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Original Article

Orbito-ocular Space Occupying Masses in Children: High Resolution CT Findings in Yemeni Patients.

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Abstract

Intra and extra orbital space occupying lesions that present with orbito-ocular manifestations in children are frequently encountered in CT department. . In this prospective study, the high-resolution CT appearances, frequency and distribution of orbito-ocular space occupying lesions in Yemeni children were described. For a period of two years 60 children with various orbito-ocular space occupying masses underwent high resolution axial and coronal (or coronal reconstruction) CT cuts (1-2mm.), using spiral CT.

The diagnosis was based on CT criteria correlated with histopathology and post-operative results. Vision defect and proptosis were the most common clinical presentations. The findings included; primary malignant masses (30%), secondary invading and metastases (10%), benign cysts masses (21.7%), Vasculogenic masses (15%), optic nerve and meningial masses (10%), inflammatory masses (11.7%), and (1.6%) for osseo- fibrogenous masses. Almost all patients underwent surgical management in the form of exisonal biopsy, incisional biopsy, enucleation or exenteration. Retinoblastoma and Rhbdomyocarcoma appear more common in Yemen compared with that reported by western studies, and delay in presentation found to be the major cause of orbital invasion. HRCT provides excellent information about nature, extent, staging and proper management planning of orbito-ocular lesions in children.

Key words: orbital lesions, children, HRCT, Yemen.

Introduction:

The pediatric patient with an orbital mass lesion differs substantially from the adult patients with a much greater incidence of congenital lesions, higher frequency of infection and unique benign and malignant tumor involving the orbit and its contents. The structure complexities of the orbit and its contents present imaging challenges. In addition, orbit and eye proper may be involved by spread from adjacent structures or from lesion affecting visual pathway. However, high resolution CT has become the preferred modality for imaging orbital pathology (1-4).

The purpose of our study is to describe the high resolution CT appearances, differential diagnosis, frequency and distribution of the orbito-ocular space occupying masses in Yemeni children.

Materials and Methods:

The present work is a prospective study that included 60 patients aged 1 month to 17 years (34 males and 27 females) with orbito-ocular space occupying lesions. The patients were referred to the CT department in three large referral centers, namely Kuwait University Hospital (Sana'a), Al-Thawrah General Hospital (Sana'a) and Sana'a Radiology Center (Sana'a) between January 2004 to January 2006. Visual acuity impairment, proptosis and muscle movement disorder are the most common presenting symptoms. All patients underwent HRCT study for the orbital region with contiguous thin section (1-

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2mm.) and high spatial resolution alghorithm using Semens somtom plus 4 spiral CT machines. Both axial and coronal (or coronal reconstruction) planes prior and after administration of IV contrast were also obtained. Non ionic low osmotic contrast medium, iopromede 240 (ultravest) was used for all patients with 1.5ml/ kg dose in addition, soft tissue as well as bone window images from all patients were also obtained. Additional routine CT examination of the skull was performed to evaluate the extension of the orbital masses and to search for extra orbital lesion. Houncefield unit measurement was obtained on the lesion on both sets of scan and other CT findings were collected and analyzed. All patients were subjected to clinical eye examination before application of CT included test for visual acuity and measurement of visual fields apart from exophthalmetry and fundoscopy. More than half of the patients underwent surgery in the form of mass resection, enucleation or exenteration.

The diagnosis was based on the CT findings and correlated with clinical and surgical findings as well as histopathological reports.

Results:

This study included 60 children who were diagnosed by HRCT as having orbito-ocular space occupying mass lesions. The patients ages ranged from 1 month to 17 years with slight male predominance, (male 56% and female 44%). The clinical presentations included visual acuity impairment 79%, proptosis 75%, pain 43%. blured vision 40%, red eye 19% and leukokoria 14.8%. Table 1 illustrated the HRCT findings of orbito-ocular space occupying masses, their nature, categories, percentage, male to female ratios and median ages. The classifications according to the origin of occulo-orbital lesions were depicted in Table 2. Table 3 showed classifications of orbito-ocular lesions according to the age groups the overall frequency of malignancies was 40% (24 of 60): 30% were primary tumors and 10% were secondary invading and metastasic. Retinoblastoma was the commonest intraocular malignancy in our study. It accounts for 40% of all malignancy in our finding. All patients of retinoblastoma presented with leukokoria and loss of vision. HRCT reveals a dense soft tissue mass with extensive calcifications involving the retina

(Fig.1). Table 4 demonstrated the staging of the retinoblastoma. Rhabdomyosarcoma accounts for more than 30% of malignant tumors in our findings and it was the most common extra-ocular orbital malignancy in children below 6 years of age in this study. Fig (2-3) demonstrate the CT appearances and extensions of the rhabdomyosarcoma. Majority of rhabdomyosarcomas presented in late stages (Table 5). Only 3 patients were operable(table 6) whereas the remainder 5 patients were only biopsied and all the eight patients of rhabdomyosarcoma were sent for chemotherapy and radiotherapy.

