Orbital CT and MR imaging in children: A pictorial review

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Learning objectives

A wide spectrum of pathologies may involve the orbit in childhood. With the clinical exam, imaging plays a major role in the etiological diagnosis, providing a precise analysis of the location of the lesion, its components, and its effects on adjacent and nearby structures. It studies tumor extension and often provides a good evaluation of its composition.

Ultrasound is proposed in case of anterior location of the mass or vascular lesion. But imaging is mainly performed with MRI. CT is needed when the lesion affects the orbital walls.

This paper presents a summary of the technical aspects of imaging. It presents the best sequences needed to depict specific pathologies and the aspects of the most common diseases encountered in child.
Background

Orbital masses have varied etiologies. Their clinical presentation is often nonspecific: exophthalmia, diplopia, decreased visual acuity, orbital pain, orbital inflammation... Imaging has a major role in the diagnostic process and is mainly based on MRI. It allows to specify the location of the lesion, its structure and its impact on the orbital components.

1- Technical aspects :

Orbital ultrasound is an atraumatic and relatively quick exam, and often associates Doppler. In tumoral pathology, it is restricted to superficial masses.

Orbital CT especially allows the analysis of orbital walls and any intralesional calcifications. It allows also the realization of positional maneuvers. Its limits are the characterization of the mass, the lesion extension mainly intracranial and the research for associated cerebral anomalies.

Orbital MRI has many indications. It allows analysis of the exact location of the lesion, its reports, the staging and the search for possible associated cerebral anomalies. the protocole includes T1 and T2 sequences, sequences with fat saturation and contrast injection, with exploration inthe three planes of space.

II- Etiologies :

1- Congenital affections :

a- Microphtalmos :

When bilateral and important, microphtalmos forms part of a more general pathological process and therefore ocular imaging has little interest. When unilateral, it represents a purely eye defect and imaging helps in etiological diagnosis.

A white reflection in the pupil (leukocoria) is suggestive of the diagnosis of persistent hyperplastic primary vitreous or PVP or PHVP (hypertrophic persistent hyperplastic primary vitreous).

Unilateral microphthalmia may be associated with coloboma or defective closure of the eye. Coloboma can be anywhere in the eye, so it has several severity. Pupillary coloboma
is a simple slot visible on the pupil without pathological consequence. Posterior coloboma reaches the papilla, then there is an excavation at the papilla instead of usual plan aspect. Retinal detachment may be associated.

On Ultrasonography, there is a fluid cavity in extension to the globe at the emergence of the optic nerve. The advantage of ultrasound is to confirm the clinical diagnosis but also to look for associated cystic malformations, mainly retro-ocular, which are not seen with the ophtalmologic examination. These cysts do not cause particular complication. Instead, they may occupy a place in the orbit and reduce the unaesthetic effects of microphthalmia on the growth of the orbit. They can also grow and cause diagnostic problems. These cysts are also explored in CT or MRI.

When is severe microphthalmia, plastics techniques are needed to correct the esthetic damage. It is in this indication that CT with 3D reconstruction can be useful (1).

b- Persistent hyperplastic primary vitreous (PHPV):

It is also called Persistence of fetal vasculature (PFV), representing the second cause of leukocoria in children. It is a congenital malformation, usually unilateral, which is usually associated with microphthalmia, a small crystalline lens with elongation of the ciliary apparatus.
Color Doppler ultrasonography should search persistence of Cloquet ligament, permeability of the hyaloid artery, retinal detachment and retinal hemorrhage frequently associated.
CT can note: a global hyperdensity of the vitreous, a discreet triangular structure connecting the intra vitreous macula to the crystalline and vitreous enhancement after injection of PDC.

On MRI, the vitreous appears hyperintense in T2-weighted images. After Gadolinium injection, it appears hyperintense in the anterior chamber and at the mass of the primary vitreous, adjoining behind the crystalline. Abnormal crystalline lens is also better visible in MRI (Figure 1 a, b, c, d).
An PHPV may be associated with other ocular anomalies (cataract, retinal dysplasia) or even retinoblastoma (1).

