Imaging in leukocoria

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Learning objectives

We discuss the imaging findings of the most common causes of leukocoria in a child.
Background

Leukocoria is a white papillary reflex produced by any intraocular abnormality that reflects incident light toward the observer.

In a child with leukocoria, the major diagnostic considerations are retinoblastoma, persistent hyperplastic primary vitreous (PHPV) and retinopathy of prematurity (ROP).

We expose the imaging characteristics of the most common causes of leukocoria in a child.
Findings and procedure details

The most common causes of leukocoria are:

1. Retinoblastoma

Retinoblastoma is the most common intraocular tumor of childhood and the third most common intraocular tumor in all ages, but it remains a rare disease.

As the name implies, it resembles or arises from immature or primitive neuroectodermal cells that are destined to become retinal photoreceptors.

Retinoblastoma may be sporadic or inherited.

Rapid diagnosis allows for minimization of both morbidity and mortality, as when a tumor is caught in the earlier stages, the risk of mortality is lower and the possibilities for ocular salvage are greater.

On sonograms, the retinoblastoma is a hyperechoic mass inside the globe. Acoustic shadowing due to calcification is common. Orbital US may also demonstrate secondary retinal detachment.

On CT scans, retinoblastoma is recognized as a hyperattenuating intraocular mass. It may also contain punctuate or nodular calcification in over 90% of cases. The lesion may be smooth or irregular and may invade or fungate into the vitreous. However, it may be difficult to distinguish the neoplasm itself from the tumor produced subretinal effusion or detachment.

On T1 weighted MR images, the retinoblastoma appears isointense to slightly hyperintense relative to the vitreous (Fig.1). There is moderate to marker enhancement of the lesion on MR images obtained after administration of gadolinium (Fig.2). On T2 weighted images, the tumor tissue may be distinctly hypointense compared with the adjacent vitreous (Fig.3). Calcification, which is readily seen on CT scans, is difficult to visualize on MR images. Subretinal exudates are usually hyperintense on both T1 and T2 weighted images because of their proteinaceous fluid. It is much easier to visualize any extension if the tumor into the optic nerve or through the choroid beyond the margins of the globe on MR images than on CT scans. The use of gadolinium coupled with fat suppression T1 weighted pulse sequences has significantly improved detection of tumors and differentiation if them from subretinal effusions, even hemorrhage.

2. Persistent hyperplastic primary vitreous (PHPV)
PHPV is caused by the arrest of normal transformation and regression of embryologic vascular connective tissue within the globe. The basic lesion is a persistence of various portions of the primary vitreous and tunica vasculosa lentis (capillary vascular network covering parts of the lens), with hyperplasia and extensive proliferation of the associated embryonic connective tissue.

It is the second most common cause of leukocoria. It is usually detected at birth or at a newborn well-baby physical examination. There is no sex prediction and the process is usually unilateral.

Bilateral PHPV is usually associated with other ocular abnormalities and genetic lesions such as trisomy 13.

PHPV can be accompanied by vitreous hemorrhage, retinal detachment, microphthalmia, and opacified lens.

The CT findings include microphthalmos, which is usually detectable, although it may be minimal or absent, and other deformities in the globe configuration, which may have been undetectable by physical examination or ultrasonography. Calcification is absent within or around the globe. There is enhancement of abnormal intravitreal tissue with intravenous administration of a contrast medium.

Decubitus positioning may show a gravitational effect on a fluid level within the vitreous chamber, reflecting the presence of serosanguineous fluid in either the subhyaloid or subretinal space. The lens may be small and irregular, and the anterior chamber may be shallow.

On MR images, there may be an abnormal hyperintensity to the vitreous due to the presence of chronic blood degradation products or proteinaceous fluid. An S-shaped retrolental soft tissue mass that is hypointense to isointense may be seen within the vitreous or along the hyaloid canal on T1 weighted images. With the use of gadolinium contrast material, there will be moderate to marked enhancement of this abnormal fibrovascular mass within the vitreous, usually seen best in the retrolental region. Overall, the imaging hallmark of PHPV is the visualization of the hypertrophic tissue in the hyaloids canal. (Fig. 4, 5)

3. Retinopathy of prematurity (ROP)

ROP is a postnatal fibrovascular organization of the vitreous humor that usually leads to retinal detachment. It is almost bilateral, but can affect the eyes with striking asymmetry.

It usually develops in premature low-birth-weight infants who receive supplemental oxygen therapy after birth.
The early stage of ROP may have no specific CT and MR imaging findings except that the eyes may be microphthalmic.

In more advanced cases, the CT and MR differentiation of ROP and PHPV, retinoblastoma, and a number of pathologic conditions associate with retinal detachment may be very difficult.

The history of incubator treatment, birth weight, bilaterality, and the ophthalmoscopy, ultrasound, and CT findings are usually sufficient to establish the diagnosis.

Calcification is rare in ROP. However, in the more advanced stage, calcification may be present.

In the most advanced cases of ROP, both eyes are microphthalmic, with very shallow anterior chambers.

ROP may on occasion present as unilateral leukocoria. However, in the majority of cases, ROP is bilateral but often markedly asymmetric.

A persistent hyaloid vascular system may be an associated finding in patients with ROP, and the recognition of a massive persistent hyaloid vascular system on clinical examination, MR imaging, CT, or ultrasound is of prognostic importance. In these cases, the surgical dissection of the retrolental membrane in the presence of a persistent hyaloid vascular system is more difficult because these vessels tend to bleed, and the retrolental membrane is tightly adherent to the detached retina. (Fig. 6, 7, 8)

4. Coat's disease

Coat's disease is an idiopathic primary vascular anomaly of the retina that accounts for about 16% of the cases of leukocoria. It is characterized by telangiectatic retinal vessels, often with areas of aneurismal dilatation, and progressive deposition of intraretinal and subretinal proteinaceous exudates that leads to massive exudative retinal detachment (exudative retinopathy)

It is a congenital condition that is present at birth, but, the symptoms may be delayed.

Coats' disease usually occurs in young boys, with the onset of symptoms in most patients occurring before age 20 years.

The CT and MR imaging findings in Coats' disease vary with the stage of the disease.

CT is useful in later stages of the disease to demonstrate the retinal detachment. There may be an overall increased attenuation in the globe due to the density of the proteinaceous subretial exudates. True calcifications are rarely, if ever, seen in coats disease.
MR imaging is superior to CT in differentiating Coats' disease from retinoblastomas and other leukocoric eyes.

The subretinal exudation of Coats' disease is usually seen as hyperintense on T1-weighted, proton density, and T2-weighted MR images.

Gadolinium enhanced MR imaging plays an important role in the diagnosis as well as in the differential diagnosis of coats disease. Gadolinium enhances MR images demonstrate abnormal enhancement of the retina and the leaves of the detached retina enhance as well. The subretinal exudates; however, does not enhance, and no enhancing mass is present.

5. Other benign etiologies

Retinoblastoma must be distinguished from a host of benign etiologies that may simulate it.

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Table 1: Intraocular Mass and Mass-like Lesions Simulating Retinoblastoma
Images for this section:

Fig. 1: T1 weighted image: rétinoblastoma

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Fig. 2: T1 Gado weighted image: rétinoblastoma

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**Fig. 3:** T2 weighted image: rétinoblastoma

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**Fig. 4:** T1 weighted image: PHPV

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**Fig. 5:** T1 Gado weighted image: rétinoblastoma

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Fig. 6: T1 weighted image: Retinopathy of prematurity

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**Fig. 7:** T1 weighted image: Retinopathy of prematurity

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Fig. 8: T1 Gado weighted image: Retinopathy of prematurity

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Conclusion

US, CT and MR imaging play important roles in the diagnosis, pretreatment staging and post therapy follow-up of patients with leukocoria. By combining clinical information with the imaging findings, a specific diagnosis is often possible in cases of childhood leukokoria.
References