Metastatic lesion was encountered in 3 patients two of them with bilateral affection and proved to be of primary neuroblastoma. The remainder of orbital malignant lesions included 2 cases of non-Hodgkin's lymphoma and one case for each of leukemic infiltrates and teratoma. (Fig. 4).

Among the benign tumors of retrobulbar region the optic nerve glioma (Fig.5-6) was seen in 4 patients whereas vascular tumors were mostly represented by capillary hemangioma (6 cases) (fig.7-8) However, shwanoma, meningioma and ossifying fibroma represented by one patient for each, who aged 15,14 and 13 years respectively (fig.9-11)

Benign cystic lesions in children form an important finding in our study. Apart from orbital abscess, dermoid cyst accounts for 30% of cystic lesions in our findings. They were located in between globes and periosteum supero-medially (Fig 12). One case of hydatid cyst was also found with retrobulbar localization and compression of orbital content (F1g.13) Three patients were found to have mucoceles extended from frontal and ethmoidal sinuses (Fig.14). 2 patients of orbital meningocele and meningoencephaloceles were seen reaching the orbit

through naso -ethmoidal and superior orbital fissure (Fig.15).

Inflammatory mass lesions were found in 7 patients, 6 of them having abscess, as sequel to neglected orbital trauma (in 2 patients) or extended from periorbital infection (in 4 patients) or post surgery (1 patient). It appeared in CT (Fig16) as hypodense centers with enhancing rims, however lesion in Fig 17 shows mycotic infection (Aspergillosis) in an immune-compromised patient. Biopsy was performed for inflammatory lesions to exclude other possibilities. Biopsy was proven in 76 % of orbito-ocular lesions. The sensitivity of was 100% and specificity was about 74%.

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Table (1) CT findings of the orbito-ocular lesions

Category	**Number**		**Percentage**	**Sex M/ F**	**Median Age(year)**
Malignant Masses	**24**		**40%**		
Retinoblastoma	9		15%	5/4	2.6
Rhabdomyosarcoma	8		13,3%	4/4	8.5
Non-Hodjkin's Lymphoma	2		3,3%	0/2	6
Teratoma	1		1,7%	1/0	3
Metastasis and leukemic infiltrates	4		6,7%	2/2	9
Benign Cystic Masses	**13**		**21,7%**		
Dermoid	7		11,7%	4/3	8
Hydatid	1		1,7%	1/0	13
Mucocele	3		5%	2/1	7
Meningocele	2		3,3%	1/1	1
Vascular	**9**		**15%**		
Capillary Hemangioma	6		10%	3/3	
Lymphangioma	2		3,3%	1/1	2
Varices	1		1,7%	1/0	5
Optic N.Sheath and Meninges	**6**		**10%**		**12**
Glioma	4		6,6%	3/1	
Meningioma	1		1,7%	1/0	8
Shwanoma	1		1,7%	1/0	14
Inflammatory	**7**		**11,7%**		**15**
Abscess	6		10%	2/4	
Aspergilosis	1		1,7%	1/0	4
Osseofibrogenic	**1**		**1,6%**		12
Ossifying Fibroma	1		1,6%	1/0	
TOTAL	**60**		**100%**	**34/26**	**13**

Table (2) Classification of the orbito-occular lesions according to the origin

Category	**Number**	**Percentage**
Malignant Lesions	**24**	**40%**
Primary malignant	18	30%
Secondary invading and metastatic	6	10%
Benign Lesions	**36**	**60%**
Primary orbital	23	38,4%
Extending from sinuses	5	8,3%
Extending from anterior cranial fossa	3	5%
Extending from periorbital bony and soft tissue structures	5	8,3%

Table (3) Classification according to age groups

Category	**0-5 years**	**6-10 years**	**11-17years**
Malignant lesions	**11**	**10**	**3**
Primary orbito –occular	9	7	2
Metastasis and secondary invading	2	3	1
Begin lesions	10	15	11

Table (4): Staging of Retinoblastoma according to Reese-Ellsworth classification

Group	I.	II.	**III.**	**IV.**	**V.**
Number of patients	**-**	**1**	**4**	**2**	**2**

Table (5) Staging of Rhabdomyosarcoma according to inter group Rhabdomysarcoma study (IRS)