2- Tumors :

a- Retinblastoma :

Retinoblastoma is the most common malignant eye tumor in children. Its incidence varies from 1/15000 to 1/20000 births, with a sex ratio of 1.5 / 1. It is a neuroectodermal tumor developed from the retina. In about 60% of cases the tumor is unilateral with a median
age at diagnosis of 2 years and most of these forms are not hereditary. In 40% of cases, the tumor is bilateral and always with a hereditary autosomal dominant with variable penetrance discovered at a median age of 1 year (3, 4, 5).

The most common signs are leucocoria and strabismus (4, 6), other signs can also be seen: nystagmus when the tumor is bilateral, a buphthalmos, anisocoria, hyphema or vitreous hemorrhage. Inflammatory signs are sometimes encountered: uveitis, eyelid edema, conjunctivitis, and sometimes a panophtalmie or orbital cellulitis with proptosis.

Three forms exist: Endophytic form, Exophytic form and Infiltrant form.

Ultrasound is the first exam to be done after the clinical examination immediately after the ocular fundus. Ultrasound with Doppler should confirm the diagnosis by showing calcifications; essential diagnostic criterion as found in over 90% of cases, and assess the size of the tumor (2, 7). The tumor typically presents as a hyperechogenic mass, vascularized on color Doppler, with either intra vitreous development (endophytic form) or subretinal development (exophytic form) with retinal detachment almost constant; containing calcifications either in clods with posterior shadow or thin and punctiform. (7)

CT should be avoided currently because, apart from rare cases of macroscopic extension to the optic nerve with enlarged nerve, the sensitivity of this technique is insufficient to detect extensions. CT is also a radiating technology; and patients may have a higher relative risk of secondary sarcoma in irradiated zone (3, 7).

When performed: its main indication is the search for tumor calcifications, strongly suggestive of retinoblastoma (Fig. 2 a) (5, 6, 7).

MRI must be performed by secondary intention after ultrasound and shall specify the uni or bilateral lesions. It is the primary exam to assess the periocular extension the optic nerve and to intracerebral structures and to detect subarachnoid dissemination (5, 7).

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The tumor is discrete hyperintense compared to the vitreous on T1-weighted sequences and hypointense on T2-weighted sequences. Enhancement of the tumor after injection is variable, generally moderate and heterogeneous (Fig 2 b, c, d), seen better in subtraction (5, 7). Intratumoral calcifications are visible on MRI (hypointense on all sequences) (6). There is usually a retinal detachment associated variable importance occurring (3) hyperintense relative to vitreous on T1-weighted sequences and T2.
The extension to the optic nerve behind the cribriform plate (retro-laminar) is relatively rare (about 10% of cases). MRI criteria extension to the optic nerve are significant enlargement of the nerve and / or its meningeal sheath (fig.2 e, f).

b- Rhabdomyosarcoma:

This is the most common cause of primary malignant orbital tumor in children if ocular tumors are excluded and represents 4% of childhood cancers (8, 9).

Orbital rhabdomyosarcoma may be primary or secondary either by direct invasion of rhabdomyosarcoma from the nasopharynx, pterygopalatine fossa, infratemporal fossa or as a metastasis (8,10). The primitive orbital rhabdomyosarcoma is a tumor that usually begins around the age of 8 years (8). There seems to be a slight male predominance sex ratio 5/3 (8,9).

Its starting point can be extraconal (37%), intraconal (17%) or both (47%) (10) with an initial preferential involvement at the upper-nasal quadrant (67%). The main clinical sign is a non axile unilateral exophthalmos (71%), often with an inflammatory character, and ptosis with especially rapid progressive (8). Epistaxis may be associated, signing extension to sinonasal cavities.

