ORIGINAL ARTICLE

CT-Scan Findings of Orbital Mass Among Pediatric Patients at a Tertiary Care Hospital in Bangladesh

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ABSTRACT

Background: CT-Scan for the detection of orbital mass among pediatric patients is very important noninvasive radiological modality. The purpose of the study was to find out CT-Scan findings of orbital mass among pediatric patients in a tertiary care hospital.

Methodology: This is a cross sectional study was carried out in Ophthalmology and Radiology and Imaging department of National Institute of Ophthalmology (NIO) from January 2012 to December 2013. All the patient below 18 years of age presented with suspected orbital mass at Ophthalmology and Radiology and Imaging department of NIO and performed CT- Scan of orbit for diagnosis of the disease and also done histopathology after operation was enrolled in this study.

Results: In this study it was observed that a total of 29 cases identified as malignant evaluated by CT, among them 27 cases were true positive and 2 cases were false positive. Benign was found in 41 cases evaluated by CT scan, out of which 1 false negative and 40 cases were true negative. The sensitivity in diagnosis of orbital tumor by CT was 93.3%.

Conclusion: The sensitivity in diagnosis of orbital tumor by CT was high and is a useful method in the differentiation between benign and malignant orbital mass.

Key Words: Orbital mass; CT-Scan findings

Introduction

A wide spectrum of orbital pathology is seen in the paediatric population.The orbital masses can be categorized based on their tissue of origin, tumors of mesenchymal origin (rhabdomyosarcoma, histiocytosis, leukemia and lymphoma), neural origin tumors (retinoblastoma, optic nerve glioma, meningioma, schwannoma, neurofibroma and neuroblastoma) and vascular malformations or tumors (Barnes et al. 1996).[1]

Retinoblastoma is the most common intraocular tumor in children. Mean age is 2 years in unilateral forms (60% of cases) and 1 year in bilateral forms (Moll et al. 1997 and Doz et al. 2004)[2,18]. Imaging plays a crucial role for intracranial tumor extension, possible midline intracranial primitive neuroectodermal tumor (PNET) and brain malformations in patients with 13q deletion syndrome (Baud et al.[3]; Ballarati et al.[20] and Rodjan et al.[4]). PNETs are associated with hereditary retinoblastoma, a combination known as trilateral retinoblastoma, which occurs in 5-15% of children in the hereditary subgroup (Rodjan et al. 2010[4]). CT detection of calcifications in retinoblastoma has a sensitivity of 81-96%, and an even higher specificity (Beets-Tan et al. 1994).[5] Rhabdomyosarcoma. CT can

differentiate between primary orbital rhabdo myosarcoma and that arising from paranasal sinuses, also demonstrate intracranial, epidural and subarachnoid extension of the tumour (Zimmerman, Bilanuik and Littman 1978).[7] Lymphangioma are most often extraconal in location. On CT they are diffuse hyperdense masses with irregular margins and show mottled contrast enhancement (Atlas et al. 1987).[8]

On CT dermoid cysts have well defined margins and appear cystic, with the density of their center ranging from CSF to that of fat (Chawda and Moseley 1999).[9] Lymphoma: on CT the lacrimal gland lymphoma appears as a hyperdense mass in the lacrimal fossa that shows enhancement on contrast administration.

Langerhans cell histiocytosis is isolated form (eosinophilic granuloma) often involves the orbital bones. CT appearance of a well-defined punched out lytic lesion with bevelled edges is characteristic. The associated soft tissue mass can encroach on the orbit or the brain, can be well-defined or diffusely infiltrative and has a nonspecific appearance on CT or MRI (Kandpal et al. 2006)[10]. Fibrous dysplasia occurs mainly in children and young adults and frequently involves the orbit. CT reveals characteristic ground glass appearance and expansion of the involved bone (Kandpal et al. 2006).[10]