Group	**I.**	**II.**	**III.**	IV.
Number of patients	**-**	**2**	**3**	**3**

Table(6) Type of surgical management

	Excisional biopsy	**Incisional biopsy**	**enucleation**	**exenteration**
No(%)	**32**	**23**	**4**	**1**

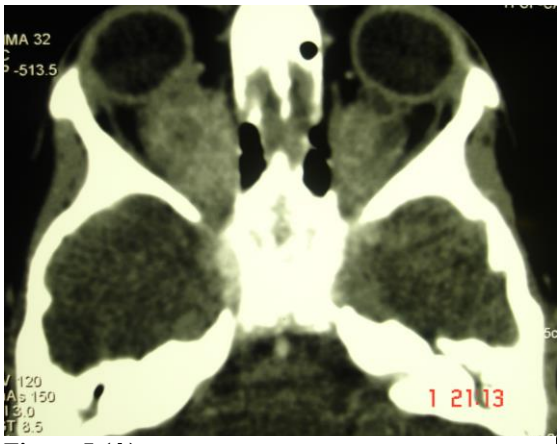

Figure 5 (A) Optic nerve glioma HRCT axial cuts of an 8 year old girl show markedly enhancing prominent fusiform enlargement of both optic nerve sheaths. Extending intracranially(B)

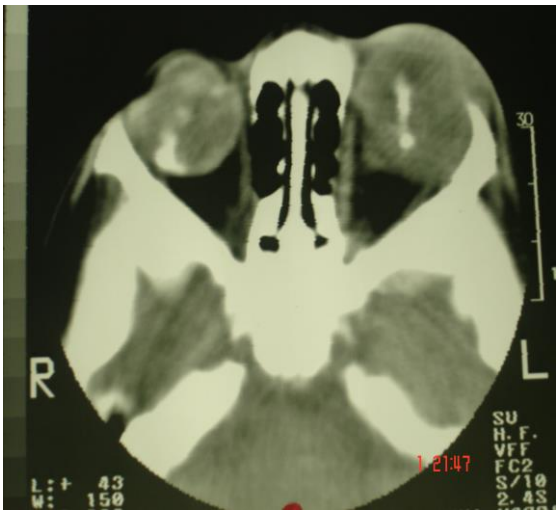

Figure 1(B)
Another patient aged 2 years with bilateral retinoblastoma with extension outside the globe .

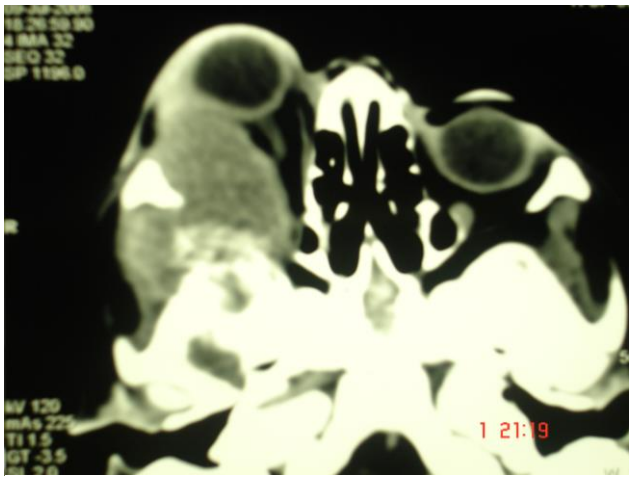

Figure 2 (A) Rhabdomyosarcoma.
The axial CT view in a 3 years age girl demonstrates large markedly enhancing right lateral retrobulbar soft tissue mass with bone destruction and extra-orbital extension.

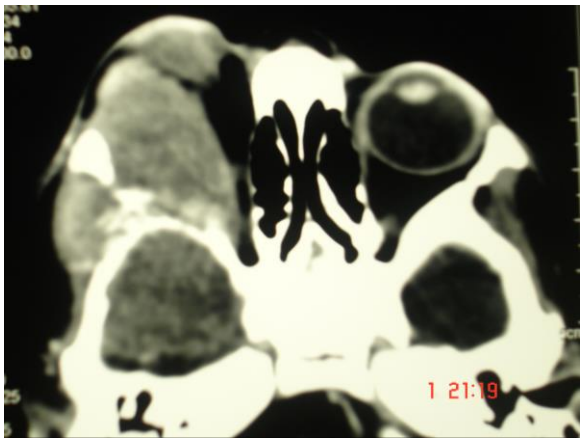

Figure 2 (B)Rhabdomyosarcoma with intra cranial extension.

