Orbital Tumors And Pseudotumors

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Learning objectives

To establish the importance of image findings in orbital tumors and pseudo tumors.

· To demonstrate computed tomography (CT) and magnetic resonance (MR imaging) appearance of various orbital tumors.

· To compare CT and MR in diagnosis of orbital lesions.
Background

Computerized techniques (CT and MR) allow precise delineation of orbital anatomy and abnormalities. Orbital tumors are well depicted by these methods, various examples are illustrated in our exhibit.

Magnetic resonance (MR) is an excellent choice for displaying high soft tissue contrast and multiplanar capability.
Findings and procedure details

1. TUMORS

1.1 Malignant tumors

1.1.1 Rhabdomyosarcoma

- Rhabdomyosarcomas are the most common mesenchymal childhood tumors. Incidence peaks at approximately 8 to 9 years of age, but is reported in all age groups. These tumors may present with rapid proptosis.

- CT (fig 1) shows moderately well defined to ill-defined margins, irregular shape, and mild-moderate contrast enhancement. Adjacent bony destruction occurs in 40%. Globe distortion and extension to the paranasal sinuses may also be seen. Regional lymph nodes are involved later in the disease course. Calcification is rare in untreated tumors.

- MR (fig 2) typically shows bright T2 signal, distinguishing rhabdomyosarcoma from other tumors such as chloroma (granulocytic sarcoma), lymphoma and metastatic neuroblastoma. MR may also better delineate the true extent of disease if surgical resection is planned.

1.1.2 Retinoblastoma

Retinoblastoma is the most common intraocular tumor of childhood. Retinoblastomas are tumors arising from the retina. They usually occur in children younger than 5 years of age and may be hereditary or nonhereditary. They may present with leukocoria in about half of patients.

Typical findings on non contrast CT are speckled calcification (Fig 3 a). MR is useful for evaluating extraocular and intracranial disease.

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 3 b, c, d), seen better in subtraction. Intratumoral calcifications are visible on MRI (hypointense on all sequences). There is usually a retinal detachment associated variable importance occurring 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 3 e, f).

1.1.3 Orbital Metastases (fig 4)
- Metastatic lesions are found more in the anterior orbit than the posterior orbit. Metastases may involve any structure in the orbit, including the intraconal or extraconal space, globe, extraocular muscles and bone. The most frequent metastases to the orbit are from: breast, lung, prostate, melanoma, carcinoid, GI, renal cell, neuroblastomas and rhabdomyosarcomas.

- CT is usually performed for orbital imaging, although MR has superior soft tissue contrast. Tumor pattern can vary from diffusely infiltrative with obscured anatomical landmarks to a focal well-defined mass. Orbital metastases from breast cancer are often diffuse and irregular growing along the rectus muscles and fascial planes. Scirrhoux (fibrotic) breast cancers are unique in their ability to produce enophthalmos and ophthalmoplegia. In these cases, the metastatic lesion is typically very T2 dark, reflecting its fibrotic nature. Metastases from carcinoid, renal cell carcinoma and melanoma tend to be circumscribed. All orbital metastases show some degree of MR enhancement.

**Metastasis of neuroblastoma** is the first cause of orbital metastasis of the child. It is the tumor of the young child, often before the age of 2 years. CT (Fig 4) shows a localized, homogeneous and regular mass, taking contrast. In case of bone involvement, there is a spiculated osteolysis or thickening of the roof or floor of the orbit. Sometimes, fine intratumoral calcifications 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.

1.1.4 **Lymphoma (fig 5)**

Lymphoma may present with unilateral or bilateral disease, and manifest in the orbit as the primary site or as part of systemic disease. Systemic lymphoma may involve the orbit in 1.5-5% of patients. Up to 75% of patients with orbital lymphoma may later develop systemic disease.

Lymphomas most commonly involve the superolateral aspect of the orbit, and bilateral disease is common. When multiple sites of disease are present, the intraconal space is most affected (85%). Lymphoma can involve virtually any part of the orbit, including the lacrimal gland, extraocular muscles, lacrimal sac, periorbital fat, and retrobulbar fat.

Though lacrimal gland involvement, it can manifest with clinical manifestation of lid swelling and a palpable mass is most common.

It may mimic orbital inflammatory mass. A more diffuse form with infiltration of the retroconal space effacing normal tissue planes may occur.

The typical presentation is low-grade proptosis with minimal pain. Malignant lymphoma of the orbit is typically B cell lymphoma of a non-Hodgkin type, arising from mucosa-
associated lymphoid tissue of ocular adnexa (MALTOMA). Suspicion of malignant lymphoma is increased in the presence of systemic disease, lacrimal duct or gland disease, and bilateral disease.

Lymphoid tumors typically appear as homogeneous, lobulated masses on CT and MRI. They mold around normal structures without deforming them, and typically do not erode adjacent bone. They may be well defined, or infiltrative in appearance. CT shows density similar to skeletal muscle or appears commonly as hyperdense contrast enhancing mass. MRI demonstrates homogenous, intermediate T1 and T2 signals, and homogeneous contrast enhancement.