Optic Nerve Glioma : Thirty per cent cases are associated with neurofibromatosis Type 1 (NF1) (Azar-Kia et al. 1987).[11] The tumor causes diffuse thickening, kinking or fusiform enlargement of the nerve, with moderate to marked enhancement which is generally less than that seen with meningioma (Gorospe et al. 2003).[12] On CT showing intracanalicular, chiasmatic or retrochiasmatic tumor extension (Mafee, Inoue and Mafee 1996).[13] Plexiform neurofibroma, on imaging it appears as an ill-defined, infiltrative, soft tissue mass which may involve the retrobulbar fat, the extraconal space including eyelid and adjacent subcutaneous tissue]. The other orbital associations of NF1 include presence of optic pathway gliomas (in 15 to 40% cases) (Davidson, Dermoid and Epidermoid 2004)[14] Secondary tumors or orbital metastasis in children is most commonly seen with neuroblastoma, in as many as 20% cases (Callizo 2000).[15] CT or MRI reveals an aggressive infiltrative soft tissue mass associated with bone destruction. Orbital metastasis may occasionally be seen in patients with Ewings sarcoma and RMS, Leukemia (Hooper, Sherman and Boal 1991). Leukemic infiltration is seen as optic nerve enlargement and enhancement on imaging (Vazquez et al. 2002).[17]

Methodology

This was a cross-sectional type of study. This study was carried out in the Department of Radiology & Imaging in collaboration with department of ophthalmology of NIO (National Institute of Ophthalmology. All the patient below 18 years of age preseented with suspected orbital mass at ophthalmology department and attending department of radiology and imaging also from NIO (National Institute of Ophthalmology), Dhaka and perfomed CT scan of orbit for diagnosis of the disease and also done histopathology after operation was enrolled in this study. CT scan is preferred due to the speed and easy availability of this modality. Subtle or small calcifications are well shown, and artifact from eye motion is minimized. Iodinated intravenous contrast may be used to assist in characterising a mass, although it is not usually necessary as intraorbital fat provides excellent natural contrast. CT scan should be performed with contiguous thin section (2-3mm) axial, and thicker section (4-5mm) coronal images, displayed in both soft-tissue and bone windows. Alternatively, coronal and oblique sagittal projections may be reformatted from axial images; the resultant image quality is improved by using overlapping sections to avoid `stair-step' artifact (e.g. 3mm thick, 2mm table increments). Evaluation of the orbital apex may require very thin section overlapping coronal images (1.5mm) thick, with 1 mm table motion), so that 3D reconstructions may be made. Spiral CT scanning, if available may also be used to provide the same data set for subsequent reconstructions. For this study 3rd generation Siemens 6 slice CT scan machine is used. Slice thickness is 2-3mm ,axial cut with sagittal and coronal reconstructions are done.

Statistical Analysis

Statistical analysis of the results were obtained by using window based computer software devised with Statistical Packages for Social Sciences (SPSS-16). The results were presented in tables, figures, diagrams. For the validity of study outcome, sensitivity, specificity, accuracy, positive predictive value and negative predictive value of CT scan evaluation for orbital mass lesion in pediatric population was calculated.

Prior to the commencement of this study, the research protocol was approved by the Ethical review committee of BSMMU, Dhaka.

Results

A total number of 70 patients clinically suspected cases orbital mass lesion who came in department of Ophthalmology, Radiology & Imaging of National institute of ophthalmology (NIO), Dhaka, during the period of January 2012 to December 2013, were included in this study.