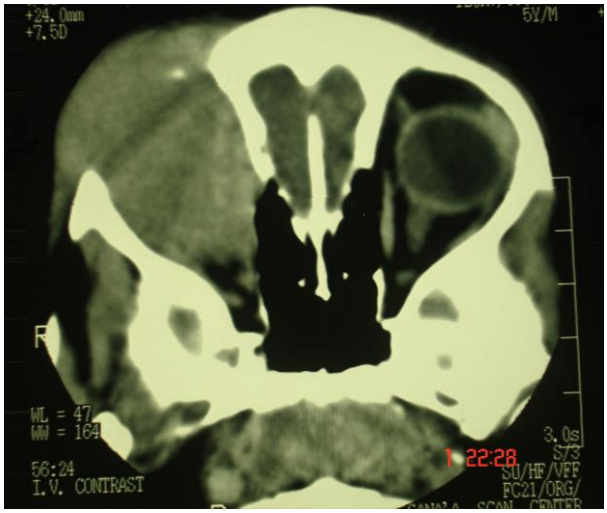

Figure 3
A large right orbital rhabdomyosarcoma in another female child aged 4 years, which was operable

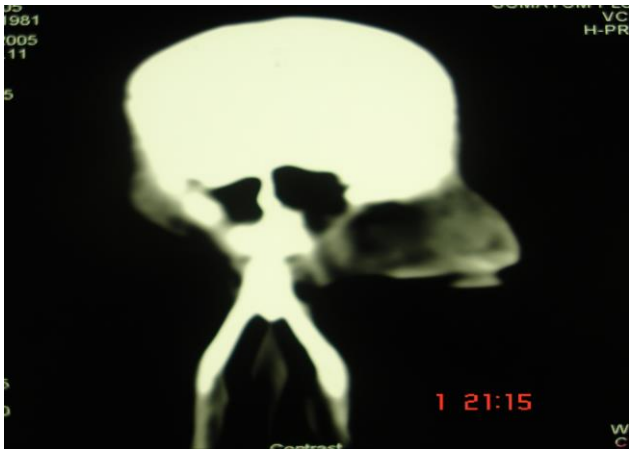

Figure 4 Teratoma 1week old girl with teratoma involving right orbit. CE axial CT reveals heterogenous mass, with central amorphous calcification (arrow) and hypodense droplets of lipid . Lesion enlarges right orbit, causing medial wall destruction superior orbital fissure expansion and massive eye proptosis.

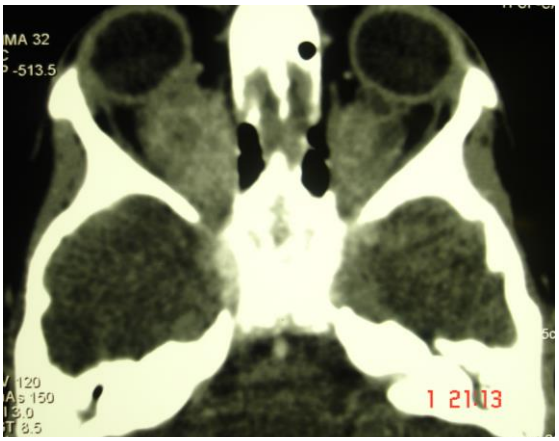

Figure 5 (A) Optic nerve glioma HRCT axial cuts of an 8 year old girl show markedly enhancing prominent fusiform enlargement of both optic nerve sheaths. Extending intracranially(B)

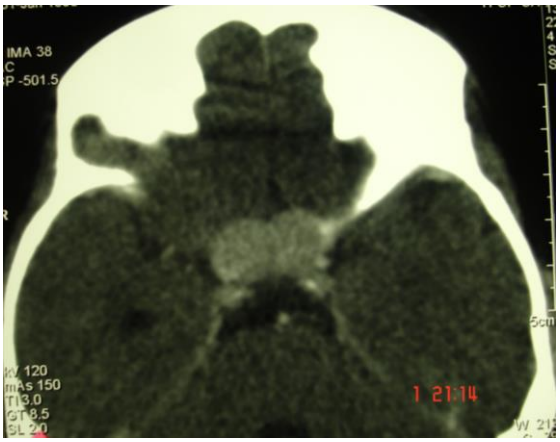

Figure 5 (B)optic nerve glioma ; intracranial extension.

