PICTORIAL
IMAGING EVALUATION OF PEDIATRIC ORBITAL PATHOLOGIES

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The orbital pathologies commonly detected during the childhood period substantially differ from the lesions that arise in adult orbit. The advance in imaging modalities including computed tomography (CT) and particularly magnetic resonance imaging (MRI) might enable the radiologists and clinicians who would be involved in either medical or surgical care of orbital pathologies, to confidently establish a definite diagnosis prior to histopathologic examination. The purpose of this pictorial assay is to present relatively common paediatric orbital pathologies with regard to CT and MRI findings.

Keywords: Orbit; Magnetic resonance imaging; Computed tomography; Ultrasonography; Paediatrics

INTRODUCTION

Paediatric orbital pathologies significantly differ from the ones that are encountered in the adults. For instance; the congenital anomalies; tumours as retinoblastoma, rhabdomyosarcoma, and optic nerve glioma; vascular pathologies including persistent hyperplastic vitreus and lymphangioma arise particularly during the childhood period.

In the last few decades, substantially due to extensive and effective use of cross sectional imaging in the head and neck region, the diagnosis and differential diagnosis of orbital pathologies prior to histopathologic examination could be feasible. Several imaging modalities exist for the radiologic examination of the orbit: 1- Plain film is limited to reveal foreign bodies prior to magnetic resonance imaging (MRI). Poor diagnostic value and the associated radiation exposure are the main concern. 2- Ultrasonography (US) is a readily available, reproducible, and radiation free method that helps discern particularly intraocular lesions and intra orbital foreign bodies with the implementation of high frequency probes (10–12 MHz). However, due to poor penetration it may be challenging to exhibit beyond the globe. 3- Computed tomography (CT) is the most versatile cross-sectional technique as it is capable of revealing bony structures, metallic foreign bodies, calcifications, masses, and inflammatory pathologies with a high temporal resolution. The main shortcoming of the CT is radiation induced cataract, especially in paediatric patients, due to repeated radiation exposure to the head region and metallic streak artefacts caused by dental fillings may interfere to explicitly exhibit the lesions.[1] 4- MRI allows better depiction of the soft tissue components of the orbit at the expense of poor temporal resolution, so that the sedation may be necessitated below a certain age. In this pictorial assay, we present relatively common paediatric orbital pathologies from a radiological perspective for the radiologists and clinicians including ophthalmologists and neurosurgeons who would be involved in the management by illustrating and emphasizing on particularly MRI and CT findings with respect to anatomical compartments.

ORBITAL ANATOMY

The bony orbit is a conical structure that is formed by frontal, sphenoid, maxillary, zygomatic, ethmoid, palatine, and lacrimal bones. It contains globe; 7 extraocular muscles (rectus and levator palpebra superioris muscles); optic, and III, IV, V_2, VI cranial nerves, ophthalmic vessels including ophthalmic artery and vein; lacrimal gland, and orbital fat.

The soft tissue of the orbit may be divided initially into two major spaces: 1- Ocular space that contains globe and 2- Extraocular space that may be further separated into three compartments by rectus muscles; conal space that is comprised of rectus muscles; extraconal space that locates externally to the rectus muscles and is confined by the orbit bony walls; lastly intraconal space that locates between the rectus muscles and contains basically optic nerve and intraconal fat (Figure-1).

Figure-1: CT anatomy of the orbit. 1: lens, 2: globe, 3: optic nerve, 4: medial rectus muscle, 5: lateral rectus muscle, 6: intraconal space, 7: extraconal space.

