

**Differential Diagnosis** - leukokoria and/or retinal masses and calcifications:
- retinoblastoma
- persistent hyperplastic primary vitreous
- Coat’s disease
- toxocara canis
- retrolental fibroplasia
- Norrie disease
- retinal astrocytic hamartoma
- choroidal osteoma
- optic drusen

**Persistent Hyperplastic Primary Vitreous (PHPV)** (Fig. 7)
- persistence and proliferation of portions of fetal hyaloid artery and primary vitreous
- is rare, usually unilateral
- clinical: blindness, leukocoria, microphthalmia (small hypoplastic globe)
- may be associated with ocular malformations, optic dysplasia, trisomy 13, and congenital syndromes especially if bilateral

**ImAGING (CT)** - small deformed globe and lens as well as a small optic nerve
- enhancing cone-shaped central retrolental density extending from the lens to back of orbit, just lateral to optic nerve along remnant of hyaloid artery (a branch of the primitive dorsal ophthalmic artery which typically regresses during the last trimester of fetal formation)
- no calcification
- complications: chronic retinal hemorrhage and retinal detachment - can cause fluid-fluid levels

**Coat’s Disease (Retinal Telangiectasia)** (Fig. 8)
- primary vascular malformation of the retina (characterized by multiple abnormal telangiectatic retinal vessels)
- results in lack of blood-retina barrier that causes exudative retinopathy that fills the retina and subretinal space with a lipoproteinaceous fluid - secondarily causes retinal detachment
- is rare, unilateral in 90%, occurs in 6-8 year old (M:F = 2:1)
- on CT, dense vitreous without focal mass or calcifications

**Imaging explorations including:** orbital ultrasonography (c), CT with contrast (d) and MRI native and with contrast (e, f) show a massive mass that fill entirely the left orbit with heterogeneously content (multiple cyst components, solid mass and calcifications). Optical nerve is not identifiable inside. After contrast administration moderate enhancement at

![Fig. 7: Persistent hyperplastic primary vitreous (PHPV): hypoplasia of ocular globe (microphthalmos) (a). At the same patient associated retinal detachment show by ultrasonography (b).](image)

![Fig. 8: Coat’s disease: CT scan shows diffuse increased density of right globe due to total retinal detachment (a), without mass or calcifications. MRI show well retinal detachment.](image)
the level of solid components. The tumor is without intracranial extension.

Postoperative diagnosis anatomo-pathological was establish - teratoma.

CONCLUSIONS

Retinoblastoma represents the most frequent intraocular malign tumor of childhood.

The oculo-orbital ultrasound allows for a supposed diagnosis.

The correct diagnosis takes superior imaging methods (CT and MRI).

CT represents the first-intention method, with a major role in evidential of calcifications.

The CT aspect is a typical one: calcified mass starting from retina, projected in the vitreous.

The MRI is superior, serving for the early detection of retro-ocular and intracranial tumoral extension.

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