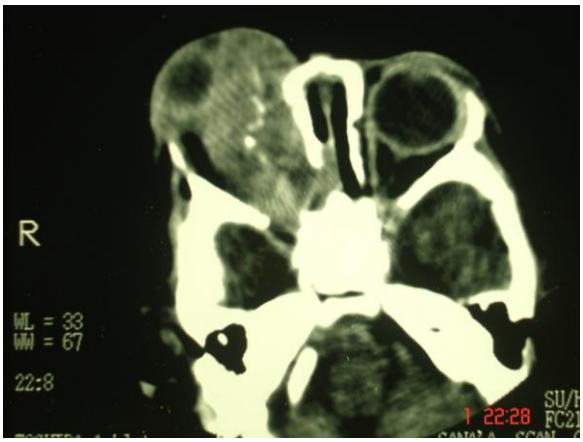

Figure 6 Another patient with optic nerve glioma
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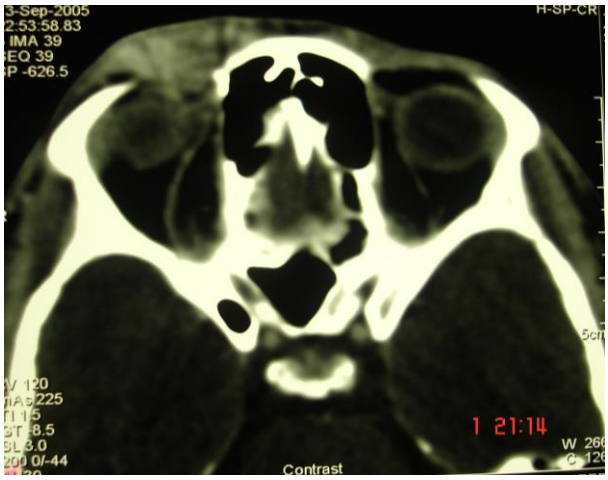

Figure7 (A) Capillary hemangioma
HRCT axial(A) and coronal (B)sections revealed an enhancing right pre and post septal hyperdense subcutaneous mass.

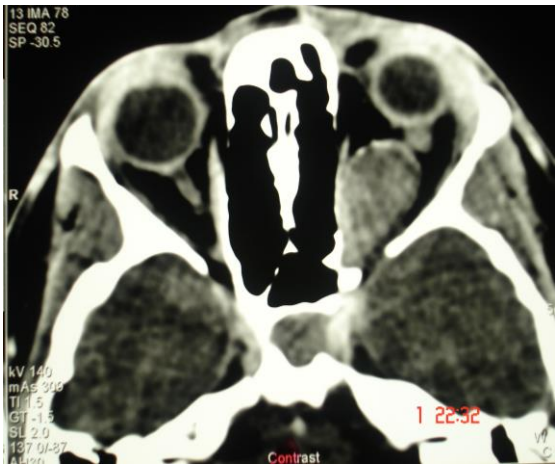

Figure 7 (B)

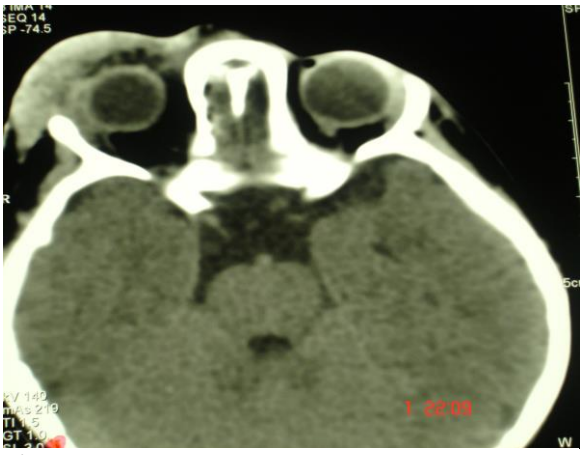

Figure 8
Another patient aged 3 years with capillary hemangioma

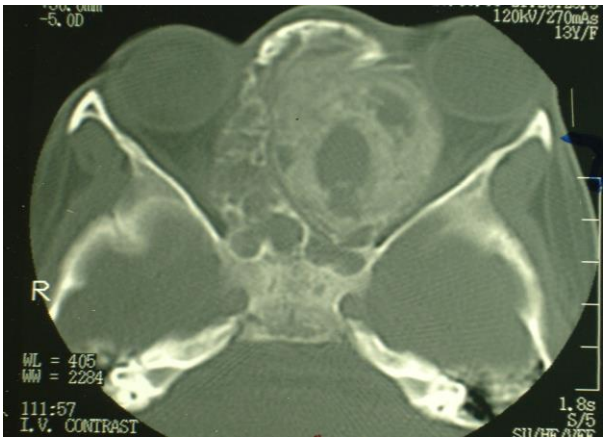

Figure 9 Ossifiying fibroma
13years old girl with ossifying fibroma involving nasal and paranasal sinuses and protruding in to the left orbit.