A-Ocular pathologies
1-Congenital Disorders
Anophtalmia and microphthalmia as both rare developmental disorders of the eyeball that occur due to insufficient formation of the optic vesicle during the first 8 gestational weeks.[2] Anophtalmia is characterized as the rudimentary eye tissue without a formed globe and lens while microhtalmia is defined as the globe with an axial length two standard deviations below the mean for age.[3]

Severe microphthalmia, with an axial length <10 mm at birth or <12 mm after age one year, and anophtalmia may be challenging to differentiate (Figure-2).[4]

Figure-2(a): Axial CT image of 2-month-old girl reveals right-sided anophthalmia. Note the absence of a well-formed globe and lens unlike the left eyeball

Figure-2(b): T2-WI of 28-month-old boy reveals bilateral micropthalmic globe oculi

Coloboma, as a congenital protrusion of the globe typically affecting inferonasal aspect, results from defective closure of embryonic choroid fissure. It may be associated with various syndromes as CHARGE and central nervous system anomalies including corpus callosum agenesis and encephalocele.[5] Radiologically, coloboma often presents as bulging of bilateral posterior globe adjacent to optic disc (Figure-3).
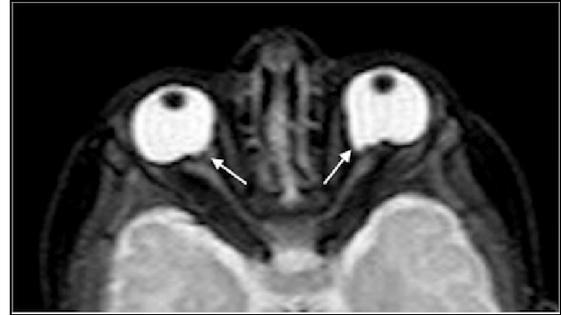

Figure-3: 7-year-old girl with bilateral colobomas. On T2-WI the colobomas located on bilateral posterior aspect of the globes medially to the optic discs are shown (arrows).

Persistent hyperplastic primary vitreus (PHPV) is a sporadic, congenital ocular disorder that is caused by the persistence and hypertrophy of the embryonic hyaloid vascular system and may be associated with microphthalmia, retinal detachment, subretinal or intravitreal haemorrhage.[6] Both CT and MRI reveal a cone shaped structure extending from the optic disc to posterior surface of the lens compatible with the persistent hyaloid artery that may enhance following contrast medium administration (Figure-4). Radiologic appearance of the vitreus may be variable due to either intravitreal haemorrhage or increased proteinaceous content.

Figure-4(a): T2-WI of 4-year-old boy reveals persistent primary hyperplastic vitreus of right globe. The right globe is seen to be microphthalmic with irregular contours

Figure-4 (b): On T1-WI, intravitreal increased signal, presumably related with hemorrhage, is seen on the right globe

Coast Disease implies congenital retinal telengiectasis that causes increased subretinal proteineous exudate and retinal detachment. Presentation in affected children is usually during the second part of first decade with leukocoria. On imaging, retinal detachment, on MRI T1 shortening, on CT increased attenuation of the vitreus, and rarely calcification may be detected.

2-Neoplasm

Retinoblastoma is the most common primary ocular tumour of childhood. Children usually present with leukocoria before the age of 3 years. Retinoblastoma is unilateral in two to thirds of the patients while inherited disease up to 90% arises bilaterally.[7] Since hereditary retinoblastoma is highly associated with radiation induced soft tissue sarcomas, MRI is recommended as primary imaging modality to evaluate and stage the disease of whom diagnoses have been previously established with fundoscopy and US.[8] The tumour manifests both T1 and T2 shortening compared to vitreus, moderate to apparent enhancement with contrast administration, and diffusion restriction with high b values (Figure-5). Gradient echo T2-weighted images (WI) are promising to exhibit the calcification. Retinoblastoma spreads into the brain and leptomeningeal space via optic nerve, so that fat suppressed post-contrast T1-W sequence allows ruling out the invasion of optic nerve. In order to evaluate intracranial extension and trilateral tumour that represents bilateral retinoblastoma and primitive neuroectodermal tumour of either pineal or supra-sellar region, routine contrast enhanced brain MRI examination should be included pre-treatment and during the follow up in addition to orbital imaging. On CT, the tumour is usually calcified, hyperattenuating, and enhances following intravenous iodinated contrast medium administration.
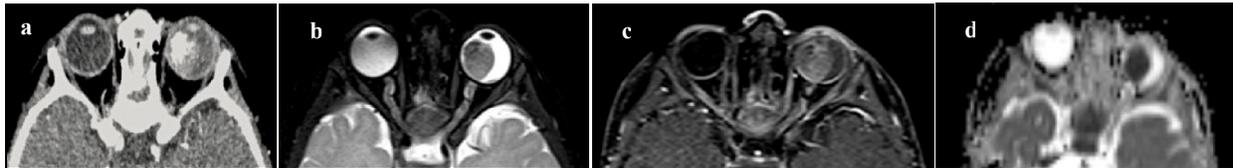