Ultrasound found an ill-defined plate mass, heterogenous with low to medium echogenicity. Vascularization on Doppler is variable.

On CT, the mass is iso or hypodense compared to muscles. It can not be distinguished from the muscle which it is derived and can encompass several muscles. When the tumor is confined to the orbit, it appears as a homogeneous mass, fairly well defined isodense compared to muscles and highly enhanced after injection of contrast (Figure 3 a). CT appreciate associated bone lysis (Figure 3 b) particular in large tumors (40% of cases) and the intracranial extension or invasion of a facial sinus (8).

On MRI, the tumor is ill-defined, hypointense or isointense to muscle on T1 and hyperintense on T2, enhancing after gadolinium injection (Fig 3 c, d, e). The ocular globe is usually repressed but rarely invaded.

Rhabdomyosarcoma is an aggressive tumor, rapidly evolving, invading frequently orbital walls and the adjacent soft tissues (10). Tumor extensions are about 3 main directions:
- To the ethmoid labyrinth and nasal cavities (20% of cases).
- Beyond the orbital apex, to the cavernous sinus and middle fossa.
- To the infratemporal fossa.

c- Glioma of the optic nerve:
the glioma of the optic nerve and / or chiasm represents 1.5 to 3.5% of orbital tumors. It is a tumor of childhood, with a maximum frequency between 2 and 8 years with an incidence of 75% in the first decade and 90% in the second decade. It represents 66% of tumors of the optic nerve, almost entirely low grade IN pediatric age:
- 60% grade I: pilocytic astrocytoma
- 40% grade II: fibrillary astrocytoma (11).

The association with Recklinghausen disease is emphasized by all authors with variable frequency: 30 to 40% of patients with optic nerve glioma have Recklinghausen disease. The clinical features include ocular signs: a decreased visual acuity, strabismus or nystagmus. The exophthalmos, often revealing, is unilateral, axile, irreducible, painless, not accompanied by oculomotor palsy, evolves very slowly; endocrine and metabolic signs and intracranial hypertension syndrome (12).

Ultrasound shows in the case of orbital location, an fusiform increase in volume of the intraorbital optic nerve, which is often elongated and sometimes angulated, reflecting mass of the optic nerve, containig cystic areas and especially having sharp margins with retrobulbar fat.

On CT, the orbital walls and optical channels are thinned but respected (5, 11). The tumor is usually isodense extending the optic nerve sometimes with hypodense areas (cystic degeneration) without calcification (unlike meningioma), and usually enhancing discretely.

On MRI, the tumor is presented as iso or hypointense on T1-weighted images with variable signal intensity on T2-weighted sequences (intense if it has an mocoïde content, discrete if it has a fibrillar structure) with a variable enhancement after gadolinium injection and fat saturation (Fig. 4 a, b, c, d). Zones of hemorrhage or calcifications are rare. MRI is also the best examination for the staging searching chiasmatic and retro-chiasmatic extension. It also enables search for NF1 lesions (12).

d- Metastasis of neuroblastoma:

It is the first cause of orbital metastasis of the child. This is a tumor of the young child, often before the age of 2 years (13). The primary tumor is at the level of the adrenal medulla (90% of cases) or sympathetic chain ganglia which gives orbital metastasis in 8 to 20% of the cases. Clinically, unilateral beginning sometimes bilateral (in 40% of cases), is brutal, eyelid edema with inflammatory and often palpebral ecchymosis (25%) very evocative, realizing Hutchinson syndrome.
The primary tumor may be responsible also of two ophthalmologic signs: a syndrome of Claude Bernard-Horner (in the cervical and mediastinal forms) and opsoclonies -myoclonus (paraneoplastic syndrome). Clinical suspicion is confirmed by measurement of catecholamines in the urine (13).