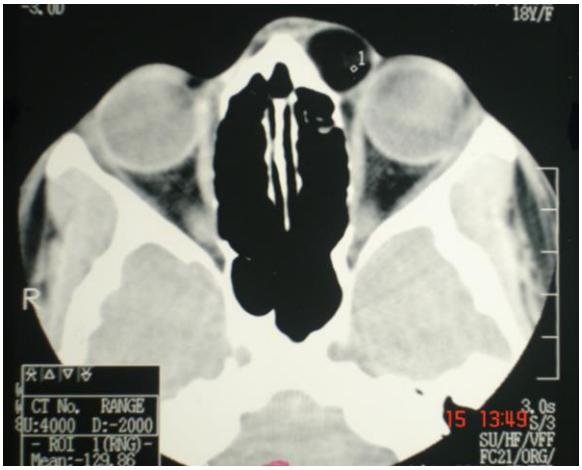

Figure 12
Dermoid. CT axial view demonstrate a low density (fat) circumscribed oval-shaped mass anteromedial to the globe in a 6 years old boy

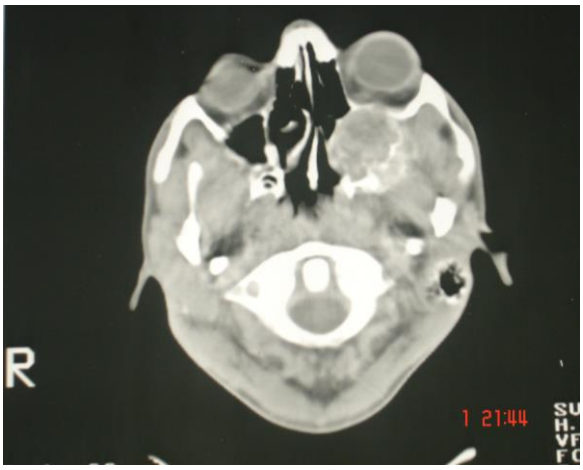

Figure 10 Shwanoma protruding on to the left oorbit

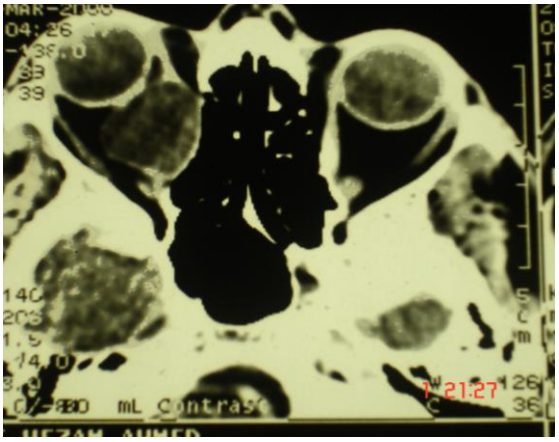

Figure 13 (A) Hydatid Cyst
An axial (A)and coronal(B) CT cuts at orbital level revealed : A well-defined relatively thick walled cyst (8-10HU) seen in right retrobulbar space producing proptosis