Figure-5 (a-d): Orbita CT of 9-month-old boy reveals enhancing and calcified solid mass within the left globe compatible with retinoblastoma (a). On T2-WI retinoblastoma is hypointense compared to vitreus (b) and is enhancing following contrast medium administration (c). Note the signal voids located on anteromedial part of the mass represents calcification on both T1 and T2 WI. Apparent diffusion coefficient map reveals low ADC value of the lesion (d).

Differential diagnosis list of retinoblastomas comprises other causes of leukocoria including PHPV, Coats disease, and toxocara canis. Presence of calcification and an enhancing solid mass help establish the diagnosis of retinoblastoma.

B- Extraocular Pathologies

1-Congenital Conditions

Neurofibromatosis (NF) type 1 is a multisystemic, genetic disorder that commonly involves orbit in affected individuals. The most common manifestations include optic pathway tumours; with a decreasing frequency, optic glioma and meningioma, plexiform neurofibroma, and sphenoid wing dysplasia. Plexiform neurofibroma is an infiltrative, nodular, and grape like soft tissue mass that usually originates from the sensory nerves and extends through the eyelids. Due to mass effect caused by the lesion, remodelling of the orbital bony structures and proptosis are closely associated. MRI is preferable to characterize and define the extent of the lesion; it appears hypointense on T1-WI, hyperintense on T2-WI, and variably enhances on postcontrast images. The dysplastic sphenoid bone is of clinic importance since it may lead to temporal lobe herniation into the orbit and pulsatile exophthalmos (Figure-6).

Figure-6 (a-c): MR images of 15-year-old girl, being followed up with the diagnosis of neurofibromatosis type 1, demonstrate a grape-shaped, nodular solid mass lesion of right orbit compatible with plexiform neurofibroma. The lesion has extraconal location and extends towards the eyelid. It's hyperintense on T2-WI (a), hypointense on T1-WI (b), and moderately enhances on postcontrast image (c). The slight dysplasia of right sphenoid bone causing proptosis of right eye is also exhibited.

Dermoid and epidermoid cysts are the most common congenital cysts of the orbit that originate from the aberrant ectoderm. They both usually present during the childhood as slow growing subcutaneous masses located adjacent to frontozygomatic and frontoethmoid sutures. On imaging, despite their similar unilocular cystic appearance, diffusion restriction of epidermoid cyst detected on diffusion weighted MRI and fatty content of dermoid cyst detected on CT help distinguish them from each other and other cystic lesions.

Septooptic dysplasia (De Morsier Syndrome), as a rare congenital disorder, is defined with any combinations of abnormalities in central neuro-structures including absence or hypoplasia of septum pellucidum and/or agenesis of corpus callosum, optic nerve hypoplasia, and hypothalamic-pituitary axis abnormalities.

2-Vascular Disorders
Capillary haemangioma is the most common orbital vascular malformation of childhood. While the diagnosis is made clinically; typical location in the upper eyelid, existence usually following birth, and gradually involution; MRI is the imaging modality of choice to show the extent of the lesion. On MRI, capillary haemangioma has lobulated contour, signal voids and exhibits hypointensity on T1-WI, hyperintensity on T2-WI, and enhances vividly on postcontrast images.