CT (Fig. 5) shows a localized, homogeneous and regular mass, taking contrast (can mimic rhabdomyosarcoma). In case of bone involvement, there is a spiculated osteolysis or thickening of the roof or floor of the orbit. Sometimes, fine intratumoral calcifications (13, 14) are observed. A sphenoid bone erosion is suggestive of the diagnosis. The MRI determines the best extension through the coronal and sagittal oblique planes. The signal is variable depending on the fleshy or cystic nature. Usually iso T1 signal relative to muscle and moderate hyperintense T2. A moderate gadolinium enhancement is usually present.

e- Hemangioma :

Capillary hemangioma is the second most common tumor in children after dermoid cyst. It is a benign tumor vasculature, resulting in the proliferation of vascular endothelium. Capillary hemangiomas occur more frequently in premature infants or children of low birth weight with a slight female prevalence. It is located most commonly in the upper nasal quadrant, it is more frequently extra than intra conical and orbito-palpebral than palpebral.

Diagnosis is clinical, based on its evolution and its characteristic morphology (cutaneous and subcutaneous forms): it is a soft mass associated with a dilated vein and capillary drainage. Imaging is performed in cases of diagnostic uncertainty (deep form).

Ultrasound is the first exam to be performed. It shows a well-circumscribed heterogeneous compressible mass, with septa. Doppler examination revealed a hypervascular tumor involving immature vessels (index of low resistance, less than 0.6). A new sonographic classification was proposed for a better therapeutic effect:
# Stage I: pre-septal
# Stage II: pre-septal and extraconal.
# Stage III: pre-septal, and extra and intraconal.

CT evaluates the posterior retro bulbar extension, to the face and to the external temporal fossa, which allows to specify the limits of the hemangioma. It appears as an isodense mass, taking contrast homogeneously and intensively and respecting the bone (15).
MRI shows a hypointense mass in comparison to the extraocular muscles on T1, and hyperintense on T2, heterogeneous, with numerous serpiginous vascular structures, hypointense, realizing the "salt and pepper" aspect (flow void phenomenon). Most signal abnormalities correspond to blood flowing slowly into the capillary spaces. An intense and homogeneous enhancement after gadolinium injection is always present (Fig. 6).

f- Dermoid cyst:

The dermoid cyst is the most common child orbital tumor. It is a tumor occurring in young patients, 7% are discovered at birth, 50% are diagnosed the age of 15.

It is often asymptomatic, as a subcutaneous mass in the upper eyelid. It rarely manifests with exophthalmia. The most common location is extraconal and superolateral, above the lateral canthus or the tail of the eyebrow. (16)

Ultrasound is performed in case of diagnostic doubt. It finds an encapsulated mass, thick-walled, oval, well-circumscribed, often more mobile compared to the deep planes and devoid of vascularization within it, confirming its cystic appearance.

CT is often required in the deep forms. It shows a well-circumscribed oval cystic lesion with a wall sometimes with calcifications and taking rarely the contrast, and with a fatty density content in less than 50% of cases. When the cyst is in contact with the bone walls, erosion with signs of reconstruction (hyperostosis) may be observed. In rare cases, the dermoid cyst may be responsible for a further erosion of the lateral wall of the orbit and become bilobed, with expansion in the temporal fossa.

On MRI, the signal depends on the content of the cyst lesion and will be hypointense T1 if it contains keratin, hyperintense if it contains fat that attenuates on the fat saturation sequences. A fluid level is possible. On T2, there is usually an intense heterogeneous hyperintensity. The size is generally greater than or equal to 1 cm (15) (Fig. 7).

3- Pseudotumors:

a- Histiocytosis X:

Histiocytosis X is a disease of the reticulohistiocytic system that manifests by generalized or localized lesions. Histiocytosis X most often affects children and young adults, with a peak incidence between 1 and 4 years. The frequency of the disease is estimated at
about 5 per million children between 1 and 15 years per year with a predominance of male involvement.

This is a rare condition which constitutes about 1% of the pathologies of the orbit whose orbital involvement is stressed in 20% of cases.