1.1.5 Optic nerve glioma

Optic nerve glioma comprises 1.5-3.0% of orbital tumors, 0.6-7.0% of all intracranial tumors, 1.7-7.0% of gliomas, and 2-5% of gliomas in the pediatric age group. The peak incidence is in 2-8 year old but can occur at any age and has been reported at up to 79 years of age. Females are more affected than males.

There is a high association (12-37%) with type I neurofibromatosis.

They may be unilateral or bilateral. Bilateral disease strongly suggests neurofibromatosis.

The tumors are slow growing with periods of growth and dormancy.

MR is optimal for imaging optic nerve glioma. The classic finding of optic nerve glioma is sharply circumscribed fusiform thickening and tortuosity of the optic nerve. Optic gliomas are typically T2 hyperintense, and usually show some enhancement, though a wide range of signal intensities and enhancement patterns may be encountered. Diffuse involvement of the substance of the nerve differentiates optic nerve glioma from optic nerve sheath meningioma, which surrounds the optic nerve. Any part of the optic nerve may be involved, from the globe to the optic chiasm. Peripheral enhancement of chiasmatic gliomas represents extraneural growth of tumor into the subarachnoid space, mixed with gliomatous tissue and non enhancing central tumor. Optic nerve gliomas that extend to the chiasm through the optic canal form a dumbbell shape.

Calcification is rare in absence of previous radiotherapy. There is no orbital hyperostosis unlike meningioma.

1.2. Benign tumors

1.2.1 Capillary haemangioma

This is the most common childhood benign orbital tumor, is more common in females, appears within first 2 weeks post partum, is non encapsulated, and may be very large. It is typically appearing as a reddish macule in the first 6 months of life. A proliferative phase occurs up to 10 months, followed by a slow involution phase for up to 10 years.
typically sporadic, they may occur as part of a genetic syndrome such as PHACES syndrome.

Contrast enhanced MRI is the preferred modality for imaging hemangioma, although CT can also be considered if it is not possible to sedate the patient for MRI. The diagnosis is typically well-established clinically, but imaging is indicated to assess the extent of the lesion and mass effect on adjacent structures. With deep lesions, the clinical diagnosis may be challenging. Capillary hemangioma typically shows:

- lobular contour borders,
- bright T2 signal with T2 dark septa between lobules,
- fine internal flow voids,
- intense, homogeneous enhancement
- preservation of adjacent bone.

Capillary hemangioma can have an atypical appearance without all of these features. Nevertheless, if these features are not all present, then alternative diagnoses should be considered, including rhabdomyosarcoma. DWI may help distinguish these lesions in challenging cases, as rhabdomyosarcoma typically shows lower ADC and brighter DWI signal than hemangioma. CT may demonstrate bony orbital expansion or scalloping with rapidly growing lesions. True bone invasion and calcifications are rarely seen.

1.2.2 Cavernous haemangioma (fig 6)

These are the most common benign intraorbital lesions in adults and most commonly present in the 2nd to 5th decades. They typically present with painless slowly progressive proptosis and are mostly intraconal.

They have smooth margins, homogeneous in density, show uniform enhancement and are easily separated from the optic nerve and extraocular muscles.

The most frequent locations are the retrobulbar muscle cone, especially the lateral aspect of the intraconal space. However, a small minority (less than 10%) of these lesions are extraconal. Cavernous hemangiomas rarely bleed due to firmer texture from the surrounding support of rich fibrous tissue.

Cavernous hemangioma typically appears as a well-circumscribed intraconal mass. Although most lesions are ovoid or round, larger lesions have lobulated margins. Larger lesions will distort surrounding structures, as opposed to lymphoma which molds around structures. CT shows homogeneous soft tissue density, and may show small calcifications or phleboliths. MR shows isointense T1 signal, bright T2 signal, dark internal septations, and a dark circumferential rim that represents a fibrous pseudocapsule. The
same findings are seen with multiphase CT, though concerns related to the increased radiation exposure of multiphase CT make this technique relatively less desirable. Small lesions often show early, uniform enhancement. MR angiography and CT angiography are generally not able to identify the feeding vessels of a cavernous hemangioma, likely due to their small caliber.

1.2.3 Meningioma

Meningiomas may arise either primarily from the optic nerve sheath (arise from capillary cells of the arachnoid around the intraorbital or intracanalicular portions of the optic nerve) or periosteum of the orbital wall, or secondarily from the sphenoid ridge or tuberculum sellae or olfactory groove and invades the optic canal and orbit by extension between the dura and arachnoid of the optic nerve.

They comprise approximately one third of primary optic nerve tumors and more commonly present in middle age females.

Bilateral meningiomas are seen in neurofibromatosis type II.

Secondary meningiomas present with hyperostosis and expansion of the bones adjacent to the orbit. They may present with visual impairment secondary to compression of the optic nerve.