Lymphangioma (venolymphathic malformation) of the orbit, albeit relatively a rare location compared to other sites in head and neck region, commonly presents with proptosis during early childhood following either sudden haemorrhage into the lesion or upper respiratory infection.[9] It's infiltrative and may involve any compartment of the orbit. Orbital US helps reveal multicystic nature, however MRI is appropriate to show the extent and establish the definite diagnosis. MRI shows fluid-fluid levels and the signal characteristics vary depending on the proteinaceous content and timing of the haemorrhage. Following contrast medium administration, in the absence of any venous component, solely the septations enhance. On CT, lymphangioma typically appears as poorly defined and hyperattenuating mass lesion (Figure-7).
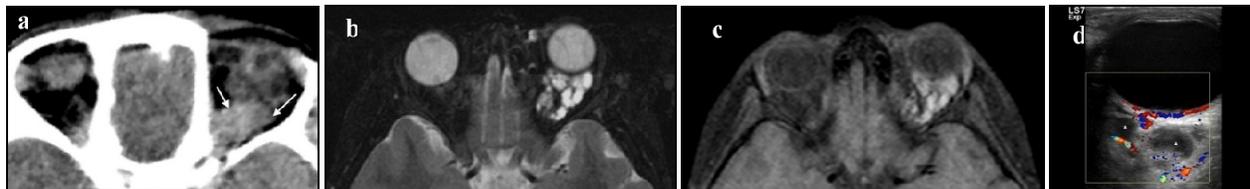

Figure-7 (a-d): 9-year-old girl with lymphatic malformation of left orbit. The CT image (a) reveals a hyperattenuating mass lesion within apex of left orbit (arrows). On T2 (b) and T1-WI (c) the lesion is shown to occupy both intra- and extraconal spaces with T2 prolongation and T1 shortening that may reflect either proteinaceous content or haemorrhage. Colour Doppler US image (d) depicts multiloculated cystic nature of the lesion located posterior to globe (asterix).

3-Neoplasms of the Orbit
Rhabdomyosarcoma (RMS) as the most common extraocular tumour arises in the children with a mean age of 6–8 years.[10] Patients usually present with abrupt proptosis due to rapidly growing nature of the RMS. MRI is the preferred imaging modality in order to assess prior to the treatment or residual/recurrent tumour during the follow-up. Since RMS is associated with bony invasion in up to 40%, CT may be complimentary to exhibit the bone involvement. Compared to muscle, the tumour appears hypointense on T1- and hyper-intense on T2-WI that is beneficial to distinguish RMS from other intra-orbital tumours including metastatic neuroblastoma, lymphoma, and granulo-cystic sarcoma (Figure-8). It shows moderate to marked enhancement on postcontrast images and low apparent diffusion coefficient values help differentiate from benign tumours as hemangioma.
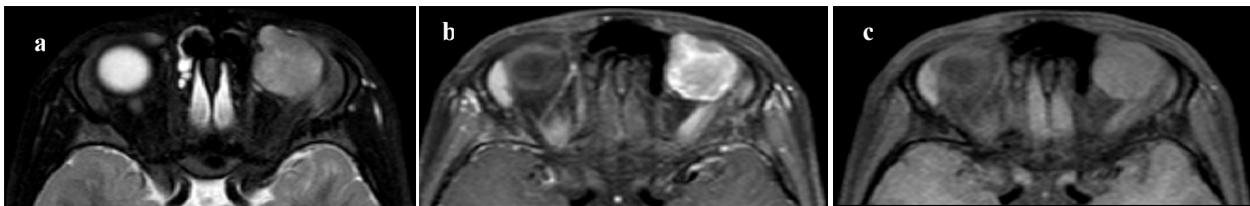

Figure-8 (a, b): The orbita MRI of 5-year-old boy diagnosed with orbital rhabdomyosarcoma. A well-defined solid mass in the left orbit superior to globe is seen. Compared to grey matter, the lesion is hyperintense on T2-WI (a) and enhances vividly on postcontrast T1-WI (b). The diagnosis of rhabdomyosarcoma was confirmed with histopathologic examination.