Depending on the severity of the condition, there are three clinical forms:

* The eosinophilic granuloma:

It corresponds to the localized form of the disease, it is the most benign, with a generally favorable prognosis.

It occurs in the first decade and presents in the orbit as a single bone lesion, usually in superotemporal (predilection for the frontal bone and the greater wing of the sphenoid).

It results clinically by progressive unilateral or bilateral exophthalmos. When the granuloma is palpable, the mass has a soft consistency and can cause a local inflammatory reaction.

On CT, we observe an osteolytic mass with clear limits whose density is that of soft tissue (Fig. 8 a, b, c). It does not enhance. Sometimes a "grelot" image is observed consisting in a bone defect of regular contour with rupture of cortical bone containing a sequestration (13).

The MRI would be more sensitive than CT in intracranial histiocytosis, in particular the hypothalamic-pituitary location, in temporal fossa and in the anterior cranial fossa. The lesion appears hyperintense T2 with a soft tissue mass in 30% of cases. In T1, the lesion is isointense relative to muscle and enhances intensely after gadolinium injection (Fig. 8 d, e, f) (14).

* The Hand-Schuller-Christian disease (HSC):

It corresponds to the form of disseminated disease.

The clinical triad is characterized by lytic lesions of the skull, exophthalmia and diabetes insipidus.

We can find skin lesions, disseminated lymphadenopathy, lung involvement and sometimes an invasion of the bone marrow. Evolution is generally favorable, but the HSC can be lethal, especially in cases of bone marrow failure, liver or lung disease (11).
*Disease Letterer-Siwe:

It reaches the infant or the very young children under 3 years. It represents the diffuse form of Histiositosis X, characterized by multifocal with disseminated cutaneous, pulmonary and hepatosplenic lesions. The orbital and bone involvement is rare. The prognosis is poor. (11)

**b- Fibrous dysplasia:**

Fibrous dysplasia is a bone dystrophy of unknown etiology that preferentially affects adolescents and young adults. There are two forms:
- monostotic (80% of cases).
- polyostotic most often diagnosed in childhood because of the frequency of complications, and the association of extra-osseous abnormalities. It is not a hereditary disorder but can be associated with a Recklinghausen disease. The risk of malignant transformation is estimated at 0.5%, but can reach 15% after radiation therapy.

Craniofacial region is affected in 20% of cases. Orbital involvement is usually unilateral and corresponds to a monostotic form (affecting in order of decreasing frequency the frontal bone, followed by the sphenoid and ethmoid). Symptoms are related to the thickening of the bones concerned. It appears usually as a painless non axile and chronic exophthalmia and facial asymmetry.

The general examination should search for factors in favor to the McCune-Albright syndrom, associating polyostotic fibrous dysplasia, skin pigmentation and endocrine abnormalities (precocious puberty). According to the seat (long or flat bones), extent and degree of ossification of the fibrous tissue, it may take a variable appearance.

Plain radiographs and CT show:
- a thickening of the orbital walls with frosted glass aspect, dense and homogeneous, or a pagetoid aspect with alternating transparent areas and areas of hyperdensity.
- a narrowing slots, fissures and channels (Fig. 9).

In MRI, the whole process of fibrous dysplasia is heterogeneous with low signal on T1- and T2-weighted sequences. The degree of the low signal depends on the condensation process.
In addition, MRI analysis more accurately a possible impact on the optic nerve particularly in the sphenoidal forms. It also allows the differential diagnosis with orbital sphenopalatine meningioma (thickening and intense contrast enhancement of the dura). (15)

4- Inflammatory conditions :

Orbital infections represent more than half of primary orbital disease processes. The location of an orbital infection is described with respect to the orbital septum, as either preseptal (peri orbital) or postseptal (orbital). The orbital septum is a thin sheet of fibrous tissue that originates in the orbital periosteum and inserts in the palpebral tissues along the tarsal plates. The orbital septum provides a barrier against the spread of periorbital infections into the orbit proper. The distinction between periorbital and orbital processes is clinically important because postseptal infections are treated more aggressively to prevent devastating complications such as cavernous sinus thrombosis and meningitis.