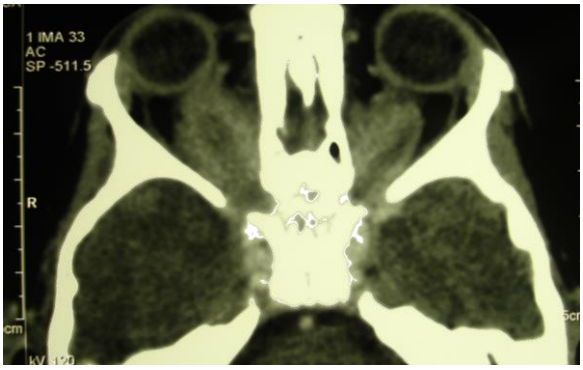

Figure 11 Bilateral nerve sheath meningioma Note enhancing sheath

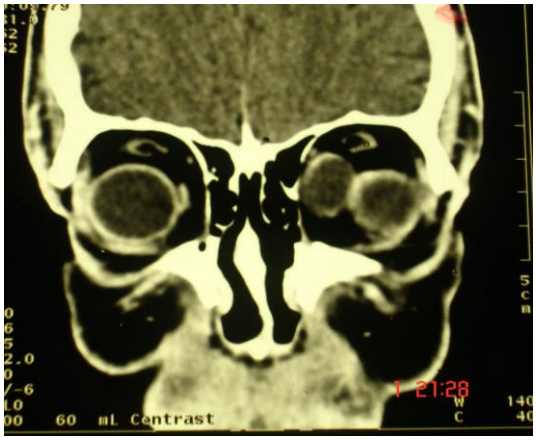

Figure 13 (B) Hydatid cyst

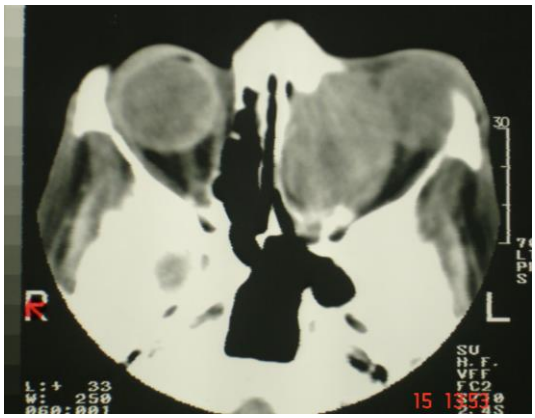

Figure14 (A) Mucocele

15years old boy with ethmoid mucocele extending into left orbit. CT shows expansion of left ethmoid sinus by a large, slightly dense, round mass eroding the lamia paperasia and protruding into the left orbit .

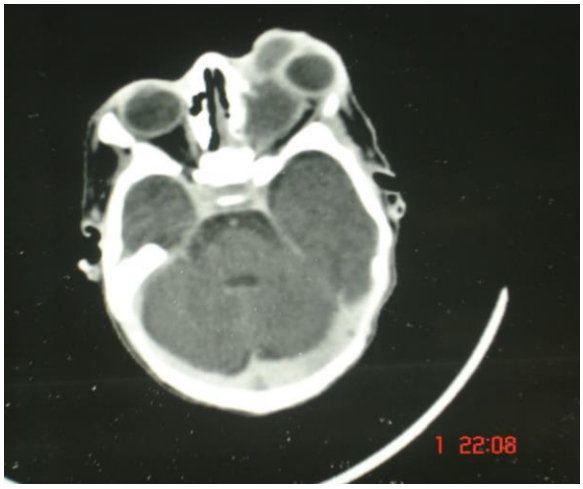

Figure 16 Axial orbital abscess

4weeks old girl with abscesses involving the left orbit. CE axial CT shows a retrobulbar mass with hypodense center and enhancing wall. Lesion extends anteriorly along medial wall of left orbit causing left eye proptosis.

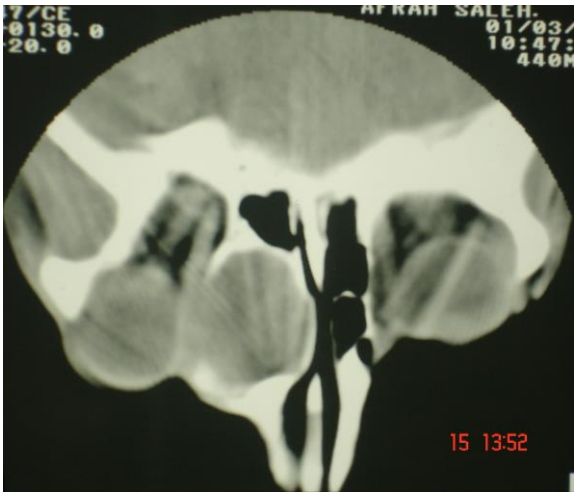

Figure 14 (B)Mucocele (coronal)

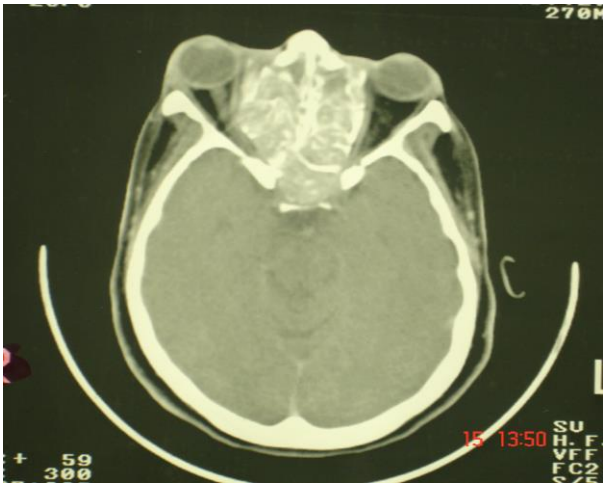

Figure 17 Aspirgillosis

15yrs old female with aspergilliosis involving the ethmoid and sphenoidal sinuses and scalloping the orbital medial wall