Optic nerve glioma (ONG) is associated with NF type 1 in up to 50% of the patients.[11] These tumours are bilateral and slow growing in the presence of NF type 1 with a common histologic subtype of pilocytic astrocytoma. Although orbital part of the optic pathway is commonly involved, MRI is optimal for the better delineation of the tumour located in any site. Involved optic nerve reveals fusiform enlargement and tortuosity with increased signal intensity on T2-WI and variable contrast enhancement. The foremost element of differential diagnosis list is optic nerve sheath meningioma despite its rarity among paediatric population. Unlike ONG, meningiomas involve 'nerve sheath' rather than 'nerve itself', may exhibit plaquelike calcifications; also, named as tram-track calcifications; vivid enhancement on postcontrast images, and hyperattenuation on CT (Figure-9).
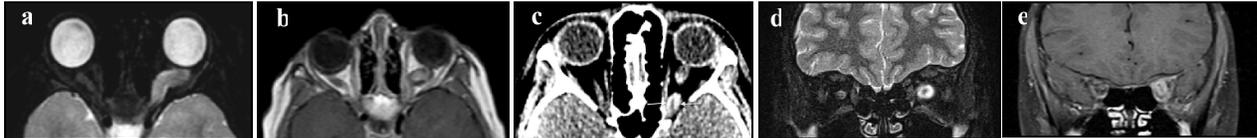

Figure-9 (a-e): Orbita MRI of 3-year-old boy followed up with the diagnosis of NF type 1 shows enlargement and tortuosity of intraorbital segment of left optic nerve compatible with optic nerve glioma. The lesion is hyperintense on T2-WI (a) enhances following contrast medium administration (b). The CT of 12-year-old girl with the diagnosis of optic nerve sheath meningioma reveals tram-track calcification of left optic nerve (arrows) (c). On coronal MR images of the same patient, optic nerve sheath is hyperintense on T2-WI (d) and enhances intensely on postcontrast T1-WI (e).

Secondary orbital tumours of childhood are comprised of granulocytic sarcoma that may occur as an initial sign of leukaemia and neuroblastoma, most common paediatric solid tumour that metastasize to the orbit. On CT, they both appear as hyperattenuating solid masses. Neuroblastoma has a predilection for the posterolateral wall of the orbit while granulocytic sarcoma may locate either in intra- or extraconal compartments. On MRI, neuroblastoma is seen isointense while granulocytic sarcoma is slightly hypointense compared to grey matter on both T1- and T2-WI. Marked enhancement is exhibited on postcontrast images (Figure-10). Clinical history of the patient, histopathologic examination of the lesion, and bone marrow aspiration may help in order to distinguish these two entities and establish definite diagnosis.
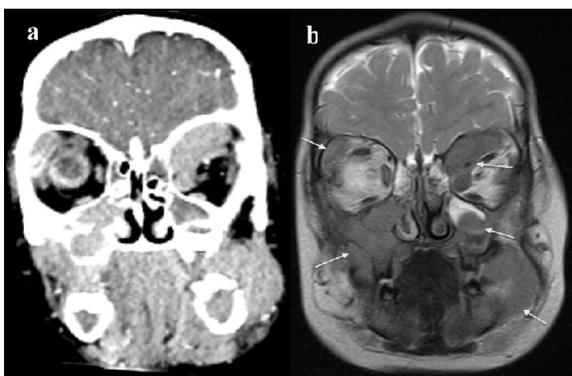

Figure-10 (a,b): Bilateral orbital granulocytic sarcoma in a 2-year-old girl with the diagnosis of acute myeloblastic leukaemia. On coronal CT (a) and T2-WI (b) multiple masses involving orbits, maxillary sinuses, and left angulus mandible are seen (arrows). The masses are hyperattenuating on CT while they exhibit hypointensity on T2-WI compared to grey matter.