Table I: Distribution of the study patients by CT scan (n=70)

	Number of patients	Percentage
Malignant		
Retinoblastoma	9	12.9
Optic nerve glioma D/D meningioma/ haemangioma	8	11.4
Rhabdomyosarcoma D/D eosinophilic granuloma, sarcoma	2	2.8
Rhabdomyosarcoma D/D Lymphoma	2	2.8
Lymphoma	3	4.3
Eosinophilic granuloma D/D haemangio pericytoma	2	2.8
Haemangioma D/D haemangio pericytoma, ,eosinophilic granuloma	1	1.4
Sarcoidosis D/D peudotumour	2	2.8
Benign		
Dermoid	15	21.4
Neurofibroma	9	12.9
Pseudo tumour D/D Sarcoidosis	3	7.1
Haemangioma D/D schwannoma	3	7.1
Schwannoma D/D maningioma	1	1.4
Peudotumour	2	2.8
Meningioma D/D Haemangioma	3	4.3
Haemangioma D/D meningioma	3	4.3
Schwannoma D/D haemangioma	2	2.8

Table I shows CT scan of the study patients it was observed that, in malignant cases majority 9(12.9%) patients done retinoblastoma followed by 8(11.1%) optic nerve glioma D/D meningioma, haemangioma. In benign cases, 15(21.4%) patients have dermoid followed by 9(12.9%) patients have-neurofibroma.

Table II: Distribution of the study patients by histopathology (n=70)

Histopathology	Number of patients	Percentage
Malignant		
Retinob lastoma	9	12.9
Optic nerve glioma	8	11.4
Rhabdomyosarcoma	5	7.1
Non hodgkin lymphoma	3	4.3
Eosinophilic granuloma	2	2.8
Haemangio pericytoma	1	1.4
Benign		
Dermoid	15	21.4
Neurofibroma	9	12.9
Pseudotumour	7	10.0
Haemangioma	6	8.6
Schwannoma	5	7.1

Table II shows histopathology of the study patients it was observed that, in malignant cases 9(12.9%) patients had retinoblastoma, 8(11.4%) had optic nerve glioma and 5(7.1%) had rhabdomyosarcoma.

In benign cases, 15(21.4%) patients had dermoid, 7(10.0%) had psedotumour, 6(8.6%) had haemangioma and 5(7.1%) had schwannoma.

Table III: CT findings of retinoblastoma (n=9)

CT finding of retinoblastoma	Number of patients	Percentage
Density		
Isodense	1	11.1
Hyperdense	8	88.9
Proptosis		
left eye	5	55.6
Right eye	1	11.1
Both eye	3	33.3
Muscle involvement		
LR, IR	1	11.1
No involvement	8	88.9
Retinal detachment	4	44.4
Optic nerve invasion	5	55.6
Contrast enhancement	1	11.1
Calcification	6	66.7

Table IV: CT findings of dermoid (n=15)

CT findings of dermoid	Number of patients	Percentage
Right orbit	8	53.3
Left orbit	7	46.7
Fat density	15	100.0
Proptosis	5	33.3

Table V: CT findings of lymphoma (n=3)

CT findings of lymphoma	Number of patients	Percentage
Density		
Soft tissue	2	66.7
Hypodense	1	33.3
Proptosis	2	66.7
Enhancement	2	66.7
Lacrimal gland involvement	1	33.3

Table VI (1): CT findings of pseudotumour(n=7)

CT findings of lymphoma	Number of patients	Percentage
Extraocular muscle involvement	6	85.7
Intraconal and extraconals pace involvement	3	42.9
Proptosis	6	85.7

Table VI (2): CT findings of neurofibroma (n=9)

CT findings of neurofibroma	Number of patients	Percentage
Density		
H yperdense	6	66.7
Isodense	0	66.7
Proptosis	8	88.9
Muscle		
LR	9	100.0
SR	6	66.7
IR	3	33.3
Enhancement	6	66.7

Table VII: Comparison between CT scan and Histopathology (n=70)

CT scan		Histopathology Malignant (n=28)		Benign (n=42)	
		N	%	N	%
Malignant	n=29	27	96.4	2	4.8
Benign	n= 41	1	3.6	40	95.2

Table VII shows CT scan malignant was found 27(96.2%) in malignant histopathology and 2(4.8%) in benign histopathology. CT scan benign was found 1 (3.8%) in malignant histopathology and 40(95.2%) in benign histopathology.