Primary meningiomas typically have a perineural location with enhancing tissue surrounding optic nerve. Presence of calcification helps confirm diagnosis.

The typical clinical presentation of optic nerve sheath meningioma is painless, gradual vision loss and proptosis in a woman between ages 30 and 50.

Visual disturbance in the affected eye is common. While some patients only suffer transient visual loss lasting a few seconds, others experience visual loss in a particular field of gaze. Optic neuropathy, proptosis and strabismus occur later.

The key imaging finding of optic nerve sheath meningioma is a homogeneously enhancing mass that surrounds the optic nerve. CT may show calcification. MRI typically shows homogeneous, intermediate T1 and T2 signals. The optic nerve may be in the center of the lesion, or may be eccentrically positioned. In some cases, the optic nerve will show abnormal T2 hyperintense signal, which is presumably related to chronic venous insufficiency.

Optic nerve sheath meningioma may produce an expansile mass. Alternatively, it may only form a thin sheet around the optic nerve, termed "en plaque meningioma", producing the classically described 'tram track' sign. In these cases, fat-suppressed post-contrast T1-weighted images in the coronal plane are essential in making the diagnosis. Any part of the nerve may be involved, and radiologic diagnosis can be challenging when only the
intracanalicular portion of the nerve is affected. Fat suppression is helpful in these cases, but can also be degraded by blooming artifact around a well-pneumatized sphenoid sinus. For this reason, coronal post-contrast images without fat suppression should also be considered.

2. PSEUDO TUMOURS

2.1 Infection

Orbital infections may originate from skin and eyelid disease, or be related to sinusitis. In the former, preseptal swelling is often seen, with intraorbital involvement later.

Sinusitis may be complicated by subperiosteal abscess formation, intraorbital extraconal inflammatory collections and cellulitis. Progression to cavernous sinus thrombosis, meningitis or sudden visual loss may occur.

CT and MRI are the main imaging modalities.

Chronic infection may result in mucocoeles which may present as orbital mass lesions with proptosis. These most commonly occur in the frontoethmoid sinuses.

2.2 Inflammation (fig 7)

Orbital inflammatory mass or orbital pseudotumor is one of the most common causes of an intraorbital mass. It is one of the most common causes of unilateral exophthalmos, with bilateral disease also common. The typical clinical triad is a patient with proptosis, pain and impaired ocular movement. Age of presentation ranges from 10 to 40 years commonly.

Typical radiological findings are contrast enhancing uveal-scleral thickening.

This may be isodense to slightly hyperdense on CT with moderate enhancement usual.

Involvement of the rectus muscles, obliteration of retrobulbar soft tissue planes, lacrimal gland, or optic nerve sheath may occur mimicking optic nerve sheath meningioma.

2.3 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. 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.

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).

* **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.

* **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.

3. **CT vs MRI in orbital imaging**
The two main methods of clinical orbital imaging are CT and MRI. Both are now widely used as primary orbital imaging techniques. CT and MRI however have different strengths and weaknesses in orbital imaging which affect selection of one or the other as the first choice, quite apart from considerations of availability and cost, with CT generally more widely available and cheaper.

**CT:**
- Provides quicker scans,
- Is able to image bone directly,
- Shows the presence of calcification better,
- Is the modality of choice in patients with suspected metallic orbital foreign bodies. Visualization of non metallic foreign bodies like wood is more problematic and both CT and MRI may have to be used.
- Provides isotropic multiplanar imaging which has increased its ability to localize the site of orbital lesions.

**MRI** has advantages over CT in
- Its superior soft tissue contrast,
- Its ability to image the orbit and intracranial structures free of beam hardening artifacts from the skull base/dental fillings,
- Its lack of ionizing radiation.

Selection of appropriate MRI imaging protocols and use of the correct surface coils is important depending on clinical question.

Use of gadolinium contrast enhancement and fat suppression aids in disease detection and characterization.

The main disadvantage of MRI is the requirement for patients to remain still due to longer imaging times, its higher cost, its inability to image bone or calcium directly, magnetic susceptibility artifacts, and claustrophobia in some patients.

**4. Diagnostic strategy**

Several approaches to the diagnosis of orbital pathology are in use. A common strategy is to localize the pathology to one of the defined compartments of the orbit. These have been described as the muscle cone, formed by the four rectus muscles, dividing the orbit into intraconal and extraconal compartments, with the optic nerve within the central part
of the muscle cone, and the extraocular muscles forming separate compartments. The extraconal compartment is bordered by the bony orbit and subperiosteal compartment.

The lacrimal gland and globe form the other compartments. The optic canal forms the apex of the pyramidal orbit, and the optic septum the base of the orbit anteriorly, with the lids forming the anterior compartment. Recently, some authors have attempted to further refine this framework by using anatomical location, bone and sinus involvement, content, shape and associated features to increase diagnostic specificity.
Images for this section:

Fig. 1

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Conclusion

CT and MR imaging are excellent, non invasive modalities for detection and diagnosis of orbital pathologies and MR is far superior to CT examination of the orbit.