4-Infectious and Inflammatory Disorders

Orbital infection may be categorized as preseptal and postseptal with respect to orbital septum as an anatomical barrier between eyelid and deeper structures of the orbit. The sinusitis constitutes the main source for the infection. CT and MRI are implemented for the evaluation of extent of the infection and the complications. Orbital abscess can be distinguished from other infectious processes with the features of fluid collections with an enhancing capsule, diffusion restriction on DWI, and gas content (Figure-11).
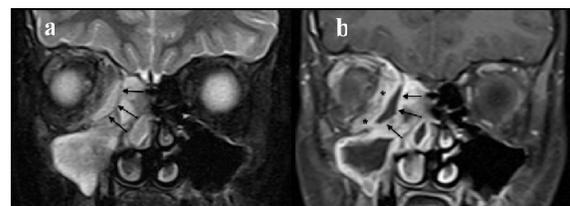

Figure-11 (a,b): 10 year-old girl presents with subperiosteal abscess of right orbit associated with same sided ethmoid and maxillary sinusitis. On T2-WI (a) subperiosteal abscess is shown adjacent to lateral wall of the ethmoid cells that has capsular enhancement on postcontrast T1-WI (b) (arrows). T1-WI image also shows the enlargement and marked enhancement of medial and inferior rectus muscles that represents infectious-inflammatory changes (asterix).

Optic neuritis implies the inflammatory condition of the optic nerve that may be associated with various etiologic factors including viral infections, autoimmune disorders, and demyelinating pathologies as multiple sclerosis and neuromyelitis optica. The clinical presentation is

constituted of sudden visual loss and pain with eye movements. Following ophthalmologic examination, MRI is the imaging technique of the choice to establish the definite diagnosis, show the extent of the optic nerve involvement, and accompanying demyelinating plaques in the brain. Fat suppressed sequences are preferable to reveal enlargement and increased signal of optic nerve on T2-WI and enhancement on postcontrast images.

Pseudotumor orbitale (idiopathic orbital inflammatory syndrome) is a nonspecific inflammatory disorder that may involve any structure of the orbit. In the common setting of sudden, painful proptosis, MRI helps reveal the inflammatory alterations affecting any or combination of preseptal region, lacrimal gland, extraocular muscles, orbital fat, optic nerve, and orbital apex. Tendinous involvement of the extraocular muscles is the best diagnostic clue to differentiate from Graves opthalmopathy.
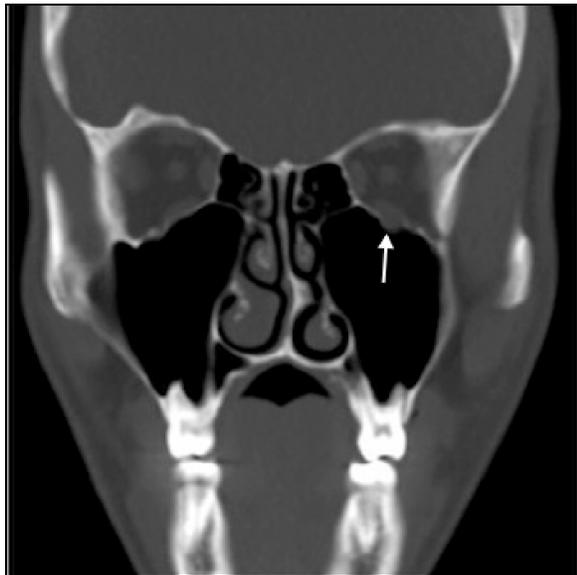

Figure-12: Left orbital trap door fracture in a 12-year-old boy following an assault. Inferior rectus muscle is partially entrapped (arrow).

5-Trauma Associated Disorders

Until age of 6 years' children are prone to orbital roof fractures whereas orbital floor fractures has a higher incidence in school-age children.[12] Trap-door fractures, as the most common type of floor fractures in children, is hinged medially allowing the orbital content get through the fracture and impinges either orbital fat or extaocular muscle. Multiplane CT scan, particularly in coronal plane, in clinically suspected patients enables to reveal the fracture and trapped soft tissue (Figure-12).

Conflict of interest: The authors state that there is no conflict of interest.

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