Table VIII: Sensitivity, specificity, accuracy, positive and negative predictive values of the CT scan evaluation for orbital mass lesion in pediatric population

Validity test	Percentage
Sensitivity	96.4
Specificity	95.2
Accuracy	95.7
Positive predictive value	93.1
Negative predictive value	97.6

Table VIII: Sensitivity, specificity, accuracy, positive and negative predictive values of the CT scan evaluation for orbital mass lesion in pediatric population

Discussion

This cross sectional study was carried out with an aim to evaluate the demographic profile of the patients having orbital mass lesion in pediatric population study population and to evaluate the association between CT scan finding with histopathological correlation and along with its validity tests by calculating sensitivity, specificity, accuracy, positive predictive value (PPV) and negative predictive value (NPV) respectively of CT scan in diagnosis of pathological variety of orbital mass.

A total of 70 patients under 18 years of age preseented with suspected orbital mass at ophthalmology and Radiology and Imaging department of National Institute of Ophthalmology (NIO), Dhaka during January 2012 to December 2013, were included in this study. Patients above 18 years of age, patient's not undergoing histopathology and patients' guardian not given consent were excluded from the study. CT scan evaluation of orbit for diagnosis of the disease and all patients underwent histopathology after operation. The present study findings were discussed and compared with previously published relevant studies.

Optic nerve glioma usually occurs in childhood (median age two to six years). In this age group, optic nerve glioma is typically a slow-growing tumor which presents with proptosis and visual loss (Azar-Kia et al. 1987).[11] In this series it was observed that majority (35.7%) patients having orbital mass lesion age belonged to≤5 years and the mean age was found 10.03±5.91 years varied from 8 months to 18 years. Moll et al. (1997)[2] and Doz et al. (2004)[18] reported in their study that mean age at clinical presentation was 2 years in unilateral forms (60% of cases) and 1 year in bilateral forms. In another study median age at diagnosis of retinoblastoma was 5 months (range, 0 to 29 months); age at diagnosis was younger among 47 children (47%) with familial retinoblastoma compared with age at diagnosis among 52 children (53%) with sporadic retinoblastoma (2 months vs 6.5 months, P <0.001) obtained by Kivela (1999).

Primary orbital rhabdomyosarcoma most often occurs in the first decade of life, with a mean patient age of 6-8 years obtained by Shields et al. (2004), Font et al. (2006), Shields and Shields (2003).[10,12]

Bidar et al.[20] reported in their study that the median age at diagnosis of orbital leukemic tumor was 8 years varied from 1-18 years. Majority of the above findings from other studies are in agreement with the present study regarding the age incidence of orbital mass lesion. In this study it was observed that orbital mass lesion was more common in male children, which were 55.7% and 44.3% patients were male and female respectively. Male to female ratio was 1.3:1. Shields et al. (2001)[19], Shields and Shields (2003)[12] and Conneely and Mafee (2005)[21] mentioned in their report that there is a slight male predilection, with a male-to-female ratio of 5:3, which are comparable with the current study.

In this present series it was observed that majority (74.1%) of the patients had decrease vision of eye followed by 34.3% had proptosis, 24.2% had swelling upper eyelid, 11.4% had swelling of eye. In this current series it was observed that 38.6% patients had decrease in left eye vision, 28.6% had decrease in right eye vision, 7.1% had decrease in both eye vision and 25.7% had normal vision. In this present study it was observed that 4.3% patients had both white eye, only 1.4% had white eye right side, 1.4% had white eye left and 92.8% patients had normal eye. In this current study it was observed that 28.6% patients had left eye proptosis, 21.4% had right eye proptosis, 2.8% had both eye proptosis and 47.1% had no proptosis. Lacrimation of the study patients it was observed that, 7.1% had right eye lacrimation, 11.4% left eye lacrimation and 81.5% had no lacrimation. Edematous eye ball was found in 1.4% patients, swelling right eye 2.9%, swelling left eye 1.4%, downward deviation of left eye 2.9%, Downward deviation of right eye 1.4%, dropping of left upper eyelid 1.4% and upward deviation of left eye 2.9% and 85.7% had normal eye.