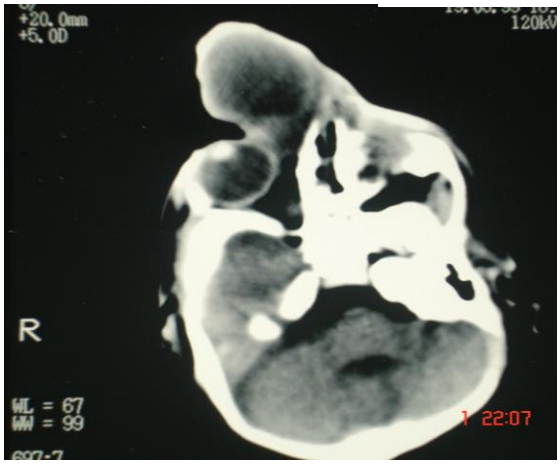

Figure 15 Meningocele

1week old boy with fronto-naso-orbital meningocele. CE axial CT shows a large, anterior herniation of meninges and low density CSF lesion extends through a fronto-naso-orbital bony defect.

Discussion

Orbito-ocular space occupying lesions in children include wide spectrum of pathology, which are challenging problems frequently faced by the radiologist and ophthalmologist alike.

The anatomic relations of the orbit to paranasal sinuses make it susceptible to pathology of these sinuses. In addition, tumor can extend to the orbit from infra-temporal fossa through infra temporal fissure and from intra-cranium through superior orbital fissure, that frequently compress the visual path ways. CT provides an excellent anatomic details and information regarding the presence, location as well as involvement of the orbit by lesions arising in an adjacent structure 1-3. The bony wall is best seen with HRCT and the presence of a well ossified bony wall and clear normal orbital fat along the margin of the lesion are extremely important CT land marks for staging and surgical planning. In children, incooperation and movement during CT examination as well as coronal imaging are also additional challenging problems in evaluating orbital lesions. However, the modern CT technology has resulted in short imaging time and in spinal scanners, we can obtain image in a fraction of a second4. In our study, a differential diagnosis of an orbital lesion was obtained by considering site, shape compartmental localization and density (attenuation) before and after administration of intra-venous contrast.

In this study malignant tumors ,namely retinoblastoma and rhabdomyosaroma, have a greater incidence compared with European and American series5-10 and this may be explained by the fact that these tumors have rapid growth and visual threat which encourage prompt referral than benign lesions. Conversely, the slow growth and the modest symptoms of benign lesions as well as the financial problems may explain the relatively low incidence of these masses in our study compared with fore-mentioned studies. Retinoblastoma should be differentiated from other benign lesions, that may simulate it and cause Leukorkoria11. However the presence of dense soft tissue mass involving the retina, extensive calcification and bilaterally or trilaterally are important differentiating CT criteria of retinoblastoma, thus, CT should be the primary imaging modality used in patients presenting with leukokoria12.

Rahbdomyosarcoma is the most common extra-ocular orbital malignant tumor in young children13 and It can spread intracranially, typically through orbital fissures into cavernous sinus, and this is best seen with HRCT14.

Orbital infections have become rare since the early and generalized use of antibiotics15. However, it is more common in our study and this can be attributed to neglected sinusal infections and health mal-education. In addition an immune compromised patient (diabetic) was presented with aspergelotic infection.

Cystic lesions of the orbit in children are frequently encountered in radiological departments. However, our number was relatively low compared to others16-19. As explained earlier Dermoid cysts represent the most common congenital lesions of the orbit20 and in our study they accounted for the majority of cystic lesions. Presence of fat contents or fat fluid level in a unilocular cyst is an important diagnostic CT criterion for dermoid. Hydatid cysts are mostly acquired in childhood and usually are not diagnosed until the third and fourth decades21,22. However in our study hydatid cyst was encountered in one patient aged 13 years. Cysts or cyst like lesions may arise from the adjacent structures and extend to the orbit and they are represented in our study by 3 cases mucoceles and 2 cases meningo-encephaloceles. However CT scan in sagittal and coronal plains illustrated well the bone defect in these lesions23,24.

Vascular lesions in our study mostly represented by capillary hemangioma with relatively low incidence as compared to other reports for the same reasons

discussed earlier. Optic nerve and its sheath tumors are mostly represented by glioma and this corresponds well to others5,9

Meningioma, shwanoma, and ossifying fibroma were all encountered in our study although they are rarely found in children25-28

Finally, the late presentation of patient of malignant tumors can explain the high incidence of advanced stages in our study and the socioeconomic condition may be responsible. In this study in Yemeni children we found that HRCT provided valuable information for evaluating orbital masses as regards their exact location, extent and proper management planning and its sensitivity can reach 100%.

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