a- Orbital cellulitis :

Periorbital cellulitis, which is defined as a preseptal process limited to the soft tissues anterior to the orbital septum, most commonly arises from the contiguous spread of infection from adjacent structures such as the face, teeth, and ocular adnexa. It also may arise from local trauma. Symptoms include swelling and erythema of the eyelids, chemosis, and, in severe cases, limitation of eye movement without proptosis. Cross-sectional imaging demonstrates diffuse soft-tissue thickening anterior to the orbital septum without abscess formation (16).

Orbital cellulitis is a postseptal infectious process most commonly caused by paranasal sinusitis, which spreads to the orbit via a perivascular pathway. Thus, bone destruction is not usually seen. The symptoms at presentation are similar to those of periorbital cellulitis; however, patients with orbital cellulitis also may present with proptosis. Visual acuity is usually maintained.

Ophthalmic ultrasonography, in skilled hands, may be a useful adjuvant for the rapid evaluation of preseptal versus postseptal involvement, as well as a useful modality for a follow-up examination. However, ultrasonography is limited in its ability to assess intracranial extension, the orbital apex, and paranasal sinuses.

MRI, especially postgadolinium-enhanced, fat-suppressed sequences, is useful for the detection of early inflammatory changes within the orbit. On MRI, an orbital cellulitis
appears hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences.

MRI is also useful for assessing intracranial extension of the infection into the cavernous sinus and for evaluating cavernous sinus thrombosis. DWI in MRI can help in the assessment of the optic nerves for developing ischemia or infarction, which can occur secondarily from orbital infections.

On CT scans, a preseptal cellulitis may appear as an area of increased density, with swelling of the anterior orbital tissues and obliteration of the adjacent fat planes. When the infection progresses, an increase in the density of the orbital fat may occur with gradual development of more discrete densities that, in turn, may progress to formation of an orbital abscess (16).

b- Sub-periosteal abscess (SPA):

Development of an orbital subperiosteal abscess is most commonly associated with ethmoid sinusitis. CT scanning is also usually the first imaging modality of choice to identify an SPA, which may be located just lateral to the lamina papyracea. Drainage of the abscess may be necessary to avoid a rapid elevation of intraorbital pressure and resultant visual impairment. Since the advent of antibiotics, intraconal abscesses secondary to paranasal sinusitis have become rare. They are now most frequently seen as a complication of a penetrating orbital injury, ocular surgery, or a metastatic process. Symptoms include marked proptosis, chemosis, ophthalmoplegia, and impaired visual acuity. Intraconal abscesses usually require surgical drainage. Additional complications of orbital cellulitis include thrombosis of the superior ophthalmic vein, the cavernous sinuses, or both; bacterial meningitis; epidural and subdural abscess; and parenchymal brain abscess.
Images for this section:

**Fig. 1:** Fig 1

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Fig. 2: Fig 2

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Fig. 3: Fig 3

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Fig. 4: Fig4

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**Fig. 5:** Fig 5

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**Fig. 6:** Fig 6

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Fig. 7: Fg 7

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Fig. 8: Fig 8

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Fig. 9: Fig 9

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Findings and procedure details

Ophtalmologic ultrasound, Orbital Computed tomography with and without contraste with multiplanar reconstructions and Orbital MRI with T1 and T2 sequences, sequences after fat saturation and injection of Gadolinium are used to perform exams.
Conclusion

Adequate knowledge of the anatomy of the eye and orbit is needed to correctly limit the differential diagnosis. Awareness and knowledge of orbital lesions in children is very important. Familiarity with imaging findings of these lesions will facilitate accurate diagnosis and help to avoid unnecessary surgery.
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