Regarding the extra ocular muscle it was observed that, normal extra ocular muscle was found 64.5%, left SR, LR involved in 11.4%, all involved in right eye 5.7%, All involved in left eye 5.7% and right LR involved 5.7%. In this series it was observed that 88.6% patients had normal retina, 8.6% retinal involvement in left eye, 1.4% retinal involvement on both side L>R and 1.4% had retinal involvement in right eye.

In this study it was observed that CT scan identified 29 malignant cases, among them 12.9% patients had retinoblastoma followed by 11.1% optic nerve glioma D/D meningioma, haemangioma, 4.3% had Lymphoma and others malignant varied from 1.4% to 2.8%. In benign cases, 21.4% patients had dermoid and 12.9% had neurofibroma. Lecompte and Langelier (1994) present in their study that the most commonly encountered tumours: uveal melanoma (observed in 50 cases), lymphoma (in 8), optic glioma (in 6), meningioma (in 6), dermoid cyst (in 5) and metastasis (in 5). In this present series it was observed that histopathology identified 28 malignant cases, among them 12.9% had retinoblastoma, 11.4% had optic nerve glioma and 7.1% had rhabdomyosarcoma. A total of 42 benign cases was identified by histopathology among them 21.4% patients had dermoid, 10.0% had psedotumour, 8.6% had haemangioma and 7.1% had schwannoma.

In this current series it was observed that a total of 29 cases identified as malignant evaluated by CT,

among them 27 cases were true positive and 2 cases were false positive. Benign was found in 41 cases evaluated by CT scan, out of which 1 false negative and 40 cases were true negative. In this present study it was observed that sensitivity 96.4%, specificity 95.2%, accuracy 95.7%, positive predictive values 93.1% and negative predictive values 97.6% of the CT scan evaluation for orbital mass lesion in pediatric population. On CT, retinoblastoma is typically a mass of high density compared with the vitreous body, usually calcified and moderately enhancing after iodinated contrast medium administration. Beets-Tan et al. (1994)5 mentioned in this study CT detection of calcifications in retinoblastoma has a sensitivity of 81-96%, and an even higher specificity. In another study (Eisen et al. 2000) reported that six or more positive criteria predicted invasion with 67% sensitivity and 80% specificity (accuracy, 72%). CT was more accurate than MRI. Invasion of the nasolacrimal system was predicted accurately (89%). Features such as panorbital involvement, orbital fat, frontal sinus opacity, molding around orbital structures, perineural involvement, and fat stranding had specificity of 97% to 100%, but low sensitivity (Ben et al. (2005). The above study findings are closely resumbled with the current study. The clinical manifestation and characteristics of CT image of 117 cases of orbital tumors in our hospital were investigated. The hemangioma had the highest incidence, and the less common tumors were, in sequence of incidence, pseudotumor, dermoid cysts, neurilemmoma, polymorphous adenoma and meningioma. The sensitivity in diagnosis of orbital tumor by CT was 93.3%. The coincidence of CT histological diagnosis with pathology were 83.3%, 82.6% and 71.4% for dermoid cysts, hemangioma, and pseudotumor respectively, but the general coincidence of CT histological diagnosis with pathology was only 67.8%. When CT was combined with ultrasound, cytological examination and clinical manifestations, the accuracy of histological diagnosis could be improved to 83.3%.

Conclusion

This study was undertaken to evaluate the CT scan finding of orbital mass in pediatric population with histopathological correlation. It can be concluded that CT scan is a useful method in the differentiation between benign and malignant orbital mass. Moreover CT scan is cost effective, available, not time consuming and real time image. To see white eye, lacrimation, proptosis, eye ball, extra ocular muscle and retina lesions are best seen in CT, which help surgeon for operative plan and decreases patient's morbidity and mortality.

Conflict of Interest: There is no conflict of interest to any of the